113 年年會 海報論文展示：病例報告
目錄

病例報告 113_C1

貌似氣胸的肺大泡：一個病例報告

Giant bulla mimicking pneumothorax: a case report

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Introduction

A bulla is an air-filled space in the lung > 1 cm in diameter. A giant bulla refers to bullae occupying at least 30% of one side of the chest. A giant bulla may exist alone or with other bullae formation. Differentiating a giant bulla or a pneumothorax is challenging and can result in misdiagnosis and improper management. Here, we present a case of a young man with shortness of breath due to a giant bulla.

Case Report

A 24-year-old young male with no previous medical history or congenital disorders, except for a 10-year habit of smoking half a pack of cigarettes daily, presented with progressive shortness of breath over the past six months. The patient denied any fever, body weight loss, cough, sputum production, chest pain, or chest tightness. Physical examination revealed an increased respiratory rate (22/min) and decreased tactile fremitus, hyperresonance on percussion, and diminished breath sounds on the right side. No wheezing was heard. A chest X-ray (Figure 1) showed an enlarged right lung with displacement of the heart, significantly reduced lung markings on the right side, but no visible pleural line. A CT scan (Figure 2) confirmed the presence of a giant bulla. Due to the significant size of the bulla and the resulting respiratory symptoms, the patient consented to surgical treatment. Postoperative follow-up showed good recovery with no recurrence.

Discussion

Giant bullae typically develop due to cigarette smoking, but can also be idiopathic, associated with marijuana smoking, HIV infection, intravenous drug use, genetic disorders such as Marfan syndrome and Loeys-Dietz syndrome, and have been reported with COVID-19 infection.

Mistaking giant bullae to pneumothorax can lead to serious consequences, such as unnecessary chest tube placement, which can cause additional complications including infection, bleeding, and further lung injury. This misdiagnosis can also result in delaying treatment for the actual condition, and potentially worsening the patient's respiratory status.

Giant bullae and pneumothorax can be differentiated by imaging characteristics. On chest radiographs, a giant bulla typically shows a concave pleural line relative to the chest wall, while a pneumothorax presents a convex pleural line. In addition, lung markings extending to the chest wall indicate that it is not a pneumothorax. Chest CT scans provide clearer differentiation, with giant bullae often located in the upper lobes and subpleural areas, whereas pneumothorax shows no such pattern.

Conclusion

Identifying giant bullae and pneumothorax is essential because their treatments are significantly different, although their presentation in physical examinations and chest X-rays is similar. Pneumothorax tends to present more acutely and is often accompanied by chest pain. Careful evaluation can reduce the risk of iatrogenic injury.

病例報告 113_C2

一位甲狀腺毒性心肌病變經由甲狀腺切除術和標準心衰竭藥物治癒的病例報告

A case report of thyrotoxic cardiomyopathy cured by thyroidectomy and guideline-directed medical treatment of heart failure

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Introduction

Although the most common cause of heart failure (HF) is ischemic heart diseases, other less common causes such as hyperthyroidism (thyrotoxicosis) should also be considered during diagnosis to improve overall clinical management of HF. Hyperthyroidism is a potentially reversible and curable cause of thyrotoxic HF, which should be excluded in every new patient with HF especially in the absence of coronary artery disease.

Case Report

A 65-year-old woman with a history of hypertension presented with a 3-week history of dyspnea, palpitation and lower legs edema. On physical examination, there were rapid irregular cardiac beats, grade 4 systolic murmur over left lower sternal region, jugular vein engorgement and palpable mass of right neck. Initial electrocardiography revealed atrial fibrillation with rapid ventricular response. Lab tests showed elevated b-type natriuretic peptide level of 662 picogram per milliliter (reference value, <100 picogram per milliliter), free T4 level of 3.13 ng per deciliter (reference value, 0.70-1.48 ng per deciliter) and low level of TSH below 0.0025 uIU per milliliter (reference value, 0.3500-4.9400 uIU per milliliter). A transthoracic echocardiogram demonstrated bilateral atrial enlargement, pulmonary hypertension and generalized hypokinesia of left ventricle, which was considered heart failure with reduced ejection fraction. Coronary angiography excluded significant coronary artery disease. Subsequent computed tomography showed a large thyroid mass extending into anterior mediastinum, along with compression of both main trachea and upper esophagus. The patient underwent unilateral thyroidectomy and guideline-directed medical treatment (GDMT) for heart failure, including beta-blocker, angiotensin receptor blocker, mineralocorticoid receptor antagonist and sodium-glucose cotransporter 2 inhibitor. A follow-up electrocardiography on 6th month after surgery showed normal sinus rhythm accompanied with restored left ventricular systolic function on echocardiography.

Discussion

Thyrotoxicosis, which is the set of signs and symptoms derived from the excess of circulating thyroid hormones, leads to increased cardiac output and decreased systemic vascular resistance, increasing the volume of circulating blood and causing systolic hypertension. In addition, the shortening of the refractory period of cardiomyocytes produces sinus tachycardia and atrial fibrillation, which leads to heart failure. Approximately 1% of patients with thyrotoxicosis develop thyrotoxic cardiomyopathy, a rare but potentially fatal form of dilated cardiomyopathy.

Conclusion

This case reminds clinicians that heart failure should be considered in patients with thyrotoxicosis, especially among those with low cardiovascular risk. Adequate surgical intervention and GDMT could accelerate the recovery timeline for both heart failure and rhythm control effect for atrial fibrillation.

病例報告 113_C3

以深部靜脈栓塞及肺栓塞臨床表現的攝護腺瀰漫性大 B 細胞淋巴瘤：一病例報告

Diffuse large B-cell lymphoma of the prostate presenting as deep vein thrombosis and pulmonary embolism --a case report

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Introduction

Primary lymphoma of the prostate is rare and the diagnosis may be difficult without tissue proof. We report the case with initial presentation of deep vein thrombosis and pulmonary embolism which prompted the survey of malignancy and further management.

Case Report

We reported a 70-year-old male who had history of hypertension, type 2 diabetes mellitus and hyperlipidemia for years with medical treatment at clinic. His surgical history includes perforated peptic ulcer s/p subtotal gastrectomy more than 40 years ago. He was brought to our emergency department due to progressive swelling of right lower limb for about two weeks followed by syncope, dizziness, dyspnea and palpitation since one day ago. Electrocardiogram showed sinus tachycardia with non-specific ST-T change and laboratory data found mildly abnormal cardiac enzymes. He was admitted to intensive care unit under the impression of deep vein thrombosis with acute coronary syndrome. Antiplatelet and anticoagulation therapy was applied. Coronary angiography found no obstructive lesions, but chest CT angiography confirmed the presence of a saddle pulmonary embolism at the bifurcation of the pulmonary trunk, extending into the left and right pulmonary arteries. Physical exam found enlarged prostate and abdominal CT scan revealed same finding alone with enlarged lymph nodes in the paraaortic region. Urologist was consulted and trans-urethral prostectomy was performed to relieve obstructive nephropathy. Pathology of the prostate specimen revealed fragments of prostatic tissue with infiltration of large atypical lymphoid cells. A diffuse large B-cell lymphoma is considered. Germinal center B-cell subtype was confirmed by immunohistochemical staining. Oncologist was consulted and he discharged with further management.

Discussion

Approximately one third of non-Hodgkin lymphomas occur in extranodal sites, but primary involvement of the prostate is rare. Less than 1% of prostate specimens demonstrate leukemia or lymphoma, and most are chronic lymphocytic leukemia or small lymphocytic lymphoma. On the other hand, venous thromboembolism is a common complication for lymphoma, especially among diffuse large B-cell lymphoma as the most prevalent non-Hodgkin lymphoma subtype. Oral direct oral anti-coagulants remain the treatment of choice for thromboembolism, along with standard management options for lymphoma. Germinal center B-cell subtype is the most curable cell subtype.

Conclusion

The cause of DVT/PE should be carefully evaluated to avoid incomplete management. Prostate lymphoma can be diagnosed by pathology and proper treatment can increase survival rate and improve life quality.

病例報告 113_C4

老年病患之單側肺動脈閉鎖：一病例報告

Unilateral pulmonary artery atresia accidentally found in a senior patient —a case report

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Introduction

Unilateral pulmonary artery atresia (UPAA) is a rare condition most commonly found during childhood. Adult patient may seek medical management due to non-specific pulmonary symptoms and diagnosed by careful evaluations. We reported a senior case accidentally found this problem during hospitalization course.

Case Report

This 75 y/o man has history of type 2 diabetes mellitus, hypertension, chronic kidney disease, benign prostate hypertrophy and gout for years which have been treated regularly at our hospital. He visited our ER for help because of erythematous swelling over left lower limb for one week. Physical examination found tenderness over left calf but no open wound or local heat. CXR showed prominent perihilar vasculature with interstitial infiltration over bilateral lung fields suspicious for pulmonary congestion with cardiomegaly. Vascular duplex of left lower limb was performed and revealed thrombus in left common femoral vein and left superficial femoral vein. Lower limb CT venography found poor contrast opacification with enlarged diameter of left femoral vein and edematous change with skin thickening of left lower extremity. He was admitted under the impression of acute deep vein thrombosis and anticoagulation was initiated. Dyspnea on exertion was noted and arterial blood gas showed hypoxemia. Tc-99m MAA perfusion scan was arranged for the possibility of pulmonary embolism which revealed large(>80%) wedge-shape segmental perfusion defect in the Left lung field and RUL. The perfusion ratio of the left/right lung field from the posterior view is 3%:97%. CTA for pulmonary artery was further performed and found sharp tapering of left main pulmonary artery with small caliber of arterial branches r/o stenosis. 3-dimentional reconstruction of pulmonary artery was done and compared with pulmonary angiography. Left side UPAA was impressed. Tracing back his history he denied specific symptoms regarding the diagnosis except shortness of breath treated by bronchodilators upon demand. His symptoms subsided by medications and pulmonary function test showed mild restrictive pattern. The patient discharged and kept outpatient management.

Discussion

UPAA is a congenital defect possibly caused by failure of the embryonic sixth aortic arch to fuse with the pulmonary trunk during development. Literature review reported right side is more common than left while the latter is often associated with other congenital cardiovascular anomalies such as tetralogy of Fallot, ventricular septal defect, patent ductus arteriosus or right side aortic arch. However our patient did not present with any of these anomalies. Patients with isolated UAPA are usually asymptomatic or suffered from recurrent respiratory infections, chronic cough, dyspnea, or hemoptysis. Management should be tailored by severity of the complications

among them pulmonary hemorrhage is the worst that surgical pneumonectomy may even be required. Our patient remained stable with mild dyspnea so medical treatment is sufficient considering his other co-morbidities.

Conclusion

UPAA may keep asymptomatic until later stage of life and be found during clinical evaluation. CTA with pulmonary angiography are appropriate tools to confirm the diagnosis. Treatment options depend on individual's presentations and underlying conditions.

病例報告 113_C5

伏立康唑治療慢性空洞性肺部麴菌疾病期間發生之嚴重低血鈉症 - 一項病例報告

Voriconazole-induced Severe Hyponatremia in Treatment during Chronic Cavitary Pulmonary Aspergillosis - A Case Report

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Introduction

Chronic cavitary pulmonary aspergillosis (CCPA) is a progressive pulmonary fungal infection with poor prognosis that typically occurs in immunosuppressed patients. Voriconazole is often used to treat CCPA, and severe hyponatremia is a rare but serious adverse side effect of the drug, which can lead to consciousness disturbance and even mortality. Here, we present a case of a 77-year-old patient who developed severe hyponatremia during voriconazole treatment for CCPA.

Case Report

A 77-year-old man with a history of lung abscess post-lobectomy with residual lung cavitation, coronary artery disease under Clopidogrel, and COPD and asthma overlapping under inhaled corticosteroid and long-acting bronchodilator therapy, experienced a persistent cough and increased sputum for 2 months. CCPA was diagnosed according to high levels of serum Aspergillus fumigatus antibodies and a progressive left upper lung cavitary lesion with fungal balls on serial chest image studies. He started oral voriconazole treatment and developed slurred speech and dizziness, progressing into severe disoriented and stupor after one week of voriconazole treatment. After general survey, including brain MRI, serum biochemistry survey, revealed a rapid decline in serum sodium level, leading to severe hyponatremia, which contributed to his consciousness disturbance. Voriconazole-induced SIADH was considered likely after exclusion other causes. His consciousness improved within 2 days after discontinuation of voriconazole and administration of 3% saline. Serum voriconazole levels later revealed a supratherapeutic concentration (5.7 μg/mL).

Discussion

Severe hyponatremia is a rare adverse event of voriconazole, which is recoverable with timely awareness and management, but severe morbidity and mortality have been reported. Previous case reports have indicated that voriconazole-induced hyponatremia may be associated with SIADH or salt-losing nephropathy (SLN), although the mechanism remains unknown. Currently, there are no documented cases of hyponatremia related to other triazole antifungals, suggesting that this reaction may be idiosyncratic to voriconazole. The metabolism of voriconazole via CYP2C19 may play a significant role, and the use of CYP2C19 inhibitors may result in achieving supratherapeutic concentrations more easily. With nearly 60% of Asians carrying at least one non-functional allele, a review of drug-drug interactions may help reduce the risk of severe adverse effects.

Conclusion

Voriconazole-induced hyponatremia is a rare adverse effect, and its mechanism remains to be elucidated. Therapeutic drug monitoring, electrolytes level monitoring, and the avoidance of drug-drug interactions may help prevent severe adverse drug reactions.

病例報告 113_C6

瀰漫性毒性甲狀腺腫患者使用 Propylthiouracil 引起黃疸的病例報告及治療策略分析

Propylthiouracil-Induced Jaundice in a Patient with Graves' Disease: A Case Report and Review of Management Strategies

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Introduction

Jaundice is a rare but serious complication of Graves' disease, particularly when associated with antithyroid drug (ATD) use. This case report discusses a unique presentation of Graves' disease complicated by jaundice due to propylthiouracil (PTU)-induced liver injury.

Case Report

A 25-year-old woman with a fifteen-year history of poorly controlled Graves' disease presented with jaundice. She had intermittently used PTU and methimazole, with a notable loss to follow-up for three years. Her free thyroxine (free T4) was above 5.8 ng/dL. Six months before this admission, she resumed PTU 100 mg three times daily after developing thyrotoxicosis symptoms. Five months after starting PTU, her laboratory results showed a free T4 of 4.28 ng/dL, triiodothyronine (T3) 4.46 ng/mL, thyroid-stimulating hormone (TSH) under 0.1 μIU/mL, and alanine aminotransferase (ALT) 39 U/L.

She experienced abdominal discomfort, diarrhea, tea-colored urine, and yellow skin and eyes for ten days before admission. Diagnosed with hepatitis and direct type hyperbilirubinemia, PTU was discontinued. Physical exam showed grade 3 thyroid goiter, icteric sclera, bulging eyes, and hand tremor.

Laboratory results indicated markedly elevated aspartate aminotransferase (AST) 201 U/L, ALT 317 U/L, bilirubin 32.9 mg/dL (direct 29.3 mg/dL), alkaline phosphatase (ALK-P) 173 U/L, albumin 3.8 g/dL, and INR 1.1. Free T4 remained above 5.8 ng/dL, TSH was 0.1 μIU/mL, T3 was above 780 ng/dL, and anti-TSH receptor antibodies were above 40 IU/L. Thyroid ultrasound showed bilateral heterogeneous and hyperechoic thyroid with increased vascularity. Abdominal ultrasound revealed no biliary tract dilatation or liver lesions. Burch-Wartofsky Point Scale (BWPS) score was 20-25 points, excluding thyroid storm. Liver biopsy later confirmed drug-induced liver injury (DILI). Due to PTU-induced jaundice suspicion, ATDs were avoided. Initial treatment included propranolol and cholestyramine. Given her severe ophthalmopathy and risk of radiation-induced thyroiditis, radioactive iodine therapy was not pursued. Ultimately, thyroidectomy was planned, but high bilirubin levels and hyperthyroidism increased surgical risks. Preoperative management with Lugol's solution and plasma exchange reduced free T4 to 2.22 ng/dL, T3 to 268.12 ng/dL, and bilirubin to 2.9 mg/dL. She subsequently underwent bilateral total thyroidectomy with parathyroid implantation, maintaining postoperative free T4, TSH, and bilirubin within normal ranges.

Discussion

This case illustrates the complex management required for Graves' disease complicated by PTU-induced liver injury. Thyrotoxicosis can independently cause hepatic dysfunction, with prevalence

rates of 15 to 76%, due to increased oxygen consumption, hypoxia, apoptosis, and oxidative stress. PTU, with a liverTox grade A, mainly causes hepatocellular injury, though its hepatotoxic mechanism is unclear.
Therapeutic plasma exchange (TPE) effectively reduces circulating T3 and T4 levels by removing hormone-bound plasma proteins and other substances like autoantibodies, catecholamines, and cytokines. The effect of TPE is transient, serving as a useful bridge to thyroidectomy for patients with refractory thyrotoxicosis or contraindications to ATDs.

Conclusion

This case underscores the importance of regular monitoring and careful management of patients with Graves' disease, especially those on ATDs like PTU. Prompt recognition and discontinuation of the offending agent are crucial to prevent further hepatic damage. TPE and timely surgical intervention effectively manage thyrotoxicosis and associated complications, providing a safe pathway to definitive treatment.

病例報告 113_C7

反覆多菌種菌血症和慢性骨盆痛：一例人為障礙症

A Case of Recurrent Polymicrobial Bacteremia and Chronic Pelvic Pain: A Factitious Disorder

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Introduction

Factitious Disorder is a psychiatric condition where a person deliberately fakes or induces illness in themselves or others, seeking medical attention without clear external rewards like financial gain. The behavior stems from a deep psychological need to be seen as ill, often leading to unnecessary medical interventions and complex interactions with healthcare providers.

Case Report

The patient is a 23-year-old female with recurrent chronic osteomyelitis after a pelvic bone fracture six years ago. She had a suicide attempt during high school. One year prior to this admission, she was admitted to the infectious disease ward with suspected osteomyelitis. During that admission, she displayed defensive and uncooperative behavior, and ultimately discharged herself against medical advice. She just graduated from a medical university.

The patient presented to the emergency department with a one-week history of fever and worsening pelvic pain. She had fever up to 39°C. Symptoms of vomiting, constipation followed the onset of fever. There was no peritoneal sign. The results of laboratory and image studies were listed (table 3). After admission, initial blood cultures were negative, but subsequent cultures following CVC (central venous catheter) use identified multiple waterborne pathogens including *Stenotrophomonas maltophilia*, *Elizabethkingia spp.*, *Alcaligenes faecalis*, *Trichosporon spp*,.and *Acinetobacter baumannii* (table 1). During the patient's hospitalization one year ago, bacteria cultured from the blood were identified and are listed in Table 2.

The patient initially refused CVC removal, leading to ongoing bacteremia and recurrent fever. The patient exhibited defensive and uncooperative attitudes. She demonstrated deceptive and disruptive behaviors including exaggerating her physical symptoms. She was often observed spending extended periods in the restroom, raising suspicion of factitious disorder. Weeks after, the patient finally consented to the removal of her CVC; however, bacteremia persisted. Concerns were raised regarding waterborne pathogen contamination through intravenous catheter use, but the patient declined alternative treatments, such as oral or intramuscular antibiotics. She eventually requested discharge against medical advice. We provided comprehensive discharge planning, emphasizing the importance of continued outpatient follow-up for ongoing antibiotic therapy and management of chronic medical conditions.

Discussion

Recurrent and multiple episodes of polymicrobial bacteremia are highly unlikely in an immunocompetent patient. Additionally, 80% of the bacteria cultured from the blood in this case were waterborne organisms, which are rarely encountered in clinical practice. What’s more, despite repeated communication, the patient persistently engaged in self-harmful behaviors,

often exhibiting anger and even aggression toward the healthcare team. Based on a literature review, the pathophysiology of factitious disorder was considered and applied to this patient (table 4). We faced many clinical challenges, including ethical, legal, and privacy issues, which are summarized in table 5. Despite these challenges, we were striving to respect and listen to her needs as much as possible, while adhering to hospital regulations and legal requirements in carrying out all medical orders.

Conclusion

The case highlights the difficulties in treating chronic conditions when psychiatric comorbidities are present and raises considerations for diagnosing factitious disorder in patients with recurrent infections and unusual behavior.

病例報告 113_C8

Deferasirox 誘發的范科尼氏症候群：案例報告

Deferasirox-induced Fanconi syndrome—a case report

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Introduction

Fanconi syndrome is a generalized proximal tubulopathy leading to electrolytes imbalance, metabolic acidosis and metabolic bone disease. Several inherited and acquired causes of Fanconi syndrome had been identified. Here we presented a rare case of deferasirox-induced Fanconi syndrome.

Case Report

A 33-year-old female with history of thalassemia, systemic lupus erythematosus (SLE) with antiphospholipid syndrome presented with erythema and swelling of the right forearm. She was diagnosed with cellulitis and started on empirical antibiotics with ampicillin/sulbactam.

During hospital stay, nephrologists were consulted for persistent glycosuria on urinalysis, despite normal blood glucose levels, no history of diabetes mellitus and sodium-glucose cotransporter-2 (SGLT-2) inhibitor use. Hypokalemia (2.8 mEq/L) was also present.

On suspicion of proximal tubulopathy, comprehensive laboratory evaluations were undertaken, which revealed hypophosphatemia (2.0 mg/dL), hypouricemia (1.2 mg/dL), and non-anion gap metabolic acidosis (pH: 7.33, bicarbonate: 19.4 mEq/L, sodium: 138 mEq/L, chloride: 110 mEq/L). Urine biochemistry confirmed potassium, phosphate, uric acid wasting and glycosuria. These findings were all consistent with Fanconi syndrome.

As for the etiology of Fanconi syndrome, screening for monoclonal gammopathy were negative. Her drug exposure history was reviewed. Deferasirox, an iron-chelating agents prescribed for hyperferritinemia in the past 6 months, was identified as a potential culprit drug, and thus was discontinued.

Following the discontinuation of deferasirox, the electrolyte imbalances and metabolic acidosis showed marked improvement within one week, allowing for the cessation of potassium, phosphate, and bicarbonate supplementation. Follow-up urinalysis also revealed complete resolution of glycosuria.

Discussion

Fanconi syndrome is a disorder of generalized proximal tubular dysfunction, leading to impaired reabsorption of potassium, phosphate, glucose, amino acids, uric acid and bicarbonate, resulting in various metabolic abnormalities, including hypokalemia, hypophosphatemia, hypouricemia, non-anion gap metabolic acidosis.

Fanconi syndrome can be inherited or acquired. Common acquired causes include monoclonal gammopathies and drugs like cisplatin and ifosfamide. In this case, deferasirox was identified as the cause. The patient's laboratory findings were typical of Fanconi syndrome. Importantly, cessation of deferasirox led to normalization of laboratory parameters within one week,

supporting the drug's causative role in the development of Fanconi syndrome.
The exact pathogenesis of deferasirox-induced Fanconi syndrome is unclear, but mitochondrial dysfunction in proximal tubular cells due to iron depletion may play an important role. This impairs ATP production needed for electrolyte transport, resulting in urinary wasting of these substances. The proximal tubule is particularly vulnerable to mitochondrial insults, since it lacks the enzymes necessary to generate ATP via anaerobic glycolysis. Additionally, the high urinary concentrations of deferasirox may damage the renal epithelium, particularly in patients with pre-existing renal impairment.
Given the rarity of this adverse effect, deferasirox remains a mainstay treatment for iron overload. This case underscores the importance of regular monitoring of renal function and electrolyte levels, and early recognition of Fanconi syndrome in individuals receiving deferasirox to prevent long-term complications.

Conclusion

This case highlights the need for vigilance in detecting Fanconi syndrome in patients receiving deferasirox therapy. Further research is needed to explore the pathophysiology of deferasirox-induced nephrotoxicity and identify potential protective strategies for vulnerable patient populations.

病例報告 113_C9

罕見真菌引起的腹膜透析腹膜炎感染: 病例報告

Neurospora Intermedia peritonitis in peritoneal dialysis: case report

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Introduction

Peritoneal dialysis is a popular technique for renal replacement therapy. The most common infection associated with peritoneal dialysis (PD) is bacterial PD peritonitis. Rarely, fungal cause of PD related peritonitis occurs with majority caused by *Candida* species. Here, we reported the first case of *Neurospora intermedia* infection in PD peritonitis patient.

Case Report

This was a 61-year-old man with history of end stage renal disease under PD for 5 years. He worked as an interior designer and helped in family farming business, which used to be a sugar refinery. Patient initially had progressive diffuse abdominal dull pain associated with cloudy dialysate fluid. He went to regional hospital and treated as PD peritonitis. PD fluid was culture negative. Cefazolin and ceftazidime were started intraperitoneally for two weeks. Condition worsened as he developed incoherent speech and confusion. He was transferred to National Cheng Kung University Hospital.

At our hospital, catheter site showed no erythema or pustular discharge. Peritoneal fluid showed 447 μL white blood cells (WBC) with 95% polymorphonuclear cells. Ceftazidime and Vancomycin intraperitoneal injection was given then shifted to intravenous form after initiation of hemodialysis with double lumen. PD solution culture later reported mold isolation. Amphotericin-B liposome was started as empiric therapy and PD tube was removed.

The identification report showed *Neurospora intermedia* three weeks after admission. Medication was shifted to voriconazole according to drug sensitive test. However, fever occurred and Abdominal CT showed lobulated fluid in pelvis which is compatible with PD peritonitis. Pigtail was inserted and drained yellowish pus like fluid. Report showed elevated WBC and negative bacterial culture. Antibiotic treatment course started for suspected pelvic abscess. Patient was discharged after improved condition. Receiving a total of 7 weeks of oral Isavuconazole for fungal infection.

Discussion

Neurospora is a genus of Ascomycete fungi. Mostly are aerobic. It lives in humid tropical regions. Ascospores that are dormant are activated by heat. It occurs in plywood factories, lumber yards, and contaminated bakeries[1,2]; on burned grass[3]; and stubble from burned sugar cane fields[4].

N. intermedia is the predominant species found on burned substrates, distinguished by its yellowish carotenoids and large conidia. It stands as the sole edible mold within the *Neurospora* genus, occurring in cooked maize cobs and onchom. Traditionally utilized in Java, Indonesia in the production of fermented foods[5,6,7]. Our case is the first report of *N. Intermedia* peritonitis in PD,

which was identified by multilocus phylogenetic analysis and internal transcribed spacer analysis. Upon literature review, there are three cases with endobronchial mass caused by *N. intermedia*[8]. Four cases of *Neurospora sitophila* infection. Two causing PD peritonitis[9,10]. One in eye infection following cataract surgery[11]. Another in pulmonary infection in HIV patient.12

Conclusion

Neurospora species could act as an opportunistic infection in immune compromised patient. We treated *Neurospora* infection using empiric antifungal medications with amphotericin-B induction followed by voriconazole and isavuconazole maintenance. These antifungal medications seems effective in treating *Neurospora* species infection and should be considered in patient with *Neurospora* related PD peritonitis.

病例報告 113_C10

脊髓旁腫塊所致之肺外結核感染引起的不明熱：案例報告

Fever of unknown origin caused by extrapulmonary tuberculosis in paraspinal masses: A case report

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Introduction

Extrapulmonary tuberculosis, though significant, is uncommon at paraspinal sites. This case report presents an instance of paraspinal soft tissue tuberculosis.

Case Report

A 42-year-old man with type 1 diabetes mellitus, hyperthyroidism, and steatocystoma multiplex presented with a six-month history of fever and right upper back pain. Chest computed tomography revealed two hypoenhancing masses in the paraspinal region. A CT-guided biopsy suggested tuberculosis, later confirmed by TB-PCR. The patient was treated with the HERZ regimen, leading to a gradual resolution of fever.

Discussion

Tuberculosis is an infection primarily caused by Mycobacterium tuberculosis. While the lungs are the most common site of infection, tuberculosis can affect other parts of the body, including the paraspinal region.

Conclusion

Tuberculosis can present with a wide range of clinical manifestations. Accurate diagnosis is essential for effective treatment.

病例報告 113_C11

位於食道黏膜下的異位性副甲狀腺腺瘤於一位三發性副甲狀腺亢進的病人：一例罕見的病例報告

A case report of tertiary hyperparathyroidism with an ectopic parathyroid adenoma located at esophageal sub-epithelial: an unusual location

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Introduction

Tertiary hyperparathyroidism is the autonomous secretion of parathyroid hormone in the setting of long-standing renal disease, leading to hypercalcemia. Although numerous cases of ectopic parathyroid adenoma associated with primary hyperparathyroidism have been reported, ectopic parathyroid adenoma in tertiary hyperparathyroidism is seldom described.

Case Report

We reported the case of a 28-year-old woman with a history of moderate mental retardation, bilateral hearing impairment and end-stage-renal-disease under hemodialysis for 5-6 years. She presented with low back pain since 2021. Initial study revealed osteoporosis and hypercalcemia, then she was referred to our hospital. Further survey demonstrated elevated intact-parathyroid hormone (iPTH), and parathyroid sonography revealed suspicious parathyroid adenoma or hyperplasia in right lower thyroid region. Tertiary hyperparathyroidism was diagnosed. The patient underwent total parathyroidectomy with right forearm auto-transplantation on November 9, 2021. Postoperatively, persistent hypercalcemia with elevated iPTH were noted. Parathyroid scan on July 6,2022 showed a Sestamibi-avid lesion in the right retro-tracheal area, raising the suspicion of parathyroid adenoma or hyperplasia. Head and neck CT performed on October 20, 2022, revealed a suspected soft tissue nodular lesion about 1.2cm at upper esophagus, which parathyroid adenoma could not be ruled out. Panendoscopy on December 27, 2022, showed a subepithelial lesion at 17 cm below incisor, measuring about 2.0 x 1.5 cm. Endoscopic ultrasound scan showed a heterogeneous hyperechoic lesion, with nodule-in-nodule pattern, arising from the 3rd layer. Whole body PET scan was done on 2023/01/13 and showed increased FDG uptake in the nodular lesion in upper esophagus about 1.2cm. Casanova test was arranged on February 2, 2023, and showed a negative result. Endoscopic submucosal dissection was done on February 17, 2023, and pathology confirmed the diagnosis of ectopic parathyroid adenoma.

Discussion

The prevalence of ectopic parathyroid is 15.9% according to a 2019 meta-analysis, with 11.6% in the neck and 4.3% in mediastinum. There were plenty of case reports presented about primary ectopic parathyroid adenoma. A primary retro-clavicular parathyroid adenoma was also reported in “journal internal medicine of Taiwan” in 2020. The retro/paraesophageal region is a common location of ectopic parathyroid adenoma, based on previous statistical findings. However, ectopic parathyroid adenoma presented as esophageal subepithelial lesion has not been reported before. We report a case of persistent hypercalcemia with elevated iPTH following total parathyroidectomy with right forearm auto-transplantation. Imaging and endoscopic study

identified the ectopic parathyroid adenoma presented as esophageal subepithelial lesion, which was successfully removed by endoscopic submucosal dissection.

Conclusion

Ectopic parathyroid tissue is common in humans and can be located from neck to mediastinum. Residue ectopic parathyroid adenoma should be considered if there is no clinical improvement following surgery for hyperparathyroidism. Adequate imaging study are necessary to identify the location of ectopic lesion.

病例報告 113_C12

金黃色葡萄球菌感染性心內膜炎完全治療後免疫複合體導致之腎小球腎炎的非典型延遲發作：病例報告

Atypical Delayed Onset of Immune-Complex Mediated Glomerulonephritis Following Complete Treatment of *Staphylococcus aureus* Infective Endocarditis: A Case Report

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Introduction

Infective endocarditis (IE) can lead to renal failure due to infective endocarditis-associated glomerulonephritis (IEAGN), with most cases showing improvement after antibiotic therapy. We present a case of delayed immune-complex mediated glomerulonephritis (GN) after complete treatment of Staphylococcus aureus-associated IE.

Case Report

A 37-year-old male with poorly controlled diabetes mellitus, hypertension, and a smoking history developed *Staphylococcus aureus*-associated IE with tricuspid valve vegetation and bilateral septic pulmonary emboli in November 2023. He received complete vancomycin treatment course for 39 days with stable renal function (creatinine 0.5-0.9 mg/dL) during hospitalization.

Two weeks post-discharge, he presented with edema and worsening renal function (creatinine 3.6 mg/dL). A renal biopsy revealed immune-complex GN, superimposed on diabetic nephropathy. No active infection was detected, His renal function gradually improved without the use of antibiotics or immunosuppressants.

Discussion

IE occurs in 30-60% of *Staphylococcus aureus* bacteremia cases, with high mortality. Immune mechanisms are implicated in 80% of cases, where circulating bacterial antigen-antibody complexes deposit in the glomerulus, triggering complement activation and renal injury. Acute renal failure is the most common clinical presentation of IEAGN, often accompanied by hematuria and nephritic or nephrotic syndrome. Historically, glomerular patterns in IEAGN have been diffuse with immune complex deposition. Our patient, however, developed immune-complex mediated GN one to two months after completing treatment for *Staphylococcus aureus*-associated IE, which is atypical as IEAGN generally occurs during the acute phase of infection.

The patient exhibited normal complement levels and renal biopsy showed crescentic GN with mesangial and subendothelial deposits, suggesting a subacute disease course. Blood cultures were negative, indicating no active infection, and the patient was treated without antibiotics or immunosuppressants. The gradual improvement in renal function without immunosuppression highlights the potential for spontaneous resolution in subacute presentations of IEAGN, especially in cases where immune activation has subsided post-infection.

Conclusion

This case underscores the atypical, delayed onset of immune-complex mediated GN after

complete treatment of *Staphylococcus aureus* associated IE. Remarkably, the patient's renal function improved without corticosteroid or immunosuppressive therapy, emphasizing the potential for spontaneous resolution in such cases.

病例報告 113_C13

廣泛期小細胞肺癌患者接受化免疫治療和緩和性放療後出現化膿性心包炎：罕見且危急的併發症病例報告

A case of pyopericardium after chemoimmunotherapy and palliative radiotherapy in extensive-stage small cell lung cancer: a rare and critical complication

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Introduction

Small-cell lung cancer (SCLC) accounts about 15% of lung cancers, with 70% of cases being metastatic at diagnosis. Radiologic findings often show centrally localized tumor in the major airways with bulky mediastinal lymphadenopathy and distant metastasis. Chemoimmunotherapy is the first-line treatment for extensive-stage SCLC, offering good efficacy with generally manageable side effects like neutropenia, anemia, and nausea. However, rare but serious complications require vigilant monitoring.

Case Report

A 54-year-old man with past history of heavy smoking presented with persistent dry cough and swelling in the upper limbs and neck for 1 month. Chest CT revealed a mass (maximal 7.8cm) in the right upper lung and extensive mediastinal lymphadenopathy (maximal 8.4cm), complicated with superior vena cava (SVC) compression. Endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) of the mediastinal lymphadenopathy confirmed a diagnosis of SCLC, which was classified as extensive-stage due to the substantial tumor burden, and malignant pleural effusion. The patient received palliative radiotherapy for SVC syndrome and chemoimmunotherapy with cisplatin, etoposide, and atezolizumab.

One week after treatment, the patient developed cardiogenic and septic shock, presenting with hypotension, tachycardia, chest pain, and fever. Chest CT revealed necrosis and shrinkage of the right upper lung and mediastinal lymphadenopathy, along with necrotic fluid and air in the pericardium and mediastinum, leading to pyopericardium and mediastinal abscess. An urgent pericardiotomy with creation of a pleuropericardial window was performed, draining frankly pus-like fluid. The effusion cultures identified *Fusobacterium necrophorum*, with no evidence of mycobacteria or fungi. Following treatment with ampicillin/sulbactam and adequate drainage, follow-up imaging confirmed resolution of the infection. The patient was discharged after three months and continued chemoimmunotherapy.

Discussion

Pyopericardium is a rare condition, accounting for less than 1% of pericarditis cases. It is typically associated with pneumonia, empyema, thoracic surgery, or hematogenous spread from sepsis. Less common causes include contiguous spread from the retropharyngeal space, cardiac valves, or areas below the diaphragm. In our case, the tumor responded well to chemoimmunotherapy and radiotherapy, but subsequent necrosis led to local inflammation and invasion of the pericardium. The effusion culture suggested possible microinvasion to the hypopharyngeal space.

Pyopericardium requires aggressive management, with 85% of cases showing good long-term outcomes when treated promptly with antimicrobial therapy and drainage.

Conclusion

Pyopericardium should be considered a potential complication during SCLC treatment when mediastinal lymphadenopathy is bulky, requiring timely and aggressive management.

病例報告 113_C14

雙重心房中膈穿刺挽救了場面：一例左心耳封堵器滑脫的病例報告

Double Transseptal Puncture Saves the Day: Securing and Retrieving a Slipping Left Atrial Appendage Occluder

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Introduction

90% of strokes related to nonvalvular atrial fibrillation result from thrombosis occurring in the left atrial appendage (LAA). Left atrial appendage closure offers a treatment option for patients with atrial fibrillation but cannot tolerate anticoagulants. During the placement of a left atrial appendage occluder (LAAO), there is about a 0.07% chance of dislodgement.

Case Report

We reported a 76-year-old man with history of atrial fibrillation (CHA2DS2-VASc score=7), embolic stroke and acute lower limb ischemia. He had recurrent upper and lower gastrointestinal bleeding likely related to anticoagulation therapy. He was admitted for left atrial appendage occlusion. Pre-procedural transesophageal echocardiography (TEE) identified a windsock-type LAA. A 32mm "Watchman" LAAO was implanted. However, post-deployment TEE revealed progressive migration of the device. Leaving the slipping LAAO in place poses the risk of its evolution into dislodgement, which may cause complications and difficulty of retrieval. Consequently, our goal is to retrieve the device before it could dislodge. A St. Jude mapping catheter was inserted through the original LAAO sheath to stabilize the device, preventing further migration. The "snaring technique" published in other case reports was not feasible since the base of the device was still partially lodged in the left atrial appendage. Additionally, the angle discrepancy between the LAAO sheath and the screw of the device make it impossible to re-engage the original delivery catheter. Thus, a second trans-septal puncture was performed with an SL1 sheath and BRK needle. The SL1 sheath was then exchanged for a steerable cryoballoon sheath with the assistance of a super-stiff guidewire. The flexible sheath was maneuvered to align with the screw at the top of LAAO, allowing successful re-engagement of the device. Once securely attached with the sheath, the LAAO was carefully retracted and removed. A new 35 mm "Watchman" occluder was successfully implanted. The patient was discharged uneventfully in the next day.

Discussion

Despite the low incidence of LAAO dislodgement, its occurrence is often accompanied by complications (58.2%) and an unacceptably high mortality (10.2%). Dislodgement of the LAAO primarily occurs during the procedure (45.3%) or within 24 hours post-intervention. The dislodged LAAO could compromise hemodynamics, and the retrieval of an embolized LAAO may cause injury to papillary muscles or valves. There is no standard management for such situation as presenting in our case. Our strategy was to stabilize the device as soon as possible before it could dislodge. A double transseptal puncture and a steerable cryoballoon sheath was utilized in our case.

Conclusion

LAAO dislodgement may cause severe complications. The strategy to immediately stabilize and retrieve a slipping LAAO is rational. Double transseptal puncture and a steerable cryoballoon sheath may be considered in this situation.

病例報告 113_C15

成功合併使用熊去氧膽酸和非諾貝特治療一名台灣原發性膽汁性膽管炎患者：病例報告

Successful Combination Therapy of UDCA and Fenofibrate in a Taiwanese Primary Biliary Cholangitis Patient: A Case Report

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Introduction

Primary biliary cholangitis (PBC), formerly known as primary biliary cirrhosis, is a chronic cholestatic liver disease. Ursodeoxycholic acid (UDCA) is the standard first-line treatment; however, patients with an incomplete response to UDCA require additional therapies. Fibrates have been used in such cases, though data on their efficacy in Taiwan is limited.

Case Report

We report the case of a 65-year-old Taiwanese man with a 5-year history of glaucoma and previously undiagnosed chronic cholangitis, who had been managed with UDCA for approximately 10 years at another hospital. He visited our hospital for a second opinion. Physical examination showed hyperpigmentation of the skin and splenomegaly. Laboratory results showed elevated levels of bilirubin (1.4 mg/dL), alkaline phosphatase (ALP, 148 U/L), and antimitochondrial antibody (AMA, 70.5 units). Abdominal imaging showed normal biliary tract system except for liver fibrosis and splenomegaly. Transient elastography (FibroScan) indicated liver stiffness of 11.4 kPa. The diagnosis of PBC was made accordingly to the clinical presentations, the laboratory results, and the image studies. Despite ongoing UDCA treatment, elevated ALP and liver enzyme levels persisted. Therefore, fenofibrate 200 mg daily was added to the UDCA regimen. Three months later, bilirubin and ALP levels decreased to 1.1 mg/dL and 85 U/L, respectively. After one year, liver stiffness decreased to 7.4 kPa.

Discussion

Approximately 40% of PBC patients do not respond to UDCA treatment, with male sex at presentation being a predictor of non-response. Fibrates have been reported to potentially improve ALP and total bilirubin levels, which are the two most important prognostic indicators in PBC. In our case, the patient showed a good response to the combination therapy, as indicated by improved liver FibroScan results and delayed progression of liver inflammation and fibrosis. Moreover, AMA may initially be within the normal range in early-stage PBC and may later become positive over time, as observed in our case. If liver biopsy is declined by the patient, annual monitoring of AMA levels may be necessary, particularly for those with UDCA-resistant cholestasis.

Conclusion

This is the first case report of a UDCA-refractory PBC Taiwanese patient underwent additional fenofibrate treatment successfully. We highlighted the possibility of applying fibrates in treatment of Taiwanese PBC patients.

病例報告 113_C16

糖尿病患者無乳鏈球菌腦膜炎併發腦梗塞

Streptococcus agalactiae Meningitis Complicated Cerebral Infarction in a Diabetic Patient

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Introduction

The incidence of *Streptococcus agalactiae*, or group B Streptococcal (GBS) disease in non-pregnant adults is increasing, particularly among the elderly and those with underlying conditions such as diabetes, neurological impairment, and cirrhosis. Common clinical presentations of GBS include skin and soft-tissue infections, pneumonia, and urosepsis. Although less frequent, meningitis and endocarditis are associated with significant morbidity and mortality.

Case Report

We report a 48-year-old previously healthy male who presented to the emergency department with 2-day duration of coma and high fever. Physical examination showed drowsy consciousness, crackles over the bilateral lungs on auscultation, and decubitus sores over the sacral region. Laboratory results were notable for hyperglycemia, ketoacidosis, metabolic acidosis, and an elevated C-reactive protein level. The chest film showed patchy opacities over the bilateral lower lung fields. The diagnosis of pneumonia and decubitus sores complicated diabetic ketoacidosis was made accordingly to the clinical presentations and laboratory results. A lumbar puncture was performed for the central nervous system infection survey. The cerebrospinal fluid (CSF) analysis revealed pleocytosis, low glucose, and a high protein level. The multiplex polymerase chain reaction (PCR) of CSF showed positive for *Streptococcus agalactiae*. The blood culture, sputum culture, and wound culture all yielded *S. agalactiae*. Magnetic resonance imaging (MRI) of brain revealed leptomeningeal enhance, ventriculitis, and cerebral infarctions over bilateral frontal lobes, parietal lobes, centrum semiovale, and corona radiata. The patient was successfully treated with ampicillin but recovered with right hemiplegia.

Discussion

S. agalactiae, which is a bacterium typically present in the normal flora of the human gastrointestinal and genitourinary tracts, can lead to severe invasive infections in both neonates and adults. These infections are uncommon in adults, representing less than 1% of all cases. Diabetes mellitus is the most frequently observed comorbidity in adults with invasive GBS disease. Generally, these infections appear as bacteremia, and there is a higher incidence of central nervous system infections, such as meningitis. The bloodstream serves as the primary route for the dissemination of the infection. Septic emboli can cause extensive or multiple infarctions, which facilitates further spread of the infection. Another suggested mechanism is immune-mediated vasculitis due to GBS, potentially leading to vascular changes and cerebral infarctions. However, additional diagnostic and histopathological studies are required to confirm the presence of thrombo-occlusive vasculopathy.

Conclusion

This case highlights a rare instance of disseminated *S. agalactiae* infection, encompassing skin and soft-tissue infection, pneumonia, meningitis, ventriculitis, and bacteremia, complicated with acute cerebral infarctions. Cerebral infarction in bacterial meningitis indicates severe disease and poor prognosis. Early identification, thorough investigation, and prompt antibiotic treatment are crucial for managing disseminated S. agalactiae disease effectively.

病例報告 113_C17

一位鏈球菌腦膜炎併發腦室炎之個案報告研究

Metagenomic Next-Generation Sequencing for Diagnosis of Group A Streptococcus meningitis：A case report

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Introduction

Streptococcus intermedius is a commensal microflora commonly found in various mucosal sites in the respiratory, gastrointestinal, and genitourinary tracts. It causes invasive suppurative infections including liver and brain abscesses along with thoracic empyema. However, it rarely causes meningitis without abscess formation.

Case Report

A 56-year-old immunocompetent man who presented with fever and headache. Bacterial meningitis was confirmed using cerebrospinal fluid analysis. Magnetic resonance imaging of the brain revealed leptomeningitis and ventriculitis. However, conventional methods failed to identify the causative pathogens. Metagenomic next-generation sequencing of cerebrospinal fluid revealed the presence of S. intermedius. The patient underwent antibiotic treatment accompanied by surgical intervention, resulting in a favorable prognosis.

Discussion

CSF culture is the gold standard for pathogen identification in CNS infections. However, the positive rate of CSF cultures was estimated to be only 30–40% in CNS infections. Although timely antibiotic treatment for CNS infections management is important, the usage of antibiotics before CSF sampling would decrease pathogen detection rates.

As a member of *Streptococcus anginosus* group (SAG), S. intermedius has high mortality rates, longer hospital stays, and usually causes suppurative infections. S. intermedius-induced CNS infections usually present as brain abscess rather than meningitis. Although, several cases of S. intermedius brain abscesses have been documented, only four cases of S. intermedius meningitis or ventriculitis have been previously reported. Three of the four involved individuals were over the age of 60, while the fourth case was a 6-year-old boy. CSF cultures yielded negative results in half (2/4) of the reported cases. The S. intermedius were identified by PCR amplification in the culture-negative cases.

While conventional microbiologic testing is insufficient to detect all pathogens, mNGS exhibits satisfactory diagnostic performance in CNS infections and has a higher detection rate than conventional culture techniques. DNA or RNA sequencing of pathogens via mNGS accelerates turnaround time in testing.

Conclusion

Herein, we report a rare case of meningitis and ventriculitis caused by S. intermedius using metagenomic next-generation sequencing. The patient recovered well after antibiotic treatment

and surgery. We present this rare case and summarize previous studies to remind clinicians that timely identification of the pathogen and optimal treatment are crucial for management of S. intermedius-induced infections.

病例報告 113_C18

懷孕女性嚴重肺炎使用高流量鼻導管及高頻胸壁振盪：病例報告

Management of Severe Pneumonia in a Pregnant Woman Using High-Flow Nasal Cannula and High-Frequency Chest Wall Oscillation: A Case Report

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Introduction

Severe pneumonia during pregnancy presents significant challenges in clinical management due to the risks to both maternal and fetal health. During the COVID-19 pandemic, the use of high-flow nasal cannula (HFNC) increased for patients experiencing respiratory distress, demonstrating its effectiveness in maintaining oxygenation. Here, we present a case of a 30-year-old pregnant woman with severe pneumonia, underwent HFNC and high-frequency chest wall oscillation (HFCWO) in achieving respiratory stability.

Case Report

A 30-year-old woman at 25 2/7 weeks of gestation, with a history of cesarean section, presented with symptoms of upper respiratory infection, fever, and low back pain. Initial evaluations confirmed right lower lobe pneumonia. Her condition deteriorated, necessitating multiple antibiotics and HFNC for oxygen support. Additionally, chest percussion therapy was used to manage severe pneumonia and facilitate sputum clearance. Betamethasone was administered to promote fetal lung maturation due to the increased risk of preterm labor. With HFNC and HFCWO therapy, the patient's respiratory condition gradually improved, allowing for a successful discharge without any major complications.

Discussion

This case highlights the complexity of managing severe pneumonia in a pregnant patient, where balancing maternal and fetal safety is paramount. The use of HFNC provided effective oxygen support, while HFCWO contributed significantly to the resolution of sputum consolidation. Early identification of pathogens using a FilmArray panel enabled targeted antibiotic therapy, which played a crucial role in stabilizing the patient. The administration of betamethasone helped mitigate potential complications associated with preterm labor, underscoring its value in such high-risk scenarios.

Conclusion

This case underscores the importance of a multidisciplinary approach and the use of advanced respiratory therapies, such as HFNC and HFCWO, in managing severe pneumonia during pregnancy to ensure positive outcomes for both mother and fetus.

病例報告 113_C19

上腸系膜動脈症候群

Superior mesenteric artery syndrome

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Introduction

Superior mesenteric artery (SMA) syndrome is an uncommon cause of upper gastrointestinal obstruction. It was caused by compression of third portion of duodenum due to narrowing of angle between SMA and aorta. In this report we will present a 42 year old man with severe vomiting due to SMA syndrome.

Case Report

A 42 years-old thin male is relatively well before. However, he felt abdominal distention and easily stuffed since two weeks ago. In addition, nausea and vomiting occurred after meal since three days. Mild dyspnea due to abdominal fullness and poor appetite were also complained. Then he was sent our emergency room. At our ER, his vital signs are unstable as hypotension, tachycardia (BP-96/45mmHg, T.P.R-36.5 °C , 112/min, 18/min). Physical examination revealed abdominal distention and diffuse tenderness without muscle guarding. Laboratory data showed hyponatremia (Na-122mmol/L) and hypokalemia (K-2.4mmol/L). KUB revealed distention with food debris in stomach and duodenum. Under the suspicion of upper GI obstruction, he was admitted to our GI ward. After admission, abdominal CT was done and revealed dilatation of stomach and proximal duodenum with an abrupt transition point at the third part of the duodenum. The aorto-mesenteric angle measures 21 degree and the aorto-mesenteric distance was about 7.43 mm. Then SMA syndrome was diagnosed. Laparoscopic duodenojejunostomy was arranged at the fifth day of hopitalization. After the procedure, eternal feeding was tried since 10th day and NG tube was removed on 14th day. He was discharged on 17th day with normal oral intake.

Discussion

SMA syndrome caused by compression of the third portion of the duodenum between the aorta and the superior mesenteric artery can induce chronic, intermittent or acute upper GI obstruction. This syndrome can occur on thin or rapid weight loss patient due to loss of the intervening mesenteric fat pad. Narrowing of aorto-mesenteric angle (≦25°) and short aortomesenteric distance (≦8mm) are the diagnostic criteria on CT scan. Conservative treatment with NG decompression, electrolytes and fluid balance and nutrition support can be tried but patients with persisted symptoms should be treated by surgery. Several surgical options such as Strong's procedure, gastrojejunostomy, and duodenojejunostomy can be selected and duodenojejunostomy is generally accepted as having superior results.

Conclusion

SMA syndrome is an uncommon cause of duodenal obstruction. Narrowing of aorto-mesenteric angle and short aortomesenteric distance can lead further compression of duodenum by SMA and

aorta. Surgical managemetn should be done if conservative treatment failed.

病例報告 113_C20

病歷報告：偽裝結核性肋膜炎與心包膜炎的縱膈腔淋巴瘤

Primary mediastinal large B cell lymphoma mimicking tuberculous pleurisy and pericariditis: a case report

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Introduction

Primary mediastinal large B cell lymphoma (PMBL) is an aggressive lymphoma that is arised from thymus. It has diverse clinical presentation including mediastinal tumor, SVC syndrome, pleural effusion, pericardial effusion, etc.

Case Report

A 36-year-old female patient without past history presented with persisted cough, chest tightness, and dyspnea for two weeks. Chest radiograph showed opacity lesion of left lung. Chest computer tomography (CT) revealed massive left pleural effusion, minimal pericardial effusion and a mass lesion in left upper lobe with invasion of the mediastinum and pulmonary vessels. Thoracentesis was done and the plural effusion analysis revealed exudate character with lymphocyte predominance. The Adenosine deaminase (ADA) level was 66 U/L (normal <40). Under the diagnosis of tuberculosis (TB) with tuberculous pleurisy and pericarditis, she started to receive anti-tuberculous treatment. However, dyspnea and cough relapsed two month later. The repeated chest CT revealed a progressed mass over anterior mediastinum with massive pericardial effusion. Pericardectomy was done by a cardiovascular surgeon. The pathologic report of anterior mediastinum tumor and pericardial peel demonstrated high grade B cell lymphoma and there was no evidence supported the diagnosis of tuberculosis. Then she received chemotherapy with a regimen of da-EPOCH-R for PMBL and soon achieved a clinico-radiographic partial response.

Discussion

In clinical practice, high ADA level in pleural effusion or pericardial effusion is plausible to establish a diagnosis of TB pleurisy or pericarditis, however, the false-positive results can occur in uncommon conditions such as lymphoma or autoimmune diseases.

Conclusion

This case indicated some limitation of ADA level as a diagnostic tool for tuberculosis. The precise diagnosis of PMBL is based on pathologic evaluation of tumor tissue in anterior mediastinum because some differential diagnosis such as extra-pulmonary TB or lung cancer may have the same clinical presentation

病例報告 113_C21

壞死梭桿菌菌血症引起的中樞神經系統併發症：兩個病例報告

CNS complications secondary to Fusobacterium necrophorum bacteremia: two case reports

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Introduction

Fusobacterium necrophorum bacteremia is often linked to Lemierre's syndrome (LS), first described by André Lemierre in 1936. LS arises from oropharyngeal infections that extend to the lateral pharyngeal spaces, causing septic thrombophlebitis of the internal jugular vein (IJV). Typically affecting young, healthy individuals aged 10 to 35, LS can impact multiple organ systems. Once the infection reaches the IJV, it can spread hematogenously, leading to various complications,and with local or distant arterial complications occurring in about 10% of patients. We report two cases of F. necrophorum bacteremia with central nervous system (CNS) complications.

Case Report

Case 1

A 62-year-old male with a history of hypoxic ischemic encephalopathy, ventilator dependence, type 2 diabetes, alcoholic liver cirrhosis, and buccal cancer was admitted with an endotracheal tube. He developed a fever (38.7°C), prompting suspicion of ventilator-associated pneumonia and treatment with ceftriaxone. Fever subsided after three days, but blood cultures revealed Fusobacterium necrophorum after five days. A neck computed tomography (CT) indicated right internal carotid artery occlusion, right cerebral anterior and middle cerebral artery territory infarction, and severe hydrocephalus. The family declined neurosurgery, opting for palliative care. Antibiotics were continued, stabilizing the patient.

Case 2

An 85-year-old male with COPD, type 2 diabetes, chronic kidney disease, and a history of cerebral infarction was admitted with an endotracheal tube. He presented with a fever (38.3°C), and ceftriaxone was given for suspected urinary tract infection. Seven days later, blood cultures showed Fusobacterium necrophorum. Two weeks later, fever returned, and new cultures revealed Prevotella disiens. Neck CT demonstrated a left cervial cord C1-C3 epidural abscess and narrowing of the proximal IJV. Antibiotic treatment continued, leading to stabilization and resolution of fever.

Discussion

Lemierre's syndrome, once rare, has re-emerged, likely due to reduced broad-spectrum antibiotic use for head and neck infections in younger populations. CNS complications are uncommon, with only 22 cases reported in one hospital series from 1980 to 2010. High clinical suspicion is crucial for diagnosis, as physical exams may not reveal the disease. Early signs often include persistently high fevers, and it should be suspected in patients with recent pharyngitis, fever, swollen neck, or pulmonary symptoms. Advanced LS has a mortality rate of 5-18%, with potential complications including metastatic disease and septic shock.

Conclusion

Our patients were significantly older with endotracheal tube in place and ventilator dependency and unable to express their symptoms, which delayed diagnosis until CT scans were performed after blood cultures confirmed F. necrophorum. Both patients were intubated, which may have facilitated oral bacteria invasion due to airway damage.

病例報告 113_C22

肝硬化病人中感染性心內膜炎的罕見且嚴重現象：病例報告

Rare and severe phenomenon of infective endocarditis in a patient with liver cirrhosis: a case report

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Introduction

Infective endocarditis (IE) can be a cause of fever of unknown origin. This case report discusses rare manifestations of IE, including pericardial effusion, aortic valve ring abscess, and ventricular septal defect (VSD).

Case Report

We present a 51-year-old male with liver cirrhosis (child-pugh class A), admitted with a 5-day fever. Initial physical examination and laboratory tests were unremarkable, but a chest CT revealed aortic valve calcification. Blood cultures identified Enterococcus faecalis, and ertapenem was administered. Despite 7 days of treatment, the fever persisted, and an inflammatory gallium scan showed negative findings. The patient left against medical advice for personal reasons. Two months later, he was re-admitted with bilateral leg edema, mild dyspnea, and painful macules on his palms. A chest X-ray indicated a water bottle-shaped heart. Laboratory tests showed leukocytosis and elevated CRP, with blood cultures again identifying Enterococcus faecalis.
Transthoracic echocardiography revealed a moderate amount of pericardial effusion, vegetation on the aortic valve, and signs of severe aortic regurgitation, moderate mitral regurgitation, and mild tricuspid regurgitation. Transesophageal echocardiography confirmed active IE with vegetation, aortic valve ring abscess, and VSD. Due to uncontrolled infection and signs of heart failure, the patient underwent aortic valve replacement and VSD repair after admission for 4 days. During surgery, a bicuspid aortic valve and vegetation were found. He was transferred to the ICU post-operation, but after 1 day, he ssuccumbed to septic shock.

Discussion

Aortic valve calcification and stenosis typically affect older adults, but in younger patients, they often result from congenital heart defects like a bicuspid aortic valve, significantly increasing IE risk. Pericardial involvement in IE is rare, primarily due to inflammation spreading from a perivalvular abscess. Aortic valve ring abscess is a serious complication, with a mortality rate of 19-25%, and its incidence is elevated in underlying heart disease like a bicuspid aortic valve.
Bacterial infections are common in cirrhosis cases, as seen in our patient with persistent bacteremia. The coexistence of liver cirrhosis and IE is rare; however, the clinical features and bacterial ecology are similar to those of the general population, with characteristics such as male predominance, preferential involvement of the aortic and mitral valves, a history of heart disease, and a predominance of Gram-positive bacteria.

Conclusion

The diagnosis and management of IE pose significant challenges, especially in patients with decompensated liver function. A bicuspid aortic valve notably increases the risk for IE. Additionally, incomplete antibiotic treatment and persistent bacteremia during the initial hospitalization compromised treatment efficacy and elevated mortality risk.

病例報告 113_C23

胰臟假性囊腫造成阻塞性黃疸：病歷報告

Obstructive Jaundice Caused by Chronic Pancreatic with Pseudocysts: a case report

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Introduction

Pancreatic pseudocysts causing external compression and jaundice are rare. Typically, this occurs in patients with alcoholic chronic pancreatitis, where jaundice arises either from hepatocellular damage or from compression and narrowing of the terminal common bile duct (CBD) due to fibrosis. In this report, we present an unusual case of chronic pancreatitis complicated by a pancreatic pseudocyst leading to obstructive jaundice.

Case Report

This 35-year-old male with chronic pancreatitis, who presented with obstructive jaundice due to compression of the common bile duct by a pancreatic pseudocyst. The patient was admitted with complaints of epigastric pain and jaundice. Initial treatment involved endoscopic papillotomy and the insertion of a plastic stent via endoscopic retrograde biliary drainage (ERBD) after percutaneous drainage of the pseudocyst. After these interventions, the patient was discharged with follow-up appointments scheduled in our GI outpatient departments.

A few weeks later, the patient returned to the emergency room with similar symptoms but without jaundice. Laboratory tests showed normal total bilirubin levels, and imaging confirmed the proper placement of the biliary stent near the pseudocyst. He continued medical treatment during hospitalization, and subsequent CT scans showed a decrease in the size of the infected pseudocyst. After completing antibiotic treatment, he was discharged in stable condition with outpatient follow-up arranged.

Discussion

Pancreatic pseudocysts often present with symptoms such as abdominal pain and nausea. When the cyst becomes infected, patients may develop fever. Typically, obstructive jaundice associated with pancreatic pseudocysts is caused by fibrotic strictures of the intrapancreatic portion of the CBD, rather than direct compression by the pseudocyst. In order to provide clarity to the patho-physiology of pseudocysts causing jaundice, Sidel et al. proposed four criteria to confirm that jaundice is caused by a pancreatic pseudocyst: (1) presence of biliary obstruction, (2) demonstration of compression of the common bile duct by the pseudocyst, (3) relief of biliary obstruction following drainage of the pseudocyst, and (4) complete resolution of jaundice during the postoperative period. The management of pancreatic pseudocysts may involve various interventional approaches, including percutaneous drainage and endoscopic retrograde cholangio-pancreatography (ERCP), among others.

Conclusion

In our case, the patient met all four criteria, as jaundice resolved following stent placement, even

though the pseudocyst recurred later without causing jaundice. This supports the conclusion that the jaundice in this patient was due solely to the pseudocyst's compressive effect on the CBD. This experience may assist us in the clinical diagnosis and treatment of similar cases.

病例報告 113_C24

與系統性紅斑性狼瘡相關的毛細血管滲漏及噬血症候群：病例報告

Systemic lupus erythematosus-associated capillary leak and hemophagocytic syndromes: A case report

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Introduction

Capillary leak and hemophagocytic syndromes are life-threatening hyperinflammatory conditions, and may rarely be associated with initial diagnosis of systemic lupus erythematosus (SLE).

Case Report

A 36-year-old previously healthy woman presented with foamy urine for 1 month and intermittent fever for 1 week. Physical examination revealed pitting leg edema and tender swelling over bilateral interphalangeal joints. Laboratory tests showed normal WBC (7900/μL), anemia (Hb: 10.8 g/dL), thrombocytopenia (platelet: 149,000/μL), normal serum creatinine, nephrotic-range proteinuria, positive ANA, elevated anti-dsDNA (>444 IU/mL), and low complement levels (C3: 41 mg/dL; C4: 7.8 mg/dL). Under the suspicion of lupus nephritis, she was admitted for renal biopsy, which was smoothly performed in the morning (day 1). That evening, she developed fever, chills, dyspnea and desaturation. Follow-up labs showed elevated creatinine (1.79 mg/dL), procalcitonin (32.76 ng/mL), and D-dimer (>100,000 ng/mL). Initial chest x-ray findings were unremarkable. Chest CTA revealed mild lung infiltrates, minimal perirenal hematoma and no pulmonary embolism. Empirical antibiotics were administered and high flow nasal cannula was applied. On day 2, dyspnea progressed and oxygenation worsened. Hypotension and oliguria developed. Repeat CXR showed dense infiltrates in bilateral mid-lower lungs. She received invasive mechanical ventilation and was transferred to the ICU. Follow-up hemogram showed hemoconcentration (WBC: 43000/μL; Hb: 15.8 g/dL; platelet: 220,000/μL), suggesting capillary leak syndrome. Fluid, albumin, hydrocortisone and norepinephrine were administered. On day 3, marked drop of the hemogram (WBC: 12500/μL; Hb: 3.8 g/dL; platelet: 61000/μL) and hepatitis (AST: 2646 U/L; ALT: 951 U/L) were noted. There was no hemosputum or gastrointestinal bleeding. Abdominal echo excluded significant perirenal hematoma. The haptoglobin level was normal. Further laboratory tests revealed a low reticulocyte count (0.4%), hyperferritinemia (71,406 ng/mL), and hypofibrinogenemia (177 mg/dL). Bone marrow biopsy confirmed the suspicion of hemophagocytic syndrome. She received transfusions, Dexamethasone (10 mg/m^2/day), and continuous veno-venous hemofiltration. All cultures had no growth. Renal biopsy confirmed class IV lupus nephritis. Her condition gradually improved. Dialysis was discontinued on day 14. She was weaned from the ventilator on day 17, left the ICU on day 21, and discharged home on day 32.

Discussion

Capillary leak syndrome is characterized by sudden leakage of fluid and proteins from blood vessels into surrounding tissues, leading to edema, hypotension, hemoconcentration, and organ dysfunction. Hemophagocytic syndrome is diagnosed when 5 out of 8 criteria are met, including

fever, hepatosplenomegaly, cytopenias, hyperferritinemia, hypertriglyceridemia or hypofibrinogenemia, hemophagocytosis, low NK cell activity, and high soluble CD25 level. Secondary causes of both syndromes are similar and include infections, autoimmune diseases, cancers, and certain medications. For those associated with autoimmune diseases, the primary treatment is corticosteroids, with intravenous immunoglobulin used if the condition is not responsive to steroids.

Conclusion

This case report is unique in that the rapid clinical decline shortly after renal biopsy initially raised suspicion of sepsis or procedure-related complications, which was ultimately attributed to capillary leak, followed by hemophagocytic syndrome associated with SLE.

病例報告 113_C25

皮肌炎引起之食道無效收縮致反覆胃食道逆流 ： 病例報告

Refractory gastroesophageal reflux due to ineffective esophageal motility cause by scleroderma : a case report

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Introduction

Ineffective esophageal motility (IEM) is defined in the Chicago Classification v4.0 as a disorder of esophageal peristalsis. Common clinical symptoms may include dysphagia, gastroesophageal reflux disease or regurgitation, but symptoms vary widely and sometimes asymptomatic. In scleroderma, an autoimmune disease that usually causes inflammation and fibrosis, esophagus is the most commonly affected part of the gastrointestinal system. In this case, we would present a case of ineffective esophageal motility related to scleroderma and the image of high resolution esophageal manometry.

Case Report

This 28-year-old female patient has a medical history of dermatomyositis (diagnosed at 2011), systemic lupus erythematosus with secondary Sjogren syndrome, digital vasculitis, class V lupus nephritis and connective tissue disease-related interstitial lung disease. She had regular follow up at our patient department and received prednisolone, mycophenolic acid, hydroxychloroquine for disease control.

This time, the patient complained about similar symptoms happened about two years ago, including cough, food regurgitation and gastric acid regurgitation in the morning. Consider symptoms persisted with refractory gastroesophageal reflux disease and scleroderma related cannot be ruled out. Panendoscopy was arranged and high resolution manometry was performed to evaluate the function of the esophagus. In high-resolution esophageal motility study, there was no elevated integrated relaxation pressure pressure, no premature contraction nor hypercontractility, but ineffective swallows (100%) with water retained in the esophagus when supine. Given her history of her autoimmune disease, she was diagnosed as ineffective esophageal motility and favor scleroderma related. After above examination, PPI was prescribed for symptomatic control and her clinical condition improved gradually.

Discussion

The diagnosis of ineffective esophageal motility requires more than 70% ineffective swallows or at least 50% failed peristalsis during high-resolution manometry (HRM) testing. Diagnostic thresholds of HRM includes weak contraction (DCI $\geq$ 100 mmHg · s · cm and < 450 mmHg · s · cm), failed peristalsis (DCI < 100 mmHg · s · cm), or a fragmented swallow.

Ineffective esophageal motility may be caused by smooth muscle disease, connective tissue disorder, opioids or other inflammations. Such as scleroderma, an autoimmune disease usually causes inflammation and fibrosis in the body, including skin, blood vessels or major organs. In scleroderma, esophagus is the most commonly affected part of the gastrointestinal system.

Esophageal involvement may lead to esophageal smooth muscle atrophy, esophageal fibrosis, dysmotility and finally significant reduction in the quality of life. Acid suppressive therapy is essential to these patients, and esophageal dilation should be considered because of the higher risk of developing stricture.

Conclusion

In ineffective esophageal motility caused by scleroderma may lead to gastroesophageal reflux and cause food and gastric acid regurgitation. In HRM, ineffective swallows and failed peristalsis may also be observed. Acid suppressive therapy usually prescribed for symptomatic treatment.

病例報告 113_C26

以經皮支架置入術治療縱膈淋巴瘤引起的肺動脈壓迫: 個案報告

Percutaneous Stenting for Pulmonary Artery Compression Caused by Mediastinal Lymphoma: A Case Report

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Introduction

Mediastinal lymphoma may compress pulmonary arteries (PA) and lead to severe pulmonary hypertension and right ventricular failure, which can be managed by percutaneous stenting.

Case Report

A 28-year-old woman presented to the emergency department with a 2-week history of cough and progressive dyspnea. She was diagnosed with stage III, intermediate risk primary mediastinal B-cell lymphoma (PMBCL) approximately 7 months ago, and treated with 6 cycles of dose-adjusted EPOCH-R (etoposide, prednisone, vincristine, cyclophosphamide, doxorubicin, and rituximab). Two months after completing treatment (8 days before the current presentation), a positron emission tomography scan was performed and showed a high probability of viable tumor in the mediastinum. Physical examination showed mild swelling of her face and neck, and no stridor, wheezes or crackles. A computed tomography scan of the chest was done and revealed right-side pleural effusion and a mediastinal tumor, compressing the lower trachea, main bronchi, superior vena cava (SVC), and bilateral PAs. This compression caused severe stenosis of the right PA. A diagnosis of PMBCL with progressive disease causing PA compression was made. The patient was admitted to the intensive care unit due to the development of hypotension and acute respiratory failure. Percutaneous stenting of the right PA was performed, with dramatic improvement of the PA hypertension and hemodynamic parameters. A subsequent bronchoscopic biopsy was conducted, and the pathology confirmed the diagnosis. She then received Nivolumab plus Brentuximab vedotin for the PMBCL and was discharged home at 36 days.

Discussion

PMBCL is frequently associated with SVC syndrome and airway compromise, but compression of the PA is relatively uncommon. PA stenting should be considered as a potential treatment option for relief of symptoms related to PA compression.

病例報告 113_C27

與結節性多動脈炎相關的噬血症候群: 個案報告

Polyarteritis Nodosa-associated Hemophagocytic Syndrome: A Case Report

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Introduction

Hemophagocytic syndrome is a life-threatening hyperinflammatory condition, and may rarely be associated with polyarteritis nodosa (PAN), a systemic necrotizing vasculitis predominantly targeting medium-sized arteries.

Case Report

A 41-year-old man presented to the emergency department due to a one-week history of fever (up to 40.1 °C). He had painful necrotic ulcers with peripheral erythema over both lower extremities for 3 months and was diagnosed with PAN one month ago. Prednisolone 10 mg per day was prescribed. On admission, physical examination revealed reduced erythema around the persistent necrotic ulcers, with some appearing scarred. Laboratory findings were remarkable for mild anemia and thrombocytopenia (Hb 8.6 g/dL, platelet 103000/μL), and mildly elevated CRP (24.4 mg/dL) and liver enzymes (GOT 109 U/L, GPT 71 U/L). Cultures of the wounds grew *Pseudomonas aeruginosa* and *Klebsiella oxytoca*. He was treated with Piperacillin/tazobactam and methylprednisolone (40 mg/day). However, relapsing fever persisted. Abdominal CT scan was done and showed splenomegaly. One week after admission, hyperferritinemia (ferritin 27,204 ng/ml), hypertriglyceridemia (triglyceride 713 mg/dL) and hypofibrinogenemia (fibrinogen 157.5 mg/dL) were noted. Bone marrow biopsy was done and revealed hypocellualr marrow with hemophagocytosis. A diagnosis of PAN-associated hemophagocytic syndrome was made. Dexamethasone (10 mg/m^2/day) was administered. Fever declined gradually. However, dyspnea, desaturation and hypotension developed. A follow-up chest x-ray showed newly developed bilateral lung infiltrates. He was intubated and transferred to the ICU on day 12. A 10-fold increase in ferritin level was noted (274,150 ng/mL), and a repeat liver function test showed direct hyperbilirubinemia and worsening hepatitis (total/direct bilirubin 9.99/7.90 mg/dL and GOT/GPT 680/226 U/L). Steroid pulse therapy (methylprednisolone 1 g/day) was administered. Despite this, his condition rapidly deteriorated and he died from profound shock and hypoxemia on day 13, when a ferritin level of 491,445 ng/ml was noted, nearly doubling in one day. All subsequent cultures showed no growth. Tests for cytomegalovirus and Epstein-Barr virus (EBV) were both negative.

Discussion

Clinical features of hemophagocytic syndrome are similar to those of sepsis and active autoimmune diseases. Serial monitoring of laboratory parameters, especially serum ferritin level, may be helpful. Hemophagocytic syndrome is diagnosed when 5 out of 8 criteria are met, including fever, hepatosplenomegaly, cytopenias, hyperferritinemia, hypertriglyceridemia or hypofibrinogenemia, hemophagocytosis, low NK cell activity, and high soluble CD25 level.

Secondary causes of the syndrome include infections, autoimmune diseases, cancers, and certain medications. For those associated with autoimmune diseases, also known as macrophage activation syndrome, the primary treatment is corticosteroids. Intravenous immunoglobulin (IVIG) and IL-1 receptor antagonist may be considered for those refractory to steroid therapy. We found only one case report of a 73-year-old Japanese woman with PAN-associated hemophagocytic syndrome. She died from internal bleeding and bowel perforation despite treatment with steroid pulse therapy, IVIG, and VP-16. Unlike our case, EBV was detected in that patient.

Conclusion

Risks for hemophagocytic syndrome in patients with PAN should be recognized.

病例報告 113_C28

後天免疫缺乏症候群患者之鳥型分枝桿菌感染合併脾臟梗塞及多處淋巴結腫大-病例報告

Disseminated Mycobacterium avium Complex (MAC) Infection in an AIDS Patient with Splenic Infarction and Lymphadenopathy

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Introduction

Mycobacterium avium complex (MAC) is a group of non-tuberculous mycobacteria that commonly causes opportunistic infections in people with advanced HIV, especially when CD4 counts drop below 50 cells/μL. MAC infection in HIV patients can disseminate to multiple organs, including the lymph nodes, spleen, liver, lungs, and bone marrow.

Case Report

The patient, a 34-year-old male with past history of hypertension, was first seen in the outpatient department(OPD) on January 10, 2024, after noticing a decline in his immunity, frequent rashes, and recurrent colds. He had tested positive for HIV in May 2023 following a self-initiated screening. His major risk factor included a history of MSM (men who have sex with men). During his first clinic visit, check HIV related lab data and Dolutegravir/Lamivudine (Dovato) was initiated. His HIV Ag/Ab Combo Test showed a high viral load(3431 copies/mL). Baktar 2 tab QD was prescribed for prophylactic antibiotic treatment. Following OPD, due to the patient's ongoing pain, an abdominal CT was arranged. The patient's abdominal pain had lessened. Physical examination continued to show a rash over his arms and chest, and new oral findings of thrush were noted. Lab results revealed a CD4 count of 38, and a positive EBV-CA IgG test. His HIV drugs were also shifted to Bictegravir/Tenofovir/Emtricit(Biktarvy). An abdominal CT showed splenomegaly and focal infarction, enlarged mesenteric lymph nodes, and mediastinal and bilateral pulmonary hilar poorly-enhanced nodules(Figure 1); which all above finding might be related to HIV infection. The patient was admitted to the hospital on January 25, 2024, due to persistent left upper abdominal pain and was found to have splenic infarction. During his hospital stay, further workup revealed mediastinal lymphadenopathy and bilateral pulmonary hilar nodules, including chest CT. A Chest CT showed multiple enlarged lymph nodes. TB culture was collected and showed Mycobacterium avium complex. He underwent a video-assisted thoracoscopic surgery (VATS) on January 31, 2024, for biopsy of a mediastinal mass. Pathological examination indicated non-caseating granulomatous inflammation, raising suspicion of a Mycobacterial infection; besides, TB culture from biopsy tissue also showed Mycobacterium avium complex (MAC) Initial negative sputum PCR results for tuberculosis, and subsequent sputum TB culture results confirmed MAC infection. The patient was discharged on February 3, 2024, with continued outpatient follow-up planned and a regimen of antiretroviral therapy. At OPD, his antiretroviral therapy was shifted to Dovato due to MAC therapy, and for MAC therapy, Ethambutol(400mg) 4 tabs QD + Rifabutin(150mg) 2 capsules QD + Clarithromycin(250mg) 2 tabs BID were prescribed since February 21, 2024. However, further follow up lab data showed slow response to CD4 level and viral load, so his antiretroviral therapy was shifted to Abacavir/Lamivudine/Dolutegrav(Triumeq). He is under regular OPD follow up.

Discussion

This case presents a 34-year-old male with HIV infection and disseminated Mycobacterium avium complex (MAC) infection. The diagnosis of **mediastinal lymphadenopathy, pulmonary nodules, and splenic infarction** indicates that the MAC infection had disseminated to several key sites, a characteristic finding in patients with low CD4 counts below 50 cells/μL (1). Besides, the splenic infarction found in this patient may reflect severe MAC involvement in the spleen, a less common but serious complication in advanced HIV patients (2).

Treatment of disseminated MAC in HIV patients involves a combination of antibiotics, typically including clarithromycin or azithromycin along with ethambutol, as recommended by established guidelines. The patient was initiated on combination antibiotics along with continuation of antiretroviral therapy (ART). Careful monitoring for immune reconstitution inflammatory syndrome (IRIS) is necessary, as IRIS can occur during immune recovery and cause exacerbation of MAC symptoms (4).

Conclusion

This case emphasizes the importance of recognizing and treating disseminated MAC infection in patients with advanced HIV, particularly those with severe immunosuppression (CD4 < 50 cells/μL). The involvement of multiple organs, such as the lungs, lymph nodes, and spleen, underscores the aggressive nature of MAC in severely immunocompromised patients.

病例報告 113_C29

伴有未識別基因突變的非典型溶血尿毒症症候群-較輕病程與治療策略

Atypical Hemolytic Uremic Syndrome with an Unidentified Gene Mutation: A Case Report of Milder Disease Course and Management Approach

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Introduction

Atypical hemolytic uremic syndrome (aHUS) is a rare disease characterized by microangiopathic hemolytic anemia (MAHA), thrombocytopenia, and damage to vital organs. The pathogenesis lies in dysregulation of the complement pathway with complement gene mutation or autoantibody. Here we presented a case of aHUS with unidentified mutation.

Case Report

A 38-year-old male without any underlying diseases was admitted to our hospital with general malaise for several days. Additionally, severe hypertension was noted. His laboratory tests revealed normocytic anemia, renal dysfunction, and thrombocytopenia. Additionally, microscopic hematuria and proteinuria were detected. A renal biopsy was performed due to the unknown cause of acute kidney injury (AKI), which revealed active thrombotic microangiopathy. We then checked markers for MAHA, showing low hemoglobin (Hb), low haptoglobin, high LDH, and Schistocyte 1+. We also examined autoimmune profiles, tumor markers, viral infection panel, and disseminated intravascular coagulation (DIC) profiles, all of which were negative. Later, the patient developed several times of tarry stool and seizure episodes. Stool O157:H7 test and panendoscopy were done, revealing negative results and a duodenal ulcer, respectively. Meanwhile, the ADAMTS13 level was checked, showing a detectable level of 62.4%. We then immediately initiated plasma exchange for 4 consecutive days; however, he remained dialysis dependent. The aHUS gene panel was checked, which revealed no complement-related genetic variants. The patient is currently receiving continuous ambulatory peritoneal dialysis. We are currently applying for eculizumab, seeking possibilities of achieving dialysis independence and a better prognosis for him.

Discussion

There are 40–60% of patients in aHUS with genetic or acquired dysregulation of the complement alternative pathway. Several mutations in genes play a role in the dysregulation. However, there are 30-50% patients with aHUS having non-identified mutations and 32-50% of them might experience end stage renal disease (ESRD) or death. Previous studies have shown that the renal outcome was not significantly different in adults regardless of genetic background. Eculizumab, a complement C5 inhibitor, was a helpful treatment in both identified and unidentified mutations of aHUS individuals. Some case studies reported on dialysis free and recovering of renal function after taking Eculizumab.

Conclusion

Given the generally poor prognosis of aHUS, and the proven benefits of Eculizumab on renal function irrespective of gene mutation status, immediate Eculizumab treatment is essential. In our case, we highlight the characteristics of milder course in unidentified mutation and choice of peritoneal dialysis for protecting patient's residual kidney function. This clinical decision could potentially increase the chances of achieving dialysis independence after Eculizumab therapy.

病例報告 113_C30

茶色胸水

Tea-colored pleural effusion: A Intriguing Case Report

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Introduction

Pleural effusion is an uncommon complication of acute pancreatitis. Typically, it presents as a transient, left-sided accumulation of straw-colored fluid. However, in rare instances, the effusion may be right-sided and hemorrhagic in nature. This atypical presentation can pose significant diagnostic challenges, particularly when thoracic manifestations predominate over abdominal symptoms.

Case Report

A 47-year-old male was admitted to the intensive care unit with complaints of epigastric pain for one day, which had aggravated over the past few hours. He reported that the abdominal pain was constant and dull. His symptoms were associated with diffuse abdominal cramping pain and were irrelevant to postural change. The patient has a history of gallstones and has not undergone cholecystectomy. The patient reported no history of alcohol exposure. There is no family history of pancreatic diseases. He denied taking any other medications or herbal remedies recently.

The initial vital signs indicate a normal sinus rhythm of 116 beats per minute, a blood pressure of 196/111 mmHg, and a saturation of 97% on ambient air. On physical examination, the patient's breath sounds were clear bilaterally, and no pitting edema was observed. Abdominal examination revealed localized tenderness on palpation in the epigastric region, without rebound tenderness and with a negative Murphy's sign. On review of systems, he denied any constitutional symptoms (fever or chills, weakness or fatigue, weight loss or gain, night sweats) as well as any cardiovascular, respiratory, neurological, musculoskeletal, hematological, or endocrinological issues.

Laboratory studies revealed an elevated white blood cell count (17,800/μL), markedly elevated serum lipase level (1426 U/L). Liver function tests indicated elevated GPT, GOT, with direct hyperbilirubinemia, while the coagulation profile was within the normal limits. Biochemical blood tests revealed no hypercalcemia or hypertriglyceridemia. The patient has no recent history of abdominal surgeries or trauma. Further laboratory tests were negative for serum IgG4 markers, ruling out IgG4-related disease.

Abdominal computed tomography revealed necrotizing pancreatitis with acute necrotic collections at peripancreatic, bilateral anterior pararenal region and omentum. There was no evidence of choledocholithiasis on CT scan. Bilateral pleural effusions developed three days later, and pigtail insertions were performed. Initially, the drained pleural fluid was tea-brown in color but later changed to yellow. His symptoms and condition improved under treatments that included aggressive intravenous hydration, Gabexate mesilate, and pain management. With the improvement of acute pancreatitis, his pleural effusions ceased to appear following drainage. After a 20-day hospital stay, the patient was discharged home.

Discussion

Pancreatitis-associated pleural effusion is a complication of pancreatic inflammation, where pancreatic fluid leaks into the pleural cavity, typically affecting the left side. The incidence is about 3-7% in patients with acute pancreatitis, but recent CT scan reports suggest that the rate has risen to nearly 50%. Treatment of pleural effusion generally begins with conservative management, but symptomatic pleural effusion often requires thoracentesis, chest tube placement, intubation, intensive care, parenteral nutrition, and administration of octreotide.

Conclusion

This case report emphasizes the importance of recognizing a rare yet critical pancreatitis complication, characterized by bilateral, large-volume, tea-colored pleural effusions.

病例報告 113_C31

部分型肺靜脈回流異常術後 13 年，發生肺靜脈狹窄導致單側肺水腫：一例罕見的病例報告

Unilateral pulmonary edema caused by stenosis of pulmonary vein, 13 years after surgery for partial anomalous pulmonary venous return : A rare case report

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Introduction

Unilateral pulmonary edema (UPE) is a rare and often misdiagnosed condition, particularly in the early stages. We present the case of a 40-year-old female with a history of partial anomalous pulmonary venous return (PAPVR) surgery 13 years ago, developed progressive right-sided pulmonary edema and respiratory failure following. The condition was caused by stenosis of the right upper pulmonary vein, leading to severe pulmonary arterial hypertension (PAH) and UPE. Surgical correction of the vein stenosis resulted in rapid resolution of symptoms, emphasizing the importance of considering venous abnormalities in UPE cases.

Case Report

e Report

A 40-year-old female with a history of PAPVR repair as rerouting of anomalous right superior pulmonary vein to left atrium 13 years ago presented to the emergency room on August 22, 2024, after sustaining fractures of the left 6th to 12th ribs. Her respiratory condition progressively worsened and pulmonary edema, necessitating mechanical ventilation and intensive care unit (ICU) admission on August 23, 2024. Despite receiving antibiotics and diuretics and her inflammatory markers (CRP 3 mg/L, Procalcitonin 0.33 ng/mL) improved, her right-sided lung congestion and respiratory failure persisted.

Preoperative transesophageal echocardiography (TEE) revealed significant stenosis of the right superior pulmonary vein (5 mm), absent visualization of the right lower pulmonary vein, increased flow velocity (179 cm/s), adequate left ventricular (LV) systolic function (LVEF 77.6%). No atrial septal defect (ASD) was detected. A CT pulmonary angiogram demonstrated a stenotic ostium, dilated pulmonary trunk, and D-shaped left ventricle, indicating pulmonary hypertension.

On September 3, 2024, she underwent successful surgical repair of the right upper pulmonary vein and atrial septum, along with pericardiotomy revision. Postoperative TEE showed improvement in the right upper pulmonary vein (8.5 mm at the ostium). The patient's endotracheal tube was removed the following day, with marked regression of unilateral pulmonary edema.

Discussion

UPE is a rare condition often mistaken for pneumonia or severe mitral valve insufficiency due to atypical radiographic findings. In this case, a patient with left-sided rib fractures developed right lung congestion, initially. Normal inflammatory markers, NT-proBNP levels and further imaging, including CTA and TEE, revealed stenosis of the right upper pulmonary vein and right lower pulmonary vein not visualized, ruling out heart failure and mitral valve insufficiency as causes.

PAPVR occurs when some pulmonary veins drain into the right atrium or its venous tributaries

instead of the left atrium. In this case, stenosis of pulmonary vein after rerouting of anomalous right superior pulmonary vein to left atrium 13 years ago for PAPVR caused impaired venous return from the right lung, leading to severe PAH and elevated pulmonary vascular resistance, ultimately resulting in right-sided pulmonary edema.
Following the re-do pericardiotomy and repair of right upper pulmonary vein, the pulmonary edema resolved dramatically within one day, confirming that the obstruction was the primary cause of the edema.

Conclusion

UPE is a reversible condition with a favorable prognosis if identified and treated promptly. This case highlights pulmonary vein stenosis, particularly in post-PAPVR patients, as a potential cause of UPE. Early recognition and intervention are crucial for optimal outcomes.

病例報告 113_C32

Octreotide 使用在難治性淋巴管肌瘤病相關乳糜胸中的病例報告

A case report on the use of octreotide in refractory chylous effusion associated with lymphangioleiomyomatosis

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Introduction

LAM (Lymphangioleiomyomatosis) is a rare disease characterized by abnormal proliferation of smooth muscle cells, affecting the lungs, kidneys, and lymphatic system. It could lead to pneumothorax, refractory pleural effusion, and lymphatic manifestations, such as chylothorax.

Case Report

A 48-year-old female patient was admitted to the hospital due to effort dyspnea. The disease had persisted for one month, presented with chest tightness, leg edema and without fever, cough, or orthopnea. The patient had been visited our Outpatient Department, where massive right pleural effusion was noted from chest X-ray imaging. Right-sided chylothorax was revealed after pig-tail drainage. No malignant cell was noted from cytology study.

Due to right-sided chylothorax, the patient underwent a CT scan of the chest on Day 2, which indicated the possibility of Lymphangioleiomyomatosis. VATS thoracic duct ligation and pleurodesis were performed on Day 16. Histopathological and immunohistochemical analyses revealed typical features of LAM. Fat-restricted diet and continued Sirolimus were given. However, refractory chylous effusion persisted in the following days. Consequently, octreotide was added for the next two weeks (Day 19-32). Finally, the drainage from the right chest was removed, and the patient was discharged in stable condition (Day 33). No effusion was noted during the outpatient department follow-up (1.5 month).

Discussion

In patients with LAM, chylous effusion is thought to arise from obstruction and destruction of the lymphatic ducts caused by proliferating LAM cells. Octreotide, a somatostatin analog peptide, reduces fat absorption from the gastrointestinal tract and decreases lymph flow. Additionally, octreotide may constrict vascular and lymphatic ducts, which could help reduce chylous effusion.

Conclusion

LAM with chylothorax refractory to conventional treatments may benefit from the combination therapy of sirolimus and octreotide, which appears effective in reducing the volume of chylous effusion.

病例報告 113_C33

胰島素瘤的診斷：一例非糖尿病患者低血糖的病例報告

Insulinoma Unveiled: A Case of Hypoglycemia in a Non-Diabetic Patient

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Introduction

Hypoglycemia is a critical condition frequently encountered in diabetic patients; however, its occurrence in non-diabetic individuals is relatively rare, necessitating an exploration of potential underlying causes.

Case Report

A 76-year-old man with a medical history of hypertension and dyslipidemia presented with acute altered consciousness and shouting early in the morning. Upon arrival, emergency medical technicians (EMTs) assessed his blood glucose levels, revealing hypoglycemia. He was promptly transported to the emergency department (ED), where intravenous glucose was administered, resulting in the rapid restoration of consciousness. The patient denied any previous history of diabetes or recent use of unknown medications, and he was subsequently admitted to the endocrinology ward for further evaluation.

A 72-hour fasting test was conducted, indicating endogenous insulin overproduction. An abdominal MRI revealed a 1.1 cm cystic lesion in the pancreatic head. A selective arterial calcium stimulation test (SACST) demonstrated a significant increase in insulin levels from the splenic artery, further corroborating the diagnosis of insulinoma. Despite treatment, hypoglycemia persisted during the hospital stay, leading to the initiation of continuous glucose monitoring (CGM). An endoscopic ultrasound (EUS) identified a hypoechoic nodule at the body-tail junction of the pancreas, measuring approximately 10.7 x 7.2 mm. A general surgeon was consulted for further management.

The patient subsequently underwent laparoscopic distal pancreatectomy, splenectomy, and cholecystectomy. The surgery proceeded without complications, and pathological examination confirmed a grade 1 neuroendocrine tumor (pT1), with no nodal involvement (0/2, pN0). The patient had an uneventful postoperative recovery, with no further episodes of hypoglycemia. Follow-up abdominal CT scans indicated no signs of tumor recurrence.

Discussion

This case illustrates a patient who presented with neurogenic symptoms of hypoglycemia, meeting the criteria for Whipple's triad. Given the patient’s health status and absence of diabetes, drug-induced hypoglycemia and critical illness were considered unlikely. A supervised fasting test was performed to evaluate for prandial hypoglycemia, revealing insulin levels of 6 μIU/ml and C-peptide levels of 0.86 ng/ml, indicating endogenous hyperinsulinism as the most probable cause. Additionally, elevated levels of neuron-specific enolase (NSE) and chromogranin A were noted. Further endocrine evaluations were undertaken to exclude the possibility of multiple endocrine neoplasia (MEN).

Imaging studies, including abdominal CT and MRI, revealed a lesion in the pancreatic head; however, SACST suggested the tumor was in the body and tail of the pancreas, a finding further substantiated by EUS. Ultimately, surgical intervention confirmed the diagnosis of insulinoma.

Conclusion

Hypoglycemia can present with neuroglycopenic symptoms and may occasionally be misinterpreted as a psychiatric disorder in healthy individuals without diabetes. Upon confirmation of hypoglycemia, a supervised fasting test is essential for differential diagnosis. The selective arterial calcium stimulation test (SACST) can further aid in localizing the tumor. For patients deemed suitable candidates, surgical resection is the preferred treatment, while medical management may be considered for those not eligible for surgery. Accurate diagnosis and management of insulinoma can significantly alleviate the patient's hypoglycemic symptoms.

病例報告 113_C34

分化不良多發性骨髓瘤個案報告

Anaplastic Multiple Myeloma – A Case report

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Introduction

Anaplastic multiple myeloma (AMM) is a rare, aggressive variant of myeloma, with challenging diagnosis. This case discusses a 71-year-old man initially misdiagnosed with peripheral T-cell lymphoma, not otherwise specified (PTCL, NOS), but later confirmed as AMM.

Case Report

The patient had a history of left inguinal spermatic cord lipoma and presented with a left inguinal mass for two months. An abdominal CT scan showed multiple enlarged lymph nodes in the aortocaval and para-aortic regions and a lobulated inguinal solid mass (5.1 x 4.5 x 8 cm). Excision pathology initially diagnosed PTCL, NOS. Immunohistochemical (IHC) staining showed diffuse positivity for CD45, while being negative for cytokeratin and SOX-10, confirming hematologic cells. Tumor cells were positive for CD3, CD4, CD8 (partial), CD56, and MUM-1, but negative for CD2, CD5, CD7, CD20, CD30, CD57, ALK, TIA-1, and granzyme B, with a Ki-67 index over 95%. Bone marrow biopsy revealed diffuse infiltration of atypical plasma cells with CD138 and light chain lambda restriction, confirming plasma cell neoplasm. IHC results included CD38+/CD43+/CD45-/CD56+/cCD79a-/CD117+/CD138+/cKappa-/cLambda+, and serum tests showed elevated IgA with decreased IgG and IgM, and a kappa/lambda ratio of 0.08. MRI detected small active lesions in sternum, bilateral scapula, bilateral ribs, sacrum, bilateral iliac bones, right pubic body, and right femoral shaft. Anemia and deteriorated renal function were also noted. Pathology review revealed monoclonality for IGH and polyclonality for TRG and TRB gene rearrangements, revising the diagnosis to extramedullary involvement by AMM.

The patient received one cycle of CHOP before the diagnosis revision, followed by four cycles of the VCTD regimen for 4 cycles and an autologous hematopoietic stem cell transplant (HSCT). He achieved complete remission and remains in remission under thalidomide maintenance treatment.

Discussion

New diagnostic molecular biological tools may aid precise diagnosis. AMM has dismal prognosis with an estimated median survival of 3 to 5 months. No standard treatment exists, with resistance to chemotherapy and novel therapies common. However, this patient had a positive outcome under aggressive treatment. Emerging drugs, such as CD38-targeting monoclonal antibodies (daratumumab, isatuximab), SLAMF7-targeting (elotuzumab), or checkpoint inhibitors (anti-programmed cell death 1/programmed cell death ligand 1), may further improve outcomes in AMM.

Conclusion

The diagnosis for AMM can be challenging. Accurately diagnosing AMM and promptly initiating anti-myeloma therapy are crucial for improving outcomes.

病例報告 113_C35

一例成人淋病結膜炎的診斷與治療經驗

A Case of Adult Ocular Infection Caused by *Neisseria gonorrhoeae*

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Introduction

Neisseria gonorrhoeae is a sexually transmitted pathogen primarily affecting the genitourinary system but can also infect other mucosal surfaces, including the eyes. The incidence of gonococcal infections has risen significantly in Taiwan, increasing from 4,000 cases in 2016 to 8,257 cases in 2023. While gonococcal conjunctivitis is more common in neonates, adult gonococcal conjunctivitis (AGC) is rare but has increased with the rising incidence of genital gonorrhea. Early diagnosis is crucial for preventing serious complications.

Case Report

A 26-year-old healthy woman presented with left eye pain and discharge after visiting a hot spring. Initial treatment with levofloxacin for conjunctivitis was ineffective. On follow-up, her left eye showed purulent discharge, and laboratory testing confirmed *N. gonorrhoeae*, resistant to fluoroquinolones. She was treated with intravenous ceftriaxone, doxycycline, and topical antibiotics. After five days, her symptoms improved, and she was discharged with continued oral and topical antibiotic therapy. At follow-up, she reported full resolution of symptoms, and further sexual history suggested potential genital gonorrhea in her spouse.

Discussion

AGC is rare but poses a significant risk to vision due to *N. gonorrhoeae*'s ability to penetrate the cornea. Diagnosis relies on culture or PCR, and prompt treatment is essential. In this case, the initial empirical treatment failed, highlighting the importance of culture and sensitivity testing. Standard treatment includes ceftriaxone, with additional doxycycline or azithromycin for suspected *Chlamydia* co-infection. The absence of early sexual history inquiry delayed the correct diagnosis, underscoring the need for a thorough evaluation in hyperacute conjunctivitis cases.

Conclusion

The increasing incidence of *N. gonorrhoeae* infections in Taiwan calls for heightened clinical awareness of AGC, particularly in younger patients. Timely diagnosis and appropriate antibiotic therapy are essential to prevent vision-threatening complications.

病例報告 113_C36

原發性副甲狀腺功能亢進症合併嚴重高鈣血症：案例報告

Severe Hypercalcemia in Primary Hyperparathyroidism: A Case Report

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Introduction

Primary hyperparathyroidism (PHPT) is a prevalent endocrine disorder characterized by excessive secretion of parathyroid hormone (PTH), resulting in hypercalcemia. While PHPT often presents asymptomatically, it can lead to various clinical manifestations, including nephrolithiasis, osteoporosis, and renal dysfunction. Severe hypercalcemia, although less common, may present with symptoms such as fatigue, weakness, and diminished appetite. This report discusses a rare case of PHPT associated with severe hypercalcemia and its effective management.

Case Report

A 57-year-old male with a history of hypertension and renal stone disease (previously treated with ureterorenoscopy in 2018) presented to the emergency department with generalized malaise and poor appetite persisting for several days. Laboratory investigations revealed significant renal impairment (serum creatinine: 3.82 mg/dL) and severe hypercalcemia (serum calcium: 18.3 mg/dL). Immediate management included intravenous hydration and calcitonin administration.

Further evaluation of the hypercalcemia showed markedly elevated intact PTH (iPTH) levels (743.1 pg/mL) and a 25-hydroxyvitamin D level of 24.0 ng/mL. Thyroid ultrasound identified a cystic mass measuring 2.3 x 2.1 x 3.2 cm in the right parathyroid region, accompanied by multiple spongiform nodules in both thyroid lobes. A parathyroid scan suggested hyperplasia of the bilateral lower parathyroid glands. Bone densitometry confirmed the presence of osteoporosis, with a T-score of -2.9.

The patient underwent a right upper parathyroidectomy, which revealed a cystic lesion measuring 2.0 x 2.0 x 3.5 cm. Pathological examination confirmed the diagnosis of a parathyroid adenoma. Postoperatively, iPTH levels normalized (9.4 pg/mL), and serum calcium levels returned to the normal range. The patient was discharged in stable condition with a scheduled follow-up.

Discussion

PHPT primarily results in hypercalcemia accompanied by elevated or unsuppressed PTH levels. This condition frequently leads to renal and skeletal complications, as evidenced by the patient's renal dysfunction and osteoporosis. Hypercalcemia in PHPT arises from increased bone resorption and enhanced renal calcium reabsorption mediated by PTH.

The diagnostic approach for PHPT typically includes measuring PTH levels, assessing vitamin D status, and employing imaging techniques such as ultrasound and parathyroid scans to localize the affected gland(s). Surgical resection of the adenoma remains the definitive treatment, often leading to normalization of calcium and PTH levels.

Interestingly, preoperative imaging in this case suggested possible bilateral lower parathyroid hyperplasia; however, intraoperative findings prompted the surgeon to excise the right upper

parathyroid adenoma. This highlights the importance of intraoperative assessment, which may differ from preoperative imaging interpretations.

Conclusion

This case exemplifies the classical presentation of PHPT with severe hypercalcemia and its successful resolution following surgical intervention. Parathyroidectomy remains the gold standard for managing symptomatic PHPT and for patients with significant complications, such as renal dysfunction and osteoporosis. Timely diagnosis and intervention are essential to mitigate long-term complications associated with hypercalcemia.

病例報告 113_C37

以反覆性血尿表現的胡桃鉗症候群：一例少見的病例報告

A middle-aged female with Nutcracker syndrome: an uncommon cause of hematuria

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Introduction

Nutcracker syndrome (NS) is caused by the compression of the left renal vein between the aorta and superior mesenteric artery, leading to impaired venous drainage. The primary symptoms are intermittent gross or microscopic hematuria, with or without associated flank pain. Despite its clinical relevance, NS is often difficult to diagnose due to its non-specific and variable presentation, resulting in frequent underdiagnosis.

Case Report

We present a 39-year-old slim female with no significant medical history, who experienced two episodes of painless gross hematuria and persistent microscopic hematuria, first noted at age 36 during a routine health examination. She also reported occasional mild lower abdominal and left flank pain. Laboratory tests revealed no significant hematologic, biochemical, or immunologic abnormalities, and urinalysis showed no dysmorphic red blood cells.

Renal ultrasonography showed no evidence of stones or tumors, and intravenous pyelography (IVP) did not identify any abnormalities in the urinary tract. Cystoscopy also showed no bladder issues. A computed tomography (CT) scan confirmed a reduced aortic-SMA angle (26°), left renal vein stenosis, and an enlarged left gonadal vein, consistent with a diagnosis of NS. Since there was no immediate risk of renal damage, the patient was managed conservatively with weight-gain nutritional support and regular follow-up to monitor hematuria and renal function.

Discussion

NS is often underdiagnosed due to its non-specific symptoms. The compression of the left renal vein increases venous pressure, resulting in hematuria, flank pain, proteinuria, pelvic congestion, and sometimes left varicocele. Hematuria occurs due to the rupture of small venous septa in the renal system.

Management of NS varies depending on the severity of symptoms and the risk of renal damage. Conservative treatment, such as nutritional support and close monitoring, is recommended for mild cases without immediate renal threats. More severe cases, especially those involving anemia, persistent pain, or renal dysfunction, may require interventional approaches like endovascular stenting (EVS) or open surgery. The decision to proceed with more invasive treatments should consider the patient's overall health, symptom severity, and availability of local expertise.

Conclusion

Nutcracker syndrome is an uncommon cause of recurrent gross or microscopic hematuria. Due to its variable and non-specific presentation, shared decision-making (SDM) is crucial to ensure

patients are informed about their treatment options, whether conservative or surgical. Tailoring management based on patient symptoms and risks remains essential for optimizing outcomes.

病例報告 113_C38

克隆氏症復發：以結節性紅斑為初始表現

A Case Report of recurrent Crohn's disease, manifesting with Erythema nodosum

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Introduction

Crohn's disease(CD) is an inflammatory disease not only involving the gastrointestinal tract, but also presenting with extra intestinal manifestations (EIMs). The EIMs can impact various organ systems, such as muscle, joints, skin, hepatobiliary tract, lung, eyes and vascular system. The skin is one of the commonly involved organs, and studies showed that approximately 5%–15% of the cases have skin involvement of erythema nodosum (EN). However, there are few reports of new-onset EIMs with skin involvement during the recurrence of Crohn's disease. We presented a case with Crohn's disease with 2nd recurrence, characterized by skin involvement in the form of EN, which had not occurred during previous flare-ups.

Case Report

A 32-year-old female had been suffering from Crohn's disease for 6 years, initially presenting with chronic diarrhea and bloody stools. She began treatment with the anti-TNF agent Adalimumab in February 2019 and responded well to the medication. Adalimumab was discontinued in December 2021 due to near-complete relief of symptoms and National Health Insurance regulations. However, five months after stopping the medication, she developed painful red nodules on both lower limbs.

Physical examination revealed red and purple, tender, and painful lumps on both shins, leading to a diagnosis of erythema nodosum (EN) by the rheumatologist. Three weeks later, she experienced a recurrence of diarrhea with blood-tinged stools and intermittent fever reaching up to 39°C.

Given the recurrence of Crohn's disease, Adalimumab treatment was resumed in May 2022. One month later, she reported significant improvement in diarrhea, along with darkening and shrinkage of the skin lesions. She continued on therapy and the skin lesions had almost completely resolved several months later.

Discussion

Erythema nodosum (EN) typically appears on the surface of the lower limbs, manifesting as a sudden onset of symmetrical, erythematous, warm, painful, and non-ulcerative nodules ranging from 1 to 5 cm in diameter. Systemic symptoms such as fever, fatigue, and arthralgia may accompany the nodules. Generally, a skin biopsy is not necessary, as the diagnosis of EN is primarily based on clinical presentation.

In this case, her previous recurrences of Crohn's disease were confined to the gastrointestinal tract. This was the first instance in which she experienced a skin manifestation of erythema nodosum (EN), which also served as an early indicator of disease recurrence. The exact correlation between Crohn's disease flare-ups and erythema nodosum remains uncertain.

Current research suggests that they may share common genetic risk factors (e.g., HLA-DR2,

PTGER4, ITGAL, SOCS5, CD207, and ITGB3) and environmental factors, such as smoking. Additionally, there are hypotheses that extraintestinal manifestations (EIMs) could be influenced by microbiota and cross-reactivity of the immune response in the gastrointestinal tract. Although the exact pathogenesis and correlation between EN and Crohn's disease are still unclear, most cases of EN are self-limiting within 3-6 weeks without specific treatment. Almost all cases of EN resolve over time, often with the aid of anti-inflammatory medications.

Conclusion

Clinicians should be aware of EIMS when managing patients with Crohn's disease. EIMs can sometimes be the first sign of recurrence and early recognition and prompt treatment are important for effective disease control.

病例報告 113_C39

使用 Vedolizumab 和 Upadacitinib 組合療法於困難治療的潰瘍性結腸炎患者：一病例報告

Combination Therapy with Vedolizumab and Upadacitinib for Difficult-to-Treat Ulcerative Colitis: A Case Report

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Introduction

Ulcerative colitis (UC) is a chronic bowel disease causing continuous colonic mucosal inflammation, typically starting from the rectum and potentially extending proximally. [1] UC incidence is rising globally with North America 8.8-23.1/100,000, Europe 0.6-24.3/100,000, Oceania 7.3-17.4/100,000 annually. It often appears in people aged 20-40, with no major gender difference. [2] Management focuses on relieving symptoms, promoting mucosal healing, and improving quality of life. First-line treatment for mild to moderate UC includes 5-aminosalicylates. [3] Immunosuppressants and biologics become necessary when first-line treatments fail. This case report explores the efficacy of combining Vedolizumab with Upadacitinib to achieve complete mucosal healing in a patient with UC who failed biologic treatments.

Case Report

A 21-year-old female was diagnosed with UC, initially presenting with bloody stools. Her treatment began with oral and rectal mesalazine and azathioprine therapy. Due to an inadequate response, a short course of methylprednisolone (4 mg twice daily) was added. Colonoscopy later revealed persistent inflammation from the rectum to the descending colon. She subsequently received biologic therapy with Vedolizumab due to ongoing bloody stools. Although there was partial symptom improvement, a follow-up colonoscopy showed persistent active inflammation. The decision was made to switch the biologic agent to Ustekinumab due to inadequate response to Vedolizumab. Despite this, endoscopy still revealed active colitis, and her clinical symptoms worsened. Ustekinumab was discontinued after the loading dose, and Vedolizumab was reused. Due to ongoing bloody stools, 15 mg of Upadacitinib was added. Following this combined therapy, the patient experienced significant improvement in stool frequency and rectal bleeding. A follow-up colonoscopy demonstrated marked improvement in mucosal inflammation, with a Mayo Endoscopic subscore of 0 in the sigmoid colon.

Discussion

Refractory UC has significant treatment challenges after first-line biologic therapies fail. Vedolizumab has shown efficacy in treating UC through its action on α4β7 integrin, which reduces leukocyte migration into the gut.[3] However, some patients do not achieve adequate remission. Additionally, combination biologics and small molecule therapy may provide some benefit in the refractory UC. Upadacitinib, a selective JAK1 inhibitor, offers a novel mechanism of action that may serve as an alternative treatment. [4] This case illustrates the potential benefits of combining Vedolizumab with Upadacitinib in refractory UC. The combination therapy led to complete mucosal healing after failure of multiple biologics treatment, highlighting its effectiveness in

achieving symptom relief and improvement in colonoscopy when the traditional approaches fail.

Conclusion

For patients with UC who have failed first-line biologics, advanced therapy combinations represent a promising therapeutic approach. This case report demonstrates that such a combination can achieve complete mucosal healing and significant clinical improvement, offering hope for patients with refractory UC. Continued research and clinical trials are necessary to validate these findings and refine treatment strategies for this challenging condition.

病例報告 113_C40

糞石未完全溶解引起的腸阻塞

Intestine obstruction causing by an incomplete resolved bezoar

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Introduction

Bezoars obstruction is a rare etiology of small bowel obstruction, and need to be remove by surgical procedure, either laparoscopic or laparotomy. In regard to the gastric bezoars, there are many ways to deal with it, we can try Coca-Cola® or digestive enzyme or remove by endoscopic devices or operation, but none of them have standard protocols.

Case Report

A 72-year-old man with type 2 diabetes mellitus and hypertension was evaluated because of a long term epigastric pain at National Cheng Kung University Hospital, where panendoscopy reorted a huge bezoar lying in the stomach, Coca-Cola® was suggested by the physician. But clinical abdominal fullness persists so he visited our hospital for help, where revealed Huge bezoar over gastric body with an estimated 1.5cm clean-base ulcer over antrum near pyloric ring via panendoscopy. Abdominal CT showed that bezoar was stuck in ileum with diffuse small bowel obstruction. Abdominal ultrasonography revealed a bezoar of curved acoustic impedence with acoustic shadow lodged within instestine. Laparoscopic ileotomy for removal of bezoar was performed on the next day. Patient was fully recovered and discharged smoothly.

Discussion

Most common cause of small bowel obstruction in adult including postoperative or inflammatory adhesive disease, herniation and malignancy. Bezoars are an unusual cause of small bowel obstruction, and only accounting for 4% of all etiology. For gastric bezoar, the treatments include dissolution by Coca-Cola®, removal by endoscopic devices, laparotomy and laparoscopic surgery. As for the intestinal bezoars, surgical procedure is the only way to removal, either by laparoscopic or milking via laparotomy. Suppose that the carbon dioxide bubbles in the beverages may penetrate into the bezoars through the microscopic pores on its surface and destruct the structure of the outer layer.

Conclusion

if gastric bezoar still remain intact and lying in the stomach, we may give it a try to carbonated drinks according to the theories. But if the shattered bezoar get stuck in the small bowel, we should early recognize the clinical symptoms and using abdominal ultrasonography or abdominal computed tomography for diagnosis and arrange operation for removal as soon as possible.

病例報告 113_C41

長隧道型開放性卵圓孔引起之直立低血氧症候群：病例報告分享

A case of Orthodeoxia syndrome caused by long tunnel patent Foramen Ovale

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Introduction

An Orthodeoxia syndrome is characterized by hypoxemia when upright position, and resolved in supine position. It may cause shortness of breath and limitation of daily activity. Orthodeoxia syndrome may caused by many conditions of patient, including intracardiac shunting, intracardiac shunting, and other rare condition.

Case Report

A 77-year-old male with history of old ischemic stroke was admitted to infection disease ward because of right femur osteomyelitis. The patient experienced progressive hypoxemia, with the lowest SpO2 of 77% occurring after being in the upright position for approximately 30 seconds, which completely resolved upon returning to the supine position. The heart and vessel CT angiography revealed no pulmonary embolism, and no obvious pulmonary parenchyma abnormality was noted. A transthoracic echocardiography showed no intracardiac shunting could be detected due to poor acoustic window. Therefore, a further agitated saline test was performed, revealing the early appearance of massive micro-bubbles in the left heart within two cardiac cycles. A transesophageal echocardiography (TEE) confirmed the presence of a tunnel-type (18 mm long and 4 mm wide) patent foramen ovale (PFO) with continuous right-to-left shunting. The patient was scheduled for percutaneous PFO closure. An Amplatzer 25mm-18mm occluder was successfully implanted under intraoperative TEE and fluoroscopy guidance. After the intervention, the patient reported no further episodes of orthodeoxia in upright position.

Discussion

An Orthodeoxia syndrome is a rare condition that characterized by arterial desaturation in the upright position, and improved in the supine position. The causes of orthodeoxia syndrome including an intracardiac shunt, intrapulmonary shunt, or abdominal cause. Upright position could stretch the interatrial communication, causing more streaming of venous blood from inferior vena cava through the interatrial defect. The agitated saline test, or ”bubble study” is a very useful procedure to detect an intra-cardiac right to left shunt, as in our case.

The long tunnel PFO is one of the various anatomical patterns of PFO. There is no clear definition of long tunnel PFO, but in general, overlap of 8-10mm between the entry and the exit point between the septum primum and secundum. The percutaneous intervention for long tunnel PFO is challenging because of the anatomy and limitation of size of PFO occluder.

Conclusion

We described a case of long tunnel PFO induced orthodeoxia syndrome which was discovered via agitated saline test and was successfully treated with percutaneous PFO closure under

intraoperative TEE and fluoroscopy guidance. The closure of long tunnel PFO is challenging because of it's anatomical complexity and limit size of device. It's important to aware that in the patient of orthodeoxia syndrome, PFO should be taken into the differential diagnosis.

病例報告 113_C42

HHV-8 相關多中心卡斯特曼病與卡波西氏肉瘤：一例病例報告

HHV-8 associated multicentric Castleman disease and Kaposi's sarcoma: a case report

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Introduction

Multicentric Castleman disease (MCD) were caused by Human Herpesvirus 8 (HHV8) in AIDS patients.1 Kaposi's sarcoma (KS) also has several types, and the most common type being AIDS-related. This case highlights the relationship between MCD, KS, and opportunistic infection (OI) in AIDS.

Case Report

We reported a 41-year-old man with fever and non-productive cough for 3 days. According to his statement, the diagnosis of human immunodeficiency virus (HIV) infection was made 17 years ago, and he did not receive combination antiretroviral treatment (cART). Bictegralvir/tenofovir alafenamide /emtricitabine (BIC/TAF/FTC), fluconazole, and trimethoprim/sulfamethoxazole (TMP/SMX) were given. The HIV viral load was 2540000 IU/ml and the CD4+ T cell count was 57 cells/μL. AIDS was confirmed. He was admitted for further management.

After admission, the septic work-up showed all negative results. His fever also improved later, and anidulafungin and levofloxacin were discontinued. He was discharged on day 14.

However, a recurrent fever occurred on day 24. An influenza rapid test showed a positive result for influenza B, and zanamivir was administered. Despite some clinical improvements, the fever persisted. He was admitted on day 30.

After admission, he experienced a fever, but his vital signs remained stable. A bone marrow study was conducted, which showed scattered human herpesvirus (HHV8)+ cells. The picture suggested an HHV8-related disease. Because the bone marrow findings were inconclusive, a lymph node biopsy was necessary to confirm the diagnosis. Therefore, a left neck lymph node excision biopsy was performed. Microscopically, HHV8+ cells were noted, especially in the mantle zones. This histological variant typically corresponds to HIV+ HHV8+ MCD clinically. 4 cycles of rituximab 500 mg were administered, and he was discharged without any complications.

On day 146, a fever was noted, and the right neck LAP recurred. He was admitted for a right neck lymph node excision biopsy on day 155.

A right neck lymph node excisional biopsy was performed, and the specimen showed diffuse HHV8 and CD31-positive cells, leading to the diagnosis of KS. 6 cycles liposomal doxorubicin 40 mg was given. The treatment with liposomal doxorubicin has been completed.

Discussion

HHV8-associated MCD is the most common MCD in HIV patients. The clinical manifestations of MCD can include fever, generalized LAPs, weight loss, hepatosplenomegaly, or leg edema. The laboratory abnormality can be elevations in inflammatory markers, hypoalbuminemia, anemia, or even to pancytopenia.[1] The symptoms and lab data are similar with OI in AIDS. KS is an AIDS-

defining malignancy, and the most common type is AIDS-related KS. The clinical manifestations of KS varied. The KS lesions are vascular nodules, which are most commonly found on the skin or mucosa. In this case, the KS lesions are only confined to lymph nodes. As a result, OI in AIDS, MCD, and KS may be hard to distinguish from each other. In AIDS patients with fever and LAP, OI should be considered first.

The standard treatment for MCD is 4-6 cycles of rituximab.[3] Fever and generalized LAPs recurred after the rituximab treatment. The possible etiologies might include MCD relapse, lymphoma, or KS.[1,2,4]

Conclusion

This case illustrated the occurrence of MCD and KS in an AIDS patient. The presentation may be similar in OI of AIDS, MCD, and KS. Definite treatment is effective with a proper diagnosis.

病例報告 113_C43

糖尿病酮症酸中毒併發血栓性血小板低下紫斑症

Immune Thrombotic Thrombocytopenic Purpura and Diabetic Ketoacidosis: A Case Report and Literature Review

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Introduction

Thrombotic thrombocytopenic purpura (TTP) is an uncommon and life-threatening disorder caused by a deficiency of ADAMTS-13, and eventually leads to microangiopathic hemolytic anemia, severe thrombocytopenia, and organ damages. Acute TTP events could be triggered by infections, or inflammations while ADAMTS-13 deficiency[1]. Moreover, diabetic ketoacidosis (DKA) represents one of the most common life-threatening complications of diabetes mellitus, and the metabolic decompensation precipitated by underlying medical illness are the cause of death[2]. Recently, several case reports showed that TTP or similar thrombotic microangiopathy could developed concomitantly with DKA. Here, we present a case with concomitant presentation of DKA and TTP.

Case Report

We presented a 37-year-old male with a history of diabetes mellitus and presented with typical symptoms of diabetic ketoacidosis. He was managed with an insulin pump and intravenous fluids. However, he developed hemodynamic instability after blood transfusion in the following days, necessitating continuous renal replacement therapy, intubation, and inotropic support. Laboratory data indicated hemolytic anemia and thrombocytopenia, and a blood smear revealed schistocytes. The PLASMIC score was 5, and ADAMTS-13 activity was 2%. The patient was diagnosed with TTP and treated with therapeutic plasma exchange, steroids, and rituximab. His platelet count stabilized above 150,000/μL, and ADAMTS-13 activity progressively improved.

Discussion

Both the PLASMIC[3] and French scores[4] are valuable tools for predicting severe ADAMTS-13 deficiency before test results are available. In this case, however, the PLASMIC score indicated a 5-24% risk, and the French score suggested a 70% risk of TTP, leading to intermediate predictions. Studies have shown that MCV <90 fL may not be an effective predictor of TTP in East Asian populations[5,6], as seen in this patient. Kidney injury, often less severe in TTP compared to hemolytic uremic syndrome (HUS)[7], was complicated by DKA in this case, potentially affecting the accuracy of the prediction models.

This case is the second documented instance of definite TTP complicated by DKA, and the first to show a clinical and ADAMTS-13 response. The interaction between TTP and diabetes mellitus remains poorly understood, but proposed mechanisms include diabetic microangiopathic changes, increased VWF expression, and decreased ADAMTS-13 levels in diabetic nephropathy. These factors may heighten the severity of TTP and increase the likelihood of acute events[8].

Conclusion

This case report highlights the association between DKA and TTP, presenting the rare complication of acute renal failure in TTP. TTP is a rare and serious disease that requires prompt recognition and management. Concurrent conditions and population should be considered when calculating prediction scores including the PLASMIC and French scores.

病例報告 113_C44

晚期肝細胞癌治療反應良好但致肝衰竭：解剖病例報告

Advanced hepatocellular carcinoma with a good treatment response but fatal liver failure: an autopsy case report

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Introduction

Hepatocellular carcinoma (HCC) remains the leading cause of cancer-related death in Taiwan. Immune checkpoint inhibitors combined with anti-VEGF therapy has transformed the treatment landscape of advanced or metastatic HCC, with around 10% patients achieving disease-free status. In this report, we present an autopsy case of advanced HCC that responded well to immunotherapy but ultimately resulted in death due to decompensated liver disease.

Case Report

An 80-year-old man presented with progressive dyspnea on exertion and chest tightness for 1 month. A chest X-ray revealed multiple nodules over bilateral lung fields. A whole body computed tomography scan (CT) disclosed multiple tumors at bilateral lung and right hepatic lobe, with direct invasion to the right portal vein, right hepatic vein, inferior vena cava and right atrium. A biopsy to the hepatic tumor revealed HCC.

A combination therapy of lenvatinib (8 mg orally QD) and pembrolizumab (100 mg intravenously every 3 weeks) was initiated. Palliative radiotherapy (52.5 Gy in 15 fractions) was also conducted to the invasive portal vein tumors. Serial CT scan assessments at 3rd, 6th and 9th months post treatment disclosed shrunken hepatic tumors, vascular and cardiac thrombi, as well as complete resolution of lung metastases. (Figure 1).

Nevertheless, decompensated liver cirrhosis developed starting from the 9th month post-treatment. A re-biopsy of the residual hepatic tumor revealed a few atypical cells, surrounded by elastic stroma and immune cell infiltrates. Lenvatinib was continued. However, the patient's liver function deteriorated, and he eventually passed away in the 12th month. An autopsy disclosed no viable tumor cells but massive liver necrosis, associated with venous thrombosis, hepatic arterial obliteration, and cirrhosis.

Discussion

We presented a first autopsy case of advanced HCC that responded well to a combination of lenvatinib plus pembrolizumab and radiotherapy. However, he eventually passed away due to decompensated liver function.

Lenvatinib, a multi-target tyrosine kinase inhibitor, is known for arterial thrombotic events (5%), but hepatitis events are very rare (<0.5%). Pembrolizumab was designed to target and block programmed cell death protein 1, helping the immune system recognize and attack cancer cells more effectively. Immune-mediated hepatotoxicity is very rare, often characterized by panlobular lymphocytic infiltration and spotty necrosis[1], which were not detected in this case. Radiotherapy is one of the locoregional therapies for unresectable or inoperable disease. Classic radiotoxicity,

presented as Veno-Occlusive Disease[2], was not observed in this patient.

Conclusion

The adverse effects of the aforementioned treatments alone cannot fully explain the cause of the patient's liver failure. Interactions between treatments may result in additive harm. More studies are warranted for a better understanding. In this case, persistent radiological lesions were observed. However, the final pathology report revealed no viable tumor cells. We have defined this phenomenon as a "ghost" tumor in our subsequent research[3].

病例報告 113_C45

次世代基因定序應用於急性呼吸道窘迫症候群的病原診斷：一例嚴重鉤端螺旋體病的病例報告

A Case Report of Leptospirosis Diagnosed by NGS_ A Useful Tool for Decision-Making in the ICU in Taiwan

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Introduction

Leptospirosis is a zoonotic infection with diverse clinical presentations, ranging from mild symptoms to severe, life-threatening disease. In critical cases, it can cause multiorgan failure and death. Diagnosis is typically based on clinical suspicion and confirmed by laboratory testing. However, in severe cases, obtaining a history of exposure and recognizing early symptoms may be challenging. Traditional diagnostic methods for infectious diseases are often slow, possibly delaying precise treatment. Next-generation sequencing (NGS) has emerged as a novel diagnostic tool that identifies pathogens using DNA or RNA from bodily fluids, offering more timely results.

Case Report

We present the case of a 49-year-old male chicken vendor with a history of diabetes mellitus and dyslipidemia, who developed progressive dyspnea over four days. He initially sought care at a local clinic for dizziness, weakness, and chest tightness, and was treated symptomatically for suspected heat stroke. His symptoms worsened, and he was transferred to hospital A due to cyanosis and altered consciousness. After developing rapidly progressive hypoxemia, he was intubated. Chest CT ruled out pulmonary embolism, and he was treated empirically for pneumonia with broad-spectrum antibiotics. However, his condition deteriorated, necessitating VV-ECMO, and he was transferred to our hospital in shock with multiorgan failure. His family provided no recent travel or living animal contact history.

CT scans revealed diffuse ground-glass opacities and patchy consolidations in both lungs, with no pleural effusion or interstitial fibrosis, raising suspicion of severe pneumonia. Despite thorough testing, conventional diagnostic tests for infectious pathogens were either negative or pending. Given the urgency, metagenomic NGS was performed on respiratory samples, which identified Leptospira interrogans within 24 hours. Antibiotics were promptly adjusted. After extubation on the 9th day of admission, the patient recalled several rat bites at work, previously unreported to his family. His condition gradually improved, and he was discharged without significant complications. Three weeks later, serologic testing from the CDC confirmed the diagnosis of leptospirosis.

Discussion

Timely identification of pathogens in critically ill patients with severe infections can be difficult due to the empirical use of antibiotics, the fastidious nature of some organisms, and the lengthy turnaround times of traditional diagnostic methods, including cultures and antibody tests. While rapid antigen tests and molecular tests, such as PCR panels, exist, they are limited in scope and

may not account for regional variations in pathogens. For critically ill patients, timely and accurate diagnosis is essential not only for guiding appropriate treatment but also for discussing prognosis and care strategies with the family, including whether an aggressive or conservative treatment approach is warranted. NGS, with its rapid turnaround time and flexibility in detecting a wide range of pathogens, offers a valuable tool in achieving a prompt diagnosis in these challenging cases.

Conclusion

In critically ill patients with severe infections, metagenomic NGS significantly improves the chances of timely diagnosis and appropriate treatment. It also provides clinicians with greater confidence when discussing prognosis and care options with family members.

病例報告 113_C46

破傷風造成之急性呼吸衰竭與心律不整：病例報告

A case report of tetanus inducing acute respiratory failure and arrhythmia

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Introduction

Tetanus is caused by exposure to the spores of the bacterium Clostridium tetani, found in soil, saliva, dust, and manure. These bacteria can enter the body through deep cuts, wounds, or burns, affecting the nervous system and leading to painful muscle contractions, notably in the jaw and neck muscles, but could also involve respiratory muscle causing respiratory failure and sympathetic tone causing arrhythmia. We present a case who received a tetanus vaccination after a car accident trauma but developed dysphagia, dysarthria, respiratory failure, and autonomic failure within two weeks, leading to a diagnosis of tetanus.

Case Report

This 82-year-old man presented to our emergency department with dysphagia and dysarthria for 3 days. Tracing back his history, he has been diagnosed with heart failure and atrial fibrillation. He had a history of a motorcycle accident two weeks prior, which resulted in abrasions on the left arm and wrist, and he received tetanus vaccination meanwhile. However, the patient returned to the hospital three days later due to worsening dyspnea. He was unable to speak due to trismus (Figure.1). Increased muscle tone over four extremities was also noted. As a result of respiratory distress and difficulty in clearing respiratory secretions, the patient had to be intubated. An electrocardiogram showed the atrial fibrillation with left bundle branch block, and the patient's heart rate was recorded at 102 beats per minute. Considering motorcycle accident history, trismus, and increased muscle tone, tetanus was suspected. Electromyography exam showed marked increased insertional activity and continuous motor unit activity in left masseter muscle compatible to typical tetanus finding (Figure.2). Human tetanus immune globulin was given.
Following injection of human tetanus immune globulin, increased muscle tone resolved in a week. However, few episodes of cardiogenic shock, complete AV block, with concurrent right bundle branch block, left bundle branch block were noted (Figure.3). Autonomic failure related to tetanus was suspected which finally leading to a permanent pacemaker implantation. Follow-up electromyography two weeks later showed less spontaneous continuous motor unit activities when compared with the previous study. The endotracheal tube was successfully removed 30 days after insertion. At the time of discharge, the patient exhibited normal limb function and was able to walk without assistance.

Discussion

Tetanus, once highly fatal, has become rare with widespread vaccination. In Taiwan, it's a notifiable disease, with fewer than 20 cases annually since 1981. Our patient, vaccinated after a car accident, still developed tetanus, progressing to respiratory failure and arrhythmias, requiring a pacemaker. This case highlights that tetanus vaccination prevents future infections but doesn't

neutralize toxins already present, as neuronal binding of tetanus toxin is irreversible. Diagnosis depends on clinical history and symptoms, especially in older patients who may lack childhood vaccinations. Treatment includes wound debridement, metronidazole, tetanus immune globulin, minimizing external stimuli, and early tracheostomy if needed.

Conclusion

Tetanus, an ancient disease, has gradually been forgotten with the widespread use of vaccines since World War II. The case emphasizes the importance of thorough epidemiological investigation and vigilant care in managing rare diseases.

病例報告 113_C47

Ramsay-Hunt 症候群併水痘帶狀皰疹病毒腦炎 – 一個罕見病例

An Unusual Case of Ramsay-Hunt Syndrome with Varicella-zoster Virus Meningoencephalitis

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Introduction

Ramsay-Hunt syndrome (RHS), or herpes-zoster oticus, is a complication of varicella-zoster virus (VZV) infection. It features a classical triad of ipsilateral peripheral facial paralysis, otalgia, and otic vesicular rashes. We herein reported a case of herpes zoster with RHS, complicated with facial, vestibulocochlear, glossopharyngeal neuritis, and medullary encephalitis.

Case Report

A 48-year-old man without known medical illness presented with a right ear pain for three weeks. He was diagnosed with acute otitis media at a clinic three weeks prior to admission, but his otic pain persisted despite antibiotic treatment. He sought care at another medical facility after vesicular rashes appeared on his ear accompanied by acute hearing loss, right facial palsy, and gait disturbance a week later. The diagnosis of RHS was made, for which he received valaciclovir (500 mg twice a day for 7 days) and oral prednisolone (80 mg/day for 4 days, 45 mg/day for 3 day). However, a fever developed with hiccups, hoarseness, progressive dysphagia, and non-productive coughs one week before admission, so he sought medical attention again. Computed tomography demonstrated dense ground-glass opacities in his right lower lung; therefore, he was referred to our hospital.

On physical examination, his consciousness was intact. Crusty vesicles in his right external acoustic canal and a peripheral-type facial paralysis were observed. Laboratory data indicated mild leukocytosis (WBC 10.35 K/μL) and an elevated alanine aminotransferase (ALT 134 U/L). A lumbar puncture was conducted with an opening pressure of 12.8 cmH2O, while the cerebrospinal fluid analysis revealed lymphocyte-predominant pleocytosis (WBC 11 x 11/9/μL, all lymphocytes), glucose 66 mg/dL, total protein 30.3 mg/dL, and detection of VZV by multiplex polymerase chain reaction. The HIV antibody-antigen combination assay was negative and his HbA1c was 6.0%. The test results for anti-cytokine antibodies, including anti-interferon autoantibodies, were negative. Gadolinium-enhanced magnetic resonance imaging (MRI) demonstrated thickening and enhancement of the right facial, vestibulocochlear, glossopharyngeal nerves, and two tiny foci of hyperintensity on FLAIR/DWI at his right posterior medulla, along with meningeal enhancement. The diagnosis of herpes zoster complicated with RHS and meningoencephalitis was made. The patient was treated with intravenous acyclovir (750 mg thrice a day, 14 days) targeting VZV infection, and intravenous ampicillin-sulbactam (1000 mg four times a day, 7 days) for aspiration pneumonia. He was discharged with a nasogastric tube, continuing his rehabilitation in the follow-up clinic.

Discussion

Neurological complications of VZV infection include meningoencephalitis, RHS, herpes-zoster

ophthalmicus, myelitis, and postherpetic neuralgia. Among which, RHS predominantly affects the facial and vestibulocochlear nerves due to the inflamed geniculate ganglion. Involvement of multiple cranial nerves is less common and is mostly seen among immunocompromised patients, implying a worse neurological prognosis. For our patient, the lumbar puncture and MRI were prompted by an unusual manifestation and severity of RHS, which raised the suspicion of a brainstem encephalitis. Despite being negative for common risk factors such as diabetes and HIV infection, other immunocompromised conditions should be carefully evaluated.

Conclusion

RHS is a neurological complication of VZV infection, whereas meningoencephalitis should be ruled out in the event of multiple cranial nerve neuritis.

病例報告 113_C48

以經口膽道鏡併電震碎石術成功處理總膽管結石合併取石網嵌頓

Basket impaction with an entrapped common bile duct stone successfully treated with peroral cholangioscopy with electrohydraulic lithotripsy: a case report

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Introduction

Choledocholithiasis is a common indication for endoscopic retrograde cholangiography and pancreatography (ERCP). Stones larger than 1 cm present a significant challenge, often resulting failure of conventional endoscopic extraction methods and necessitating mechanical lithotripsy. Entrapped mechanical lithotripsy is a severe complication, which requires rescue procedures to avoid surgery. This case report describes the successful use of cholangioscopy-guided electrohydraulic lithotripsy (EHL) after the failure of various rescue methods.

Case Report

A 64-year-old female with a history of recurrent cholangitis and choledocholithiasis presented for further management of residual common bile duct (CBD) stones. She had previously undergone multiple ERCP procedures, including sphincterotomy, endoscopic papillary balloon dilation, and mechanical lithotripsy. Residual CBD stones were detected on a follow-up imaging, prompting her current admission.

During the ERCP, cholangiography revealed multiple filling defects in the CBD, with the largest stone measuring 1.6 cm. Endoscopic sphincterotomy, followed by endoscopic papillary balloon dilatation was performed without complication. However, the large CBD stone could not be removed by balloon lithotripsy. Mechanical lithotripsy was subsequently attempted, but the basket became impacted with captured stone. Additional attempts failed, including using balloon lithotripter to manipulate of the impacted basket. A rescue lithotripter handle was also unsuccessful due to challenging angulation at the ampulla.

Peroral cholangioscopy using the Boston Scientific SpyScope DSII was then employed, and EHL was performed, delivering a total of 964 pulses. This approach successfully fragmented the stone. Follow-up cholangiogram showed multiple smaller filling defects in the middle to distal CBD. Fragmented stones, and the trapped basket were successfully removed. The patient was discharged two days later without complications.

Discussion

Mechanical lithotripsy is a standard technique for managing large bile duct stones, but can lead to significant complications, such as the entrapment of both the stone and the lithotripsy basket.[1] Traditional salvage methods, including larger balloon dilation and the use of rescue extra-endoscopic mechanical lithotripters, are not always effective in resolving these issues. Cholangioscopy combined with EHL offers a promising alternative, providing direct visualization and precise stone fragmentation using high-energy shock waves. This minimally invasive approach reduces the need for surgical intervention, although its high cost remains a limitation.[2,3]

Nonetheless, EHL is an effective rescue technique when conventional methods fail.

Conclusion

This case highlights the novel approach of using cholangioscopy combined with EHL as an effective rescue method. This technique provides timely and minimally invasive management in the endoscopic room, potentially avoiding the need for further surgical intervention.

病例報告 113_C49

個案報告: 以溶血性貧血表現之 B12 缺乏

A patient of Vitamin B12 deficiency presented as hemolytic anemia

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Case Report

A 67-year-old man presented to the emergency department with progressive exertional dyspnea that had developed over the past two weeks.

The patient suffered from decreased appetite, abdominal pain, nausea, vomiting after meals, and altered taste perception for three months. Despite being prescribed Simethicone, Mosapride, and Oxethazaine, his condition improved only minimally. After that, he began experiencing intermittent diarrhea and exertional dyspnea, along with unintentional weight loss from 110 kg to 86 kg over the three-month period. Consequently, he visited our emergency department, where the initial examination showed pale conjunctiva, icteric sclera, and epigastric tenderness.

Laboratory findings indicated pancytopenia, a low Reticulocyte Production Index, and indirect hyperbilirubinemia. The anemia was macrocytic, with markedly elevated lactate dehydrogenase (LDH) levels and decreased haptoglobin.

A comprehensive workup for pancytopenia was conducted. The patient's vitamin B12 level was significantly low, while folic acid levels were normal. A blood smear revealed immature myeloid cells and hypersegmented neutrophils, with no fragmented red blood cells. Imaging studies, including a whole-body CT scan, showed no splenomegaly. The patient tested negative for antinuclear antibodies (ANA), anti-extractable nuclear antigens (Anti-ENA), and Coombs' test. Flow cytometry of peripheral red blood cells showed no PNH clones. A bone marrow study demonstrated hypercellularity, with a myeloid-to-nucleated erythroid cell (ME) ratio of approximately 1:4, megaloblastic changes, no evidence of hemophagocytosis, negative iron and reticulin stains, and CD34 and CD117 positive blasts not increased. Overall, the bone marrow study was compatible with megakaryocytic hyperplasia, typically associated with thrombocytopenia.

A subsequent upper gastrointestinal endoscopy identified multiple ulcerative lesions in the gastric mucosa or submucosa. A biopsy revealed atrophic gastritis and a neuroendocrine tumor. Following component therapy and parenteral vitamin B12 supplementation, the patient's symptoms improved within two days, and his hemogram and LDH levels gradually normalized over the next two months.

Discussion

We present a case of pernicious anemia manifesting as pancytopenia, leukoerythroblastosis and hemolytic anemia, initially raising concerns about hematological malignancy. Pernicious anemia is characterized by anemia associated with vitamin B12 deficiency and gastric mucosal atrophy, resulting from autoantibodies targeting intrinsic factor and/or gastric parietal cells. Although often overlooked, the prevalence of pernicious anemia is estimated to be 2-3% among the elderly [1]. While it typically presents as megaloblastic anemia, vitamin B12 deficiency can, in rare instances, present with pancytopenia. Laboratory findings may include markedly elevated levels of lactate

dehydrogenase (LDH), attributable to both ineffective intramedullary erythropoiesis and a reduction in red blood cell lifespan [2]. Symptoms of vitamin B12 deficiency can be effectively resolved through parenteral supplementation, with minimal side effects [3]. Nevertheless, patients with pernicious anemia have an increased risk of gastric adenocarcinoma and neuroendocrine tumors; therefore, endoscopic surveillance is recommended every 3-5 years [4]. There is a pressing need to raise awareness of this condition among clinical practitioners.

病例報告 113_C50

首例亞洲家族謎樣症候群

First Reported Case of RIDDLE Syndrome in an Asian Family: Unraveling the Riddle

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Introduction

Riddle syndrome is a rarely identified yet potentially life-threatening hereditary disease entity. The clinical phenotypes are characterized by the constellation of compromised body figure development and intellectual maturation, while immunodeficiency and recurrent infections are common. The presentations are nevertheless largely heterogeneous, and the molecular mechanism contributing to such heterogeneity remained enigmatic. No definitive treatments are available thus far, leading to dismal prognosis in severe cases.

Case Report

A 37-year-old Amis lady presented with insidiously onset of dyspnea on exertion that worsened over the past two years. Concomitant productive cough, anorexia, dysphagia, slurred speech with reduced volume, progressive binocular diplopia, and unintentional body weight loss were experienced as well. She could neither sit nor stand up. Her family history was significant for cluster of infertility. Due to hypoprolactinemia-related premature ovarian insufficiency, she tried oral steroid with hormone replacement therapy, predisposing repeat hospitalization for recurrent pneumonia and empyema. Upon presentation, she was cachectic with body mass index at 16 kg/m^2. Physical examination revealed a short stature, microcephaly, stiff truck, bibasilar crackle breathing sound on auscultation, digital clubbing, and absent ankle jerks. Pulmonary function test showed restrictive ventilatory pattern, and World Health Organization group 3 pulmonary hypertension was complicated. Nerve conduction study revealed prolonged distal latencies, while electromyography was compatible with myopathic changes. Muscle biopsy reported mixed pattern with predominant neuropathic changes. Pleuroscopy was performed concerning refractive exudative effusion, which demonstrated thick infiltrative mucosa over parietal pleura, multiple adhesions, and fibrinous tissues, raising suspicion for malignancy. Tumor marker panels intriguingly showed high alpha-fetoprotein level at 443 ng/mL. Despite decortication and prolonged antibiotics, mechanical ventilation was necessitated and difficult weaned because of acute on chronic type II respiratory failure. Finally, whole genome sequencing was performed to assess genetic disorder. Germline mutation at Really Interesting New Gene Finger Domain (RNF) 168 c.91 T>C was present, confirming diagnosis of Riddle syndrome. Family screening revealed the same homozygous mutation in her two sisters as well, while the father was an asymptomatic carrier. Currently, the patient remained ventilator-dependent and under active evaluation for lung transplantation.

Discussion

We herein described the first reported family cluster of Riddle syndrome in Asia, characterized by recurrent pulmonary infection, gastrointestinal involvement, and polyneuropathy. Loss-of-

function truncation and frameshift mutation of RNF168 have been intertwined by *in vivo* model with compromised genomic integrity due to defects in ubiquitin-mediated repair mechanisms of deoxyribonucleic acid double strand break and altered p53-binding protein 1 recruitment, resulting in impacted lymphocyte maturation and immunodeficiency. Interestingly, our case presented constantly normal level of immunoglobin, and her two siblings remained otherwise asymptomatic aside from dysmorphic body figures. Additional mutations, *e.g.* ATM heterozygosity as suggested in a previous case, might account for the observed variability in clinical presentations. As for clinical implication, the constellation of these phenotypes conjunctionally with elevated serum level of alpha-fetoprotein may serve as screening criteria for further genetic testing to identify Riddle syndrome.

Conclusion

Riddle syndrome has been increasing recognized through next-generation sequencing to identify RNF168 deficiency. However, the clinical presentation remained highly variable across different cases.

病例報告 113_C51

病例報告：70 歲女性伴隨肝功能衰竭–IgG4 相關自體免疫性肝炎

Case report: IgG4-associated autoimmune hepatitis in a 70-year-old woman with progressive liver failure

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Introduction

Autoimmune hepatitis (AIH) is a chronic liver disease causing inflammation and potentially leading to cirrhosis if untreated. IgG4-related disease (IgG4-RD), on the other hand, is a chronic fibroinflammatory disorder affecting various organs, characterized by elevated serum IgG4 levels and infiltration of IgG4-expressing plasma cells. Hepatic involvement of IgG4-RD is less frequently documented, with the first report of hepatic manifestations appearing in 2007.[1] Since then, similar cases have been reported, leading to the proposal of a new disease entity known as IgG4-associated AIH (IgG4-AIH). This condition is identified by the significant presence of IgG4-expressing plasmacytes in the liver of patients who meet the criteria for classical AIH. Here, we present a case of this rare condition.

Case Report

A 70-year-old woman without underlying diseases presented with elevated liver enzymes over six months. Despite discontinuing herbal medicine, her condition persisted, escalating to malaise, anorexia, and hyperbilirubinemia (total bilirubin [T-bil] 2.74 mg/dL), with AST/ALT levels exceeding 1000 U/L. A CT scan suggested liver cirrhosis without biliary tract abnormality. Blood tests showed an ANA titer of 1:1280 and a serum IgG level of 2587.63 mg/dL, while being negative for viral hepatitis, Wilson's disease, and alpha-1 antitrypsin deficiency. A simplified AIH score was at least 6 points, indicating a probable diagnosis of AIH. She was treated with methylprednisolone and later azathioprine. Liver biopsy 3 days after treatment confirmed chronic active hepatitis with AIH characteristics, and scored as F3 in the METAVIR system. Immunostaining showed IgG4-bearing plasma cells at 4 cells/high power field (HPF) and an IgG4/IgG ratio of 16%. A high serum IgG4 level (262 mg/dL) was also observed. The patient's T-bil peaked at 6.58 mg/dL and, after two days, dropped to 3.41 mg/dL. It normalized by day 41, while serum IgG and IgG4 were within normal limits by day 55, and AST/ALT fell below 100 IU/L by day 69. The patient showed a drastic recovery, effectively managing IgG4-AIH.

Discussion

IgG4-AIH is a new disease entity first reported by Umemura et al in 2007,[1] with several case reports following.[2-7] Our case presented characteristic pathological findings as previous reports, including interface hepatitis, lymphoplasmacytic infiltrates, and emperipolesis. The cutoff for IgG4-expressing plasma cells in the liver varies across studies, with different thresholds such as >10 or >5 cells/HPF. In our case, we observed 4 cells/HPF, possibly influenced by steroid treatment. Although reports show severe inflammation and advanced liver fibrosis, the prognosis is not

poorer.[7] In our case, we observed dramatic improvements under glucocorticoid treatment. This emphasizes the importance of familiarity with IgG4-AIH, as additional immunostaining may be necessary for diagnosis, can predict a favorable response to glucocorticoid therapy, and necessitates further monitoring for systemic IgG4-RD manifestations.

Conclusion

We presented a case of a rare disease: IgG4-AIH, which demonstrated an impressive response to glucocorticoid therapy. This is the first report of IgG4-AIH in Taiwan, providing greater understanding and familiarity with this disease. In patients with AIH who are suspected of having concurrent IgG4-RD, evaluating serum IgG4 levels and conducting additional staining for IgG4-expressing cells may be beneficial.

病例報告 113_C52

次世代基因定序及時診斷及治療廣東住血線蟲腦膜炎：病例報告

Timely Diagnosis and Treatment of Angiostrongylus cantonensis Meningitis Using Next-Generation Sequencing: A Case Report

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Introduction

Angiostrongylus cantonensis is the most common cause of eosinophilic meningitis, which can lead to severe neurologic deficit. Rapid diagnosis and timely treatment are essential to minimize these risks.

Case Report

A 59-year-old woman with a history of hypertension presented with progressive headaches and declining performance status over the past two weeks. Upon arrival at the emergency department, her vital signs were stable, but neurological examination revealed impairments in high cortical functions, including speech, writing, and calculation. Laboratory tests showed eosinophilia in peripheral blood and brain magnetic resonance imaging disclosed diffuse leptomeningeal enhancement. A lumbar puncture revealed an opening pressure of 22 cm H2O, with cerebrospinal fluid (CSF) analysis showing lymphocyte-predominant pleocytosis and elevated total protein, which is compatible with meningitis. Empirical treatment with intravenous ceftriaxone, vancomycin, and acyclovir were initiated. CSF was sent to metagenomic next-generation sequencing (mNGS, Asia Pathogenomics) and nucleic acid amplification test (NAAT, Taiwan Centers for Diseases Control). Both tests subsequently disclosed the presence of Angiostrongylus cantonensis. The patient was treated with albendazole and methylprednisolone. After two weeks of treatment, she was discharged with clear consciousness and no pleocytosis on repeat lumbar puncture. Six months post-discharge, she was fully independent in her daily activities.

Discussion

Research indicates that eosinophilic meningitis from Angiostrongylus cantonensis may result in significant sequelae, including death. Traditional diagnostic methods, like cerebrospinal fluid (CSF) serology, are accurate but time-consuming. In contrast, metagenomic next-generation sequencing (NGS) provides a hypothesis-free approach, delivering faster results. Studies have shown that metagenomic NGS can impact clinical decision-making and expedite diagnostic information. Thus, in cases where the pathogen remains unidentified and the patient's condition worsens, metagenomic NGS is a valuable diagnostic tool. In this case study, the use of NGS reduced the diagnostic time by about one week, enabling more precise treatment for the patient.

Conclusion

Subacute meningoencephalitis can present without fever or meningitis signs, often leading to a diagnostic delay. Parasitic meningoencephalitis such as Angiostrongylus cantonensis should be considered if peripheral eosinophilia. Metagenomic NGS is a novel diagnostic tool that can aid in pathogen identification and improve patient prognosis.

病例報告 113_C53

神經內分泌腫瘤釋放 ACTH 引起的異位性庫欣氏症：一例罕見的病例報告

A case report of ectopic Cushing's syndrome caused by an ACTH-secreting neuroendocrine neoplasm of unknown origin

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Introduction

Ectopic Cushing's syndrome (ECS) is a rare cause of endogenous Adrenocorticotropic hormone (ACTH)-dependent Cushing's syndrome. Tumor detection remains a challenge. We present a rare case of ectopic Cushing's syndrome caused by an ACTH-secreting neuroendocrine neoplasm.

Case Report

We reported a 44-year-old man with clinical features of Cushing's syndrome including leg weakness, bilateral leg edema, round face, purple striae and buffalo hump. His 24-hour urine free cortisol amount was 484.4 μg and morning cortisol level was not suppressed (19.8 μg/dL) under 1 mg overnight dexamethasone suppression test. A pituitary magnetic resonance imaging showed a suspicious lesion. However, the inferior petrosal sinus sampling results showed no evidence of an ACTH-secreting pituitary adenoma. No tumoral lesion was detected on a whole-body computed tomography (CT). On the ^{68}Ga-SSTR PET/CT and ^{18}FDG-PET/CT, a left mediastinal nodule showed excessive somatostatin receptor expression and moderate hypermetabolism. The mediastinal nodule and thymus were excised, and the histopathological findings indicated an ACTH-secreting neuroendocrine neoplasm of anterior mediastinal lymph node, grade 1. The possible origin was still unclear. The patient then received steroid supplement for adrenal insufficiency and follow up image showed no signs of recurrence.

Discussion

ECS is rare and occurs in 3.2~6% of neuroendocrine neoplasms. The primary tumor is most often intrathoracic, but 20% are of unknown origin. With the superiority of ^{68}Ga-SSTR PET/CT and 18FDG-PET/CT over octreoscan, detection of the primary tumor has improved.

Conclusion

This is a challenging case of diagnosing ECS and detecting its primary tumor. This report highlights the use of ^{68}Ga-SSTR PET/CT and ^{18}FDG-PET/CT to detect ACTH-secreting neoplasms.

病例報告 113_C54

以藥物治療併標準化療控制小細胞肺癌併異位性 ACTH 分泌症候群

Paraneoplastic Cushing syndrome with ectopic ACTH of small cell lung cancer successfully controlled with medical management and standard cancer treatment: a case report

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Introduction

Paraneoplastic Cushing syndrome with ectopic ACTH is a rare presentation of small cell lung cancer associated with a high mortality rate. Prompt intervention to lower cortisol levels should be administered to reduce the risk of complications, including infection, hypokalemia, and hyperglycemia. We present a case of a patient managed with chemotherapy and pharmacologic treatment for hypercortisolism.

Case Report

This is a 51-year-old man with hypertension, hyperlipidemia, and type 2 diabetes mellitus. One month before admission, he developed general malaise, occasional chest pain, poorly controlled hypertension, and unintentional weight loss over three weeks. He visited a local hospital ten days prior to admission, where a computed tomography scan of the chest revealed a lung tumor in the left upper lobe. He came to our emergency room one day before admission for a second opinion.
Physical examination revealed obesity, hypertension, and hyperpigmentation over the chest and abdomen. Hypokalemia (2.0 mmol/L) with metabolic alkalosis was noted. Elevated levels of CEA and Chromogranin A were also detected. A bronchoscopy biopsy was performed, and pathology confirmed small cell lung cancer. Further tumor staging indicated cT4N1M0. Given the hypokalemia and uncontrolled hypertension, Cushing syndrome was suspected. Elevated ACTH and cortisol levels were observed. A brain MRI reported no pituitary lesions, leading to a suspicion of paraneoplastic Cushing syndrome with ectopic ACTH. Staining of the previous pathology later confirmed positivity for ACTH, thereby confirming the diagnosis.
Systemic chemotherapy with cisplatin and etoposide, plus concurrent radiotherapy, was initiated. Various cortisol-lowering agents were administered, including fluconazole, etomidate, and metyrapone, with varying treatment responses. However, one episode of diabetic ketoacidosis and septic shock developed during treatment but soon resolved. Five cycles of chemotherapy and a full course of radiotherapy were completed over the following 6 months. A subsequent computed tomography scan showed a significant decrease in the size of the chest tumor. However, cortisol and ACTH levels remained only partially controlled under metyrapone.

Discussion

Paraneoplastic Cushing syndrome with ectopic ACTH is a rare manifestation of small cell lung cancer. A study using the Surveillance, Epidemiologic, and End Results (SEER) database reported that the median survival of patients with ectopic ACTH syndrome in SCLC was much shorter compared to those without paraneoplastic syndrome [1]. Another study that included five patients with ectopic ACTH in SCLC treated at a single medical center showed similar manifestations and

prognosis [2]. They used various cortisol-lowering agents in addition to anticancer treatment, but with partial or no treatment responses. Only one patient was successfully managed for both hypercortisolism and cancer, and this patient had a significantly longer survival. This aligns with our experience that managing both cancer and hypercortisolism leads to better outcomes.

Conclusion

Ectopic ACTH syndrome in small cell lung cancer is rare and associated with a poor prognosis. In addition to anticancer treatments, cortisol-lowering treatment is essential.

病例報告 113_C55

反彈性酮酸中毒發生於使用超長效胰島素的糖尿病酮酸中毒患者：病例報告

Rebound ketoacidosis in a patient with diabetic ketoacidosis using ultra-long-acting insulin: a case report

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Introduction

Diabetic ketoacidosis (DKA) is a life-threatening condition necessitating careful management, particularly in the transition from intravenous (IV) insulin to subcutaneous (SC) insulin to prevent complications. The American Diabetes Association (ADA) in 2009 recommended a 1-2 hour overlap between IV insulin discontinuation and SC insulin initiation. However, with the advent of ultra-long-acting insulins, this overlap may be insufficient. Recent guideline suggested starting SC basal insulin within 60 minutes of IV insulin initiation, though specific overlap durations are not well-defined.

Case Report

A 24-year-old woman with a history of type 1 diabetes mellitus presented with DKA, triggered by missed insulin doses and SARS-CoV-2 infection. She was managed with IV insulin infusion, fluids and electrolytes replacement. After 30 hours of IV insulin, ketone clearance achieved, SC insulin glargine U-300 (IGlar-300) was administered with a 6-hour overlap with IV insulin. Despite the extended overlap hours, the patient developed rebound ketoacidosis 7 hours after IV insulin discontinuation, necessitating the resumption of IV insulin and additional doses of IGlar-300. Ketone clearance was eventually achieved 48 hours after restarting IV insulin, with stabilization of blood glucose levels and resolution of metabolic acidosis.

Discussion

This case report describes the transition strategy from IV insulin infusion to SC ultra-long-acting insulin in the management of DKA. The patient experienced rebound ketoacidosis despite a prolonged overlap period. When using ultra-long-acting insulin as the transition strategy, a longer overlap may be necessary to achieve stable therapeutic levels in the bloodstream to avoid rebound ketoacidosis. The updated consensus report in 2024 till suggested an overlap of 1-2 h between administration of SC insulin and the discontinuation of IV insulin; but, this consensus report did not include the transition to ultra-long-acting insulin (e.g., degludec, glargine U300). Recent studies suggest that early administration of basal insulin, particularly long-acting types, can improve outcomes by speeding DKA resolution and reducing hyperglycemic rebound. In 2023, Joint British Diabetes Societies (JBDS) recommended that the usual basal insulin should be restarted within 60 minutes of commencing IV insulin infusion in DKA management. However, guidelines are less clear on the optimal management for newer, ultra-long-acting insulins, leaving clinicians to navigate this transition on a case-by-case basis.

Conclusion

Transitioning from IV insulin to SC ultra-long-acting insulin in DKA management may require an extended overlap period to prevent rebound hyperglycemia and ketoacidosis. Early administration of long-acting basal insulin can result in a faster DKA resolution and less hyperglycemic rebound. The required duration of overlap remains uncertain. This case illustrates the importance of tailoring insulin transition strategies to the specific pharmacokinetics of the insulin used and the patient's clinical status. The findings emphasize the need for further research to establish evidence-based guidelines for the use of ultra-long-acting insulins in the management of DKA.

病例報告 113_C56

以關節炎及卵巢輸卵管膿瘍表現的古德氏症候群--- 個案報告

A case of Good syndrome with presentation of tubal-ovarian abscess and arthritis--a case report

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Introduction

Good syndrome (GS) is a rare immunodeficiency found in 6-10% of patients with thymoma, a slow-growing tumor of the thymus with an incidence of 0.05 per 100,000 person-years. In about half of the cases, thymoma diagnosis precedes the onset of immunodeficiency, although the time interval between the two varies. The pathogenesis of GS is unclear, but it is thought to result from disordered thymic epithelium leading to defective thymic selection. GS differs from other immune deficiencies, such as common variable immune deficiency (CVID) and X-linked agammaglobulinemia (XLA), due to its late onset, lack of familial occurrence, and absence of lymphoid hyperplasia.

Case Report

This case report describes a 50-year-old woman who presented with a tubo-ovarian abscess and later developed arthritis. Initially, she experienced fever, chills, and lower abdominal pain, which was diagnosed as sepsis caused by a ruptured tubo-ovarian abscess. She underwent surgery, and a chest X-ray incidentally revealed a mediastinal mass, later identified as a type AB thymoma. After resection, the patient had an uneventful recovery.

Seven months after surgery, the patient developed arthritis in her left foot without other associated symptoms. Laboratory tests revealed hypogammaglobulinemia and undetectable CD19 B-cells, with reduced CD4 T-cell count and a low CD4/CD8 ratio. This confirmed the diagnosis of Good syndrome. The patient was treated with intravenous immunoglobulin (IVIG), which improved her arthritis, and follow-up CT scans showed stable thymoma.

Discussion

Inflammatory arthritis is more common among patients with T-cell immunodeficiencies, though the mechanisms remain unclear. Dysregulated T-cell function is believed to play a crucial role in the development of joint inflammation. Joint issues are also seen in humoral primary immunodeficiency disorders (PIDs), such as agammaglobulinemia, CVID, and hyper-IgM syndromes, though they are rare in GS. Arthritis can sometimes be an initial symptom of these immunodeficiencies and may even reveal an underlying PID.

Studies have shown that rheumatologic disorders are present in 10.1% of CVID patients, with juvenile idiopathic arthritis (JIA), rheumatoid arthritis (RA), and juvenile spondyloarthritis (JSpA) being the most common. While joint involvement in GS is rare, it is important for clinicians to be aware that joint symptoms in patients could signal an underlying immunodeficiency.

Conclusion

joint manifestations are uncommon in Good syndrome but are frequently observed in other T-cell

immunodeficiencies. The role of T-cell dysfunction in causing arthritis is an important area for further research. Early diagnosis and management of joint complications in GS are crucial to improve patient outcomes. Further studies are needed to understand the pathogenesis and treatment of arthritis in Good syndrome.

病例報告 113_C57

縱膈囊腫併發大量心包膜積血 - 個案報告

Mediastinal cyst complicated with massive hemopericardium - A Case Report

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Introduction

Mediastinal cysts can range from asymptomatic cases to life-threatening complications. We report a case of a 47-year-old woman with a large mediastinal cyst, massive hemopericardium, and pleural effusion.

Case Report

A 47-year-old Taiwanese woman, previously healthy, presented with 2-3 weeks of shortness of breath, palpitations, and a nighttime cough that worsened with exertion and lying down. She denied chest pain, fever, weight loss, night sweats, medication, tobacco use, or drug allergies.
Chest radiography showed a mass in the anterior mediastinum with left-sided pleural effusion, and the patient was admitted for further investigation.
A grade III/VI systolic murmur was heard at the left lower sternal border. Initial labs, including beta-HCG, alpha-fetoprotein, and squamous cell carcinoma antigen, were unremarkable. Echocardiography revealed a large pericardial cyst (10 x 10 cm), pericardial effusion, moderate tricuspid regurgitation, and moderate pulmonary hypertension (54 mmHg). Chest computed tomography showed a large, well-circumscribed cystic mediastinal mass within the pericardium, approximately 12 cm, heterogeneous with coarse calcifications, and left pleural effusion.
The patient underwent thoracoscopic resection. During the operation, the mass was found to be densely adherent to the left lung, necessitating a wedge resection. After drainage, the anterolateral wall of the large cyst was resected using Ligasure. Analysis of the cyst's contents revealed it was rich in RBCs. Pathological examination reported atrophic thymic tissue and a fibrotic-walled cyst adhered to the lung tissue, without an epithelial lining. The final diagnosis was a hemorrhagic pericardial cyst. A follow-up echocardiogram revealed relief of tricuspid regurgitation and pulmonary hypertension.

Discussion

Pericardial cysts account for approximately one-third of mediastinal cysts, with an incidence of about 1 in 100,000 people. Most cases of pericardial cysts are asymptomatic and are commonly discovered incidentally in patients in their 40s or 50s. Common symptoms, when present, include pleuritic chest pain, dyspnea, palpitations, cough, dysphagia, fever, weight loss, and paroxysmal tachycardia. Dizziness and syncope have also been reported. [3,4]
Compression of nearby structures can lead to complications such as right ventricular outflow tract obstruction, superior vena cava syndrome, or obstruction of the right mainstem bronchus.[5] Rarely, pericardial cysts can be associated with serious complications such as heart failure, cardiac tamponade, hemorrhage, rupture, constrictive pericarditis, and, in some cases, sudden cardiac death. [6,7]

For symptomatic pericardial cysts, management options include minimally invasive procedures such as percutaneous aspiration (recommended by the European Society of Cardiology) or ablation/ethanol sclerosis. There is a significant likelihood of recurrence after aspiration, with the recurrence rate of pericardial cysts being approximately 30%. Other treatment options include resection via thoracotomy, VATS, or mediastinoscopy. [1]
Our patient had symptoms and elevated systolic pulmonary artery pressure, likely due to compression. After thoracotomy and cyst drainage, her symptoms improved. The cyst's location suggests it was originally a pericardial cyst, with its epithelial lining destroyed by inflammation. Regular follow-up is needed due to recurrence risk.

Conclusion

Mediastinal cysts are often asymptomatic but can cause serious complications. Early detection and treatment are essential to prevent adverse outcomes.

病例報告 113_C58

即刻診斷與補體抑制劑治療：在懷孕誘發的非典型溶血性尿毒症候群併多重器官衰竭中的救援策略

Rapid Diagnosis and Eculizumab: A Lifesaving Strategy in Pregnancy-Associated aHUS with Multi-Organ Failure

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Introduction

Atypical hemolytic uremic syndrome (aHUS) is a rare, rapidly progressing, and life-threatening thrombotic microangiopathy. Diagnosing aHUS during the puerperium is challenging due to overlapping symptoms with preeclampsia and HELLP syndrome. This case highlights a patient with pregnancy-associated aHUS affecting multiple organs, including the brain, lungs, and kidneys, who rapidly recovered with prompt diagnosis and Eculizumab treatment.

Case Report

A 40-year-old female patient had regular and healthy prenatal check-ups (at 24 weeks of pregnancy: GFR 130 mL/min/1.73 ㎡, Cr 0.52 mg/dL, urine analysis without proteinuria, Hb 11.7 g/dL, platelet count 264 x10^3/μL, and negative for Anti-Cardiolipin IgG and Beta2-glycoprotein IgG). At 34 weeks of pregnancy, she experienced blurred vision and reduced urine output for two weeks, with a blood pressure of 167/106 mmHg. At 36 weeks of pregnancy, she had sudden vaginal bleeding at home and discovered that the fetus had no heartbeat upon arrival at the hospital. After placental removal, her oliguria worsened, and her renal function acutely deteriorated, accompanied by hemolytic anemia and thrombocytopenia. She was transferred to our emergency department with a GFR of 9 mL/min/1.73 ㎡, Cr 5.05 mg/dL, Hb 9.2 g/dL, platelet count 27 x10^3/μL, and LDH 1053 U/L. Despite four plasma exchanges, her condition did not improve, and she even experienced a 108-second-long grand mal seizure, leading to a diagnosis of atypical hemolytic uremic syndrome (aHUS). Brain MRI later confirmed posterior reversible encephalopathy syndrome (PRES). She received four doses of a complement inhibitor, resulting in a rapid improvement in platelet count and renal function. Genetic testing for 25 aHUS-related genes revealed no abnormalities. The patient's condition gradually stabilized, confirming aHUS despite the absence of detectable genetic mutations. The patient was discharged after 10 days, able to return to self-care, underscoring the effectiveness of timely treatment in severe aHUS cases.

Discussion

Diagnosing thrombotic microangiopathy (TMA) in pregnancy is challenging due to its overlapping features with conditions like preeclampsia, HELLP syndrome, acute fatty liver of pregnancy (AFLP), thrombotic thrombocytopenic purpura, atypical hemolytic uremic syndrome (aHUS), disseminated intravascular coagulation, systemic lupus erythematosus, and antiphospholipid syndrome. Notably, after placental removal, conditions such as preeclampsia, HELLP syndrome, and AFLP usually stabilize. However, in aHUS, placental removal can act as a 'second hit,' triggering complement activation and further destabilization of TMA. When this worsening occurs, aHUS

should be promptly considered, with rapid diagnosis and treatment. Despite life-threatening multi-organ failure due to aHUS, the patient recovered and was discharged after 11 days, thanks to timely full-dose Eculizumab under compassionate use. This contrasts with the extended hospital stays, often exceeding 21 days, seen in patients without full-dose and regular frequency of anticomplement treatment.

Conclusion

This case highlights the vital need for rapid diagnosis and timely Eculizumab treatment in pregnancy-associated aHUS, which can swiftly lead to multi-organ failure. Early intervention not only stabilizes the condition but also dramatically shortens recovery time, as seen in this patient's discharge after just 11 days. It underscores the importance of clinical vigilance and timely access to therapy to improve outcomes in severe aHUS cases.

病例報告 113_C59

Pembrolizumab 合併使用 Cetuximab 造成的免疫相關不良事件：重症肌無力之個案報告

Myasthenia Gravis as an Immune-Related Adverse Event Following Pembrolizumab and Cetuximab Treatment for Advanced Gum Cancer: A Case Report

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Introduction

Pembrolizumab, a programmed death receptor-1 inhibitor, has shown promising results in the treatment of advanced gum cancer. However, immune-related adverse events are a significant concern. This report presents a case of a patient with advanced gum cancer who developed myasthenia gravis following treatment with pembrolizumab and cetuximab.

Case Report

The patient, a 67-year-old male, has a medical history of moderately differentiated squamous cell carcinoma located in the left upper gum. He underwent surgical resection on July 13, 2022, which was classified as T4aN2bM0. Postoperatively, the patient was treated with weekly cisplatin from August 15 to September 23, 2022, with total six doses, and radiotherapy from August 1 to September 23, 2022, with a total dose of 5400cGy/27Fx. Despite concurrent chemoradiotherapy, local recurrence was observed in March 2023. Subsequently, the patient was treated with immune checkpoint inhibitor therapy, including pembrolizumab on April 14 and May 5, 2023, and cetuximab on April 19, 2023. In May 2023, the patient reported symptoms of generalized weakness and right eyelid ptosis. Additionally, he suffered from head drooping during prolonged sitting, challenges in rising from a supine position, and an increased propensity for choking. As a result, the patient was admitted for comprehensive evaluation.

Upon admission, no organic brain lesions were identified via brain MRI. Further investigations revealed elevated creatine phosphokinase levels (1615 U/L), anti-Ach receptor levels (4.06 nmol/L), a decremental response in the repetitive stimulation test on the right biceps brachii following repetitive 3 Hz stimulation, and the presence of Titin antibodies. After ruling out autoimmune inflammatory myopathies, a diagnosis of myasthenia gravis was confirmed. Consequently, we initiated therapy with methylprednisolone (500mg per day) in conjunction with pyridostigmine. Despite this, the patient presented persistent generalized weakness and thus we commenced plasma exchange therapy seven times accordingly. Following the completion of plasma exchange, the patient's ptosis, muscle strength and swallowing performance improved subsequently. Thus, we gradually tapered off prednisolone in an outpatient setting and continued with cetuximab as antineoplastic therapy.

Discussion

This case illustrates the emergence of myasthenia gravis (MG) as a potential immunotherapy-related adverse event (irAE) linked to immune checkpoint inhibitors (ICI) therapy. To our understanding, this is the inaugural reported instance of concurrent MG secondary to

antineoplastic therapy with pembrolizumab in conjunction with cetuximab. The temporal correlation between ICI administration and the onset of MG suggests a plausible causal connection. Although MG is a recognized paraneoplastic syndrome associated with various malignancies, its manifestation following ICI therapy is less frequent. This patient exhibited improvement following the discontinuation of pembrolizumab and the initiation of corticosteroids and plasma exchange. Regarding advanced gum cancer, we maintained antineoplastic therapy with cetuximab for disease control.

Conclusion

We report a rare case of immune checkpoint inhibitor-related myasthenia gravis associated with the concurrent use of pembrolizumab and cetuximab. We managed the condition by administering corticosteroids and plasma exchange for immune-related adverse events. While pembrolizumab has revolutionized cancer treatment, clinicians must remain vigilant for the development of irAEs such as MG. Early recognition and appropriate management are crucial to prevent serious complications.

病例報告 113_C60

心包穿刺術併發同時醫源性氣胸、心包積氣、腹腔積氣-案例報告

Pericardiocentesis complicated with concurrent pneumothorax, pneumopericardium and pneumoperitoneum– case report

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Introduction

Pericardiocentesis is a procedure performed to remove pericardial fluid from the pericardial sac. It is often performed in the setting of cardiac tamponade to correct hypotension. Nonetheless, there may be multiple complications including arrhythmia, cardiac perforation, cardiac tamponade and injury to surrounding structures such as diaphragm, liver, lung, peritoneum. We present a rare case with pericardiocentesis related concurrent pneumothorax, pneumopericardium and pneumoperitoneum. Surgery intervention is performed with subsequent resolution.

Case Report

A 71-year-old man had been admitted for severe community acquired pneumonia. However, shock developed and echocardiogram showed cardiac tamponade. Emergent pericardiocentesis via sub-xiphoid anterior approach was performed. The patient had improving hemodynamics after pericardial effusion drainage. Nonetheless, progressive subcutaneous emphysema and abdominal distention occurred 6 hours later. A computed tomographic was obtained and showed pneumoperitoneum, pneumomediastinum, pneumopericardium and subcutaneous emphysema in the chest and abdominal wall. Exploratory laparoscopic surgery then showed iatrogenic diaphragm penetration of the pericardial drainage catheter through peritoneal cavity to pericardium. No hollow organ perforation was observed. We removed drainage catheter and made diaphragm repair by suture. However, left side tension pneumothorax developed after diaphragm repair and chest tube was inserted via the left 4th intercostal space. After further treatment, chest tube was removed without residual pneumomediastinum, pneumopericardium and pneumoperitoneum 3 weeks later. The patient was discharged under ambient air.

Discussion

Pericardiocentesis is an invasive procedure that carries potential risks. Major complications reported in the largest series (0.3–3.9% cases), include death, laceration of cardiac chambers, and coronary arteries or intercostal vessels, puncture of abdominal viscera or peritoneal. Blind pericardiocentesis should be restricted to emergencies and other critical circumstances, where image guidance is not readily available. Echocardiography, which allows real-time monitoring, is now favored for its safety and effectiveness during pericardiocentesis.

In our case, we performed emergent pericardiocentesis due to cardiac tamponade. Nonetheless, puncture through peritoneal cavity and diaphragm causes pneumothorax, pneumoperitoneum and even pneumopericardium.

There is no consensus regarding treatment of pneumopericardium and the clinical course is highly variable. Treatment depends on etiology and clinical severity, which is determined by the amount and speed of the of the accumulation of air in the pericardium. Hypotension, radiographic intrapericardial air, and absent alternative causes of shock constitute the diagnosis of fatal pericardial tamponade due to pneumopericardium. However, usually pneumopericardium heals spontaneously within a few days. If there is no hemodynamic imbalance, clinical monitoring of patients with pneumopericardium waiting for absorption of air is possible. On the contrary, if pericardial tamponade develops, pericardiocentesis or surgically closing the channel between air and pericardium should be performed urgently.

Conclusion

Pericardiocentesis is a crucial procedure for treating symptomatic pericardial effusions, but it carries risks. Concurrent pneumothorax, pneumopericardium and pneumoperitoneum are rare complications and surgery intervention with adequate air drainage, tissue defect repair may lead to improving condition.

For prevention of these complications, blind pericardiocentesis should be limited to emergencies, as image-guided techniques offer greater safety and reduce recurrence rates.

病例報告 113_C61

下壁急性心肌梗塞的夢魘:右冠狀動脈全阻塞併發乳突肌斷裂-案例報告

Nightmare in inferior wall AMI: RCA total occlusion complicated with papillary muscle rupture – case report

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Introduction

Right coronary artery (RCA) occlusion causes acute myocardial infarction (AMI)and papillary muscle rupture (PMR) is one of the catastrophic mechanical complications following MI. Failed percutaneous coronary intervention(PCI) with papillary muscle rupture leads to rapid hemodynamic deterioration and cardiogenic shock. Timely diagnosis and rapid, multidisciplinary management are crucial for improving outcomes. We present a challenging case with inferior wall ST-elevation MI complicated with papillary muscle rupture. PCI failed and mechanical support with emergent surgical rescue were performed.

Case Report

A 45-year-old male with untreated hypertension and obesity presented to the emergency department with intermittent chest pain over two weeks with exacerbation for one day. At our emergency department, there was unstable vital sign with blood pressure: 91/60mmHg, heart rate: 110/min, respiratory rate: 30/min wit SpO2:88% under ambient air. Electrocardiogram showed lead II, III, aVF, V4R ST elevation. Primary PCI was performed after intubation and dual antiplatelet therapy initially.

Nonetheless, there was occlusion since proximal right coronary artery. We could not establish coronary flow due to severe calcification and failed wiring to true lumen. The patient deteriorated with unstable hemodynamics and oxygenation during the procedure. Veno-arterial extracorporeal membrane oxygenation (VA-ECMO) and intra-aortic balloon pump (IABP) were applied. Subsequent transesophageal echocardiography (TEE) confirmed posteromedial PMR with severe mitral regurgitation(MR). Emergency mitral valve replacement (MVR) and coronary artery bypass grafting (CABG) with great saphenous vein to PDA were performed. After surgery, the patient had stable hemodynamics but developed severe ischemic stroke and following septic shock. Due to unconsciousness and poor neurologic outcome, the family withdrew life-sustaining treatment. The patient expired following these events.

Discussion

PMR due to ischemia is a severe complication of AMI. The posteromedial papillary muscle with a single supply from the PDA is more frequently affected, especially in inferior or lateral STEMI. Our patient has inferior STEMI and RCA occlusion was noted. Nonetheless, we could not restore coronary artery flow and the patient had deteriorated condition. Mechanical support was used and further TEE showed posteromedial PMR with severe MR. Emergent surgery is indicated for hemodynamically unstable patient with AMI and failed PCI. Besides, mitral valve repair is primarily indicated for severe MR due to papillary muscle rupture, especially in hemodynamically unstable

patients or those in cardiogenic shock.

Conclusion

Timely diagnosis of PMR is crucial for AMI patients. Mechanical support and emergent surgery play an important role for failed PCI and acute MR. Nonetheless, there is still high complication and mortality rate.

病例報告 113_C62

一例血管免疫細胞 T 細胞淋巴瘤患者 COVID-19 感染後自發緩解的病例報告

A Case Report of Spontaneous Remission in a Patient with Angioimmunoblastic T-Cell Lymphoma Following COVID-19 Infection

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Introduction

Angioimmunoblastic T-cell lymphoma (AITL) is a rare and aggressive subtype of peripheral T-cell lymphoma characterized by systemic symptoms, generalized lymphadenopathy, and a poor prognosis. Treatment typically involves aggressive chemotherapy, but spontaneous remission in AITL is extremely rare. The COVID-19 pandemic has introduced novel medical scenarios, including unexpected disease remissions. This article discusses a remarkable case of spontaneous AITL remission following a COVID-19 infection, exploring potential mechanisms and future research implications.

Case Report

A 59-year-old woman presented with nodular enlargements in the left inguinal and left cervical lymph nodes. Biopsy confirmed the diagnosis of Angioimmunoblastic T-cell lymphoma (AITL). The patient underwent six cycles of CHOP chemotherapy (Cyclophosphamide, Doxorubicin, Vincristine, and Prednisone) and achieved complete remission. Following initial remission, the patient received conditioning chemotherapy with the BEAM regimen (Carmustine, Etoposide, Cytarabine, and Melphalan) and underwent autologous peripheral blood stem cell transplantation as consolidation therapy. After five years, palpable lymph nodes in the left supraclavicular fossa were noted by the patient. PET scan revealed recurrent lymphoma in the bilateral lower neck, left axillary, and left iliac regions. The patient received Pralatrexate for recurrent peripheral T-cell lymphoma (PTCL) for 18 cycles, achieving partial remission (PR).

During follow up, the patient was diagnosed with COVID-19 pneumonia and was treated with Molnupiravir. Neither chemotherapy nor target was administered during this period. Surprisingly, complete remission was found in following PET scan.

Unfortunately, about 1 year later, PET scan indicated FDG-avid malignancy in multiple supra- and infra-diaphragmatic lymph nodes, spleen, multiple bones, both lungs, and liver. The patient was admitted for salvage chemotherapy. Finally, she succumbed to acute liver failure and infection during her hospital stay.

Discussion

Viral infections can significantly disrupt the immune systems of cancer patients in ways that are not fully understood. Some cases of tumor regression during the pandemic have been reported. These cases involve various factors, including different types and stages of cancer, different COVID-19 treatments, and comorbidities, with limited long-term follow-up data available.

Several hypotheses could explain this occurrence: 1.Immune Activation: COVID-19 infection triggers a strong immune response, activating cytotoxic T-cells and natural killer (NK) cells, which

might target and destroy lymphoma cells. 2.Cytokine Storm: The severe cytokine storm associated with COVID-19 may inadvertently target lymphoma cells. High levels of cytokines like IL-6, IL-10, and TNF-alpha might create an unfavorable environment for lymphoma cell survival. 3.Corticosteroids: Used to manage inflammation in COVID-19, corticosteroids may also possess anti-lymphoma properties, contributing to remission.

Patients with recurrent or refractory peripheral T-cell lymphomas treated with traditional Pralatrexate have only 1.8 months median PFS. In our COVID-19 infected patient, the PFS was 12 months. It appears that COVID-19 has altered the immune response, leading to a later recurrence in our case. Nevertheless, further monitoring for recurrence is still necessary and consolidation therapy is warranted.

Conclusion

This report shows that COVID-19 infection could induce regression of lymphoma. Further consolidation therapy is warranted to prevent recurrence.

病例報告 113_C63

肝細胞癌患者接受 Atezolizumab 和 Bevacizumab 後造成腫瘤溶解症候群的案例分析

Tumor Lysis Syndrome After Atezolizumab and Bevacizumab in Hepatocellular Carcinoma

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Introduction

Hepatocellular carcinoma (HCC) is the most common primary liver cancer. According to the updated Barcelona Clinic Liver Cancer (BCLC) guidelines, the atezolizumab and bevacizumab (Atezo-Bev) combination become the first-line systemic therapy, with better survival over Sorafenib [1]. Common adverse events of the Atezo-Bev regimen have been described, but tumor lysis syndrome (TLS) has rarely been reported with this treatment. Here, we present a case of TLS after Atezo-Bev treatment.

Case Report

A 53-year-old man with HIV (under highly active antiretroviral therapy) and hepatitis B virus-related liver cirrhosis (Child-Pugh score 6) was diagnosed with advanced HCC, staged as cT4N1M0, BCLC stage C, with a 15.3 cm tumor and invasion into the portal vein and inferior vena cava.

Three weeks after receiving the first dose of the Atezo-Bev regimen, a marked decrease in α-fetoprotein (AFP) levels (from >303,000 ng/mL to 1,650.43 ng/mL) was noted. However, the patient started to develop symptoms of general malaise and poor appetite, with laboratory data revealing significant metabolic disturbances, including hyperkalemia (7.1 mmol), hyperphosphatemia (7.3 mg/dL), hyperuricemia (13.9 mg/dL), and acute kidney injury (AKI), as evidenced by a rise in creatinine from 1.05 mg/dL to 4.00 mg/dL over three weeks. The above findings led to a diagnosis of tumor lysis syndrome (TLS) complicated by AKI.

He was promptly treated with Rasburicase for hyperuricemia, aggressive hydration for AKI, and management for hyperkalemia. While these measures improved hyperuricemia and renal function, the patient later developed signs of liver decompensation, including progressive jaundice and ascites. Owing to this condition, the planned Atezo-Bev regimen was discontinued. Despite medical interventions, the patient's condition worsened, and he passed away a few days later under hospice care.

Discussion

According to the 2022 BCLC update, the Atezo-Bev regimen is now the first-line systemic therapy for patients with advanced HCC, as it provides better overall and progression-free survival compared to sorafenib in the IMbrave150 trial [1,2]. The adverse events of the Atezo-Bev regimen included proteinuria, hypertension, elevated AST levels, fatigue, and pruritus [3].

Tumor lysis syndrome (TLS) is a severe oncologic emergency caused by extensive tumor cell breakdown, leading to metabolic abnormalities (hyperkalemia, hyperphosphatemia, hypocalcemia) and AKI. Although TLS is mainly linked to rapidly proliferating hematologic malignancies, it can also occur in patients with solid tumors, including HCC, spontaneously or after treatment [4]. A large tumor burden was reported as a known risk factor for TLS in HCCs [5,6]. Our

case, with a tumor measuring 15.3 cm, supports this, suggesting the substantial tumor burden likely contributed to TLS.

Despite the increasing use of immune checkpoint inhibitors (ICIs), TLS is not mentioned in the National Comprehensive Cancer Network (NCCN) guidelines or drug inserts. A real-world pharmacovigilance study revealed that ICIs, including atezolizumab, can induce TLS in solid and hematologic malignancies [7]. Two case reports described TLS in metastatic urothelial carcinoma after a single dose of Atezolizumab [8,9]. Another study reported TLS in metastatic breast cancer after Atezolizumab and Nab-Paclitaxel, attributing it solely to immunotherapy since the patient had not experienced TLS with prior chemotherapy [10]. To our knowledge, TLS has not been reported in HCC patients treated with the Atezo-Bev regimen.

Conclusion

We presented a rare case of a patient with HCC and a large tumor burden who developed TLS after receiving the Atezo-Bev regimen. Physicians should consider TLS as a possible differential diagnosis in patients receiving the regimen, as TLS is a life-threatening condition that may require urgent management.

病例報告 113_C64

子宮內膜癌導致之難治性危及生命的高血鈣

A Patient of Endometrial Carcinoma Associated with Refractory Life-Threatening Hypercalcemia

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Introduction

Hypercalcemia occurs in 20-30% of cancer patients during the disease course[1], with the most common manifestations being nausea, vomiting, renal impairment, and even coma. Gynecologic neoplasms were responsible for approximately 20% of malignancy-associated hypercalcemia[2]. Yet, the ovarian tumors account for the majority of gynecologic neoplasms associated with hypercalcemia[3]. Here, we present a patient of endometrial carcinoma (EC) with hypercalcemia, caused by ectopic parathyroid hormone (PTH) secretion. To our knowledge, only one patient of ectopic PTH and hypercalcemia caused by endometrial carcinoma has been reported previously[4].

Case Report

A 53-year-old woman with past medical history of leiomyoma uteri came to the emergency department due to abdominal pain and general discomfort. Laboratory data showed severe hypercalcemia (calcium = 13.4 mg/dL), acute pancreatitis (elevated lipase level: 429 U/L) and acute kidney injury (creatinine = 4.49 mg/dL, blood urea nitrogen = 89 mg/dL). The hypercalcemia surveys showed elevated iPTH levels up to 462 pg/mL. Thyroid sonography and Tc-99m-MIBI parathyroid scan did not reveal any abnormalities. Whole body Computed Tomography (CT) showed an enlarged uterus (Figure 1) and nodules in the paraaortic and paracaval regions. Diagnostic fractional dilatation and curettage (D&C) was performed, and the pathology reported adenocarcinoma. Endometrial cancer and ectopic PTH secretion from endometrial cancer were confirmed.

She received a series of chemotherapy and antihypercalcemic therapy, including cinacalcet and zoledronate. However, the disease was refractory to multiple lines of chemotherapy, including paclitaxel plus carboplatin. cisplatin plus 5-FU and concurrent chemoradiotherapy with carboplatin (Figure 4). Ultimately, she died on multiple organs failure caused by tumor progression with hypercalcemia.

Discussion

Most patients with endometrial cancer initially present with abnormal uterine bleeding. In this patient, however, hypercalcemia complicated with pancreatitis and abdomen distress was discovered.

Malignancy-associated hypercalcemia can be divided into two major subgroups: local osteolytic hypercalcemia (LOH) and humoral hypercalcemia of malignancy (HHM). Less commonly, ectopic production of PTH or 1–25-dihydroxy (OH)2 vitamin D3 secretion also demonstrate the result. Elevation of serum parathyroid hormone-related protein (PTHrP) is often detected in most cancer patients of humoral hypercalcemia.

Despite the complex mechanisms, volume expansion with isotonic saline is the initial treatment of

choice. Bisphosphonates are also commonly used in patients with hypercalcemia. On the other hand, the primary goal is to treat the underlying malignancy. Another case report[5] described a patient with hypercalcemic endometrial cancer requiring renal support therapy. Both calcium and PTH levels dropped immediately to normal range after receiving cytoreductive surgery. Our patient underwent several kinds of antihypercalcemic therapy, which included massive hydration, loop diuretic use, the new-generation bisphosphonates of Zoledronic acid, and a combination of antineoplastic chemotherapy/ CCRT and endocrine therapy; however, the hypercalcemia relapsed because of refractory cancer.

Conclusion

We reported a rare case of endometrial cancer with hypercalcemia. The disease was refractory to most treatments, leading to a dismal prognosis.

病例報告 113_C65

腸道假性阻塞之少見原因：胸腺瘤與乙醯膽鹼受體抗體引發腸道假性阻塞之病例報告與臨床啟示

Thymoma-Associated Acetylcholine Receptor Antibodies Causing Intestinal Pseudo-Obstruction in the Absence of Myasthenia Gravis: A Case Report and Clinical Insights

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Introduction

Thymoma is often associated with paraneoplastic syndromes, particularly myasthenia gravis (MG), where acetylcholine receptor (AChR) antibodies disrupt neuromuscular transmission. Although MG primarily affects skeletal muscles, there is evidence that AChR antibodies can influence other systems, including the gastrointestinal (GI) tract. This case discusses the relationship between thymoma, AChR antibodies, and ileus in a patient without typical MG symptoms.

Case Report

A 35-year-old man with no significant medical history presented with a one-day history of abdominal pain, accompanied by abdominal fullness, absence of bowel movements, and poor appetite. Four months earlier, he had experienced intermittent fever lasting over a week, for which he was treated for an intra-abdominal infection. He denied chest pain, dyspnea, diarrhea, or urinary symptoms.

At a local hospital, abdominal CT revealed left pleural nodular thickening, ascites, and dilated bowel loops without obstructive factor, prompting his transfer for further evaluation. On admission, chest X-ray and abdominal film revealed an oval mediastinal mass and signs of ileus. Despite prokinetic therapy, the intestinal obstruction persisted. Colonoscopy revealed no points of mechanical obstruction. Following up laboratory finding of an elevated acetylcholine receptor antibody level of 0.93 nmol/L, raised the concern for thymoma-associated paraneoplastic syndromes, though the patient had no symptoms suggestive of myasthenia gravis.

A chest CT scan identified a 9 cm mass in the left anterior mediastinum, suspicious for invasive thymoma or thymic carcinoma, along with pleural metastases and a left upper lung nodule. A lung biopsy revealed lymphocytic infiltration without malignancy. Video-assisted thoracoscopic surgery (VATS) was performed, and pathology confirmed Type B2 thymoma with direct invasion of the mediastinal pleura. Immunohistochemical staining showed CD5, CK (AE1/AE3), p63, and TdT positivity, consistent with thymoma. The tumor was staged as pT1bNx, Masaoka Stage IVA.

The patient was started on chemotherapy with cisplatin, doxorubicin, and cyclophosphamide. He was discharged in stable condition. There were no clinical signs of thymoma-associated myasthenia gravis, despite the elevated acetylcholine receptor antibody level. Further chemotherapy and monitoring were planned.

Discussion

Thymoma is commonly associated with paraneoplastic syndromes such as myasthenia gravis (MG), where acetylcholine receptor (AChR) antibodies play a central role. These antibodies interfere with neuromuscular transmission, typically causing muscle weakness, but can also lead

to gastrointestinal dysfunction, including intestinal pseudo-obastruction, though this is rare. In our case, the presence of AChR antibodies with ileus suggests a multifactorial process.
Acetylcholine is crucial for smooth muscle contraction in the gastrointestinal (GI) tract, mediated through muscarinic receptors. Disruption of acetylcholine signaling due to AChR antibodies may impair GI motility, contributing to intestinal pseudo-obastruction.(1)Thymoma is also associated with autoimmune dysfunction, which can affect autonomic nerves and also extrinsic enteric nervous system, potentially leading to bowel dysmotility(4). Although rare, metastases from thymoma to the GI tract have been reported, which could cause intestinal pseudo-obastruction (2).
In addition, immunosuppressive therapies and acetylcholinesterase inhibitors used to manage MG could also contribute to gastrointestinal issues. Overuse of these medications may lead to cholinergic crisis, which can paradoxically cause autonomic dysfunction, including intestinal pseudo-obastruction.(3)
Overall, intestinal pseudo-obastruction in patients with thymoma and positive AChR antibodies is likely multifactorial, involving both immune dysregulation and potential direct effects on gastrointestinal motility. Close monitoring and a multidisciplinary approach are essential for managing such complex presentations.

Conclusion

Thymoma-associated AChR antibodies may lead to intestinal pseudo-obastruction through impaired acetylcholine signaling in the GI tract. Clinicians should remain alert to such atypical manifestations in thymoma patients to ensure timely diagnosis and treatment.

病例報告 113_C66

偉大的模仿者-令人頭痛的紅斑性狼瘡

The Great Imitator: Lupus Erythematosus, A Headache for Doctors and the Patient

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Introduction

Systemic lupus erythematosus (SLE) is a multi-organ autoimmune disease, with neuropsychiatric SLE (NPSLE) being a severe complication, presenting with headaches, cognitive dysfunction, confusion, psychosis, and seizures. NPSLE can appear early in the disease. However, given the prevalence of stress-related headaches today, diagnosing lupus when only neurological symptoms are present can be challenging. Here, we present a case where a headache was the initial sign of SLE.

Case Report

This 33-year-old female hairdresser had no known systemic diseases or family history of autoimmune disorders. She experienced six months of headaches and increasing agitation, which her family physician, coworkers, family, and fiancé attributed to work stress and her upcoming wedding. Despite advice to relax, she struggled with irritability, and her family suggested psychiatric consultation for pre-wedding anxiety.

A week before visiting our clinic, she developed upper respiratory symptoms and mild leg edema. Urinalysis showed microhematuria and mild proteinuria (UPCR: 199.52 mg/g), with chest tightness. COVID-19 PCR and influenza tests were negative, and her EKG and troponin I levels were normal. Symptomatic treatment and further tests were arranged. Two weeks later, her symptoms worsened, including headaches, foamy urine, eyelid swelling, dyspnea, fever, and leg edema. Labs revealed prolonged aPTT, positive antibodies (anti-ribosomal P, anti-RNP, anti-SM, APS), elevated opening pressure, and worsening proteinuria (UPCR: 17,311 mg/g) with hypoalbuminemia (2.2 g/dL). Following intravenous steroids, her headaches and irritability improved, allowing a kidney biopsy that confirmed mesangial lupus glomerulonephritis. After continued treatment, her emotional stability returned, and she was ready for her wedding, with proteinuria reduced to 99.17 mg/g at discharge.

Discussion

The patient was newly diagnosed with SLE but only sought nephrology care after kidney involvement and significant proteinuria. In just two weeks, her proteinuria worsened from 199.52 mg/g to 17,311 mg/g. Treatment alleviated the six-month-long headaches and emotional instability. Fortunately, the NPSLE had not progressed to severe symptoms like seizures. NPSLE often appears at or shortly after SLE diagnosis, typically within the first year. While its pathogenesis remains unclear, it is thought to involve inflammatory and vasculitic mechanisms. In this case, the patient's headaches, a focal NPSLE symptom, were initially overlooked due to modern life stressors, delaying proper diagnosis.

Conclusion

This case highlights the diagnostic challenges of NPSLE when neurological symptoms, such as headaches, precede other systemic manifestations. In this patient, six months of persistent headaches and emotional instability were initially attributed to stress, delaying the recognition of SLE. Only after severe kidney involvement with nephrotic syndrome was the underlying SLE identified and treated. This emphasizes the importance of considering autoimmune etiologies, like NPSLE, even when common symptoms such as headaches may be easily dismissed due to lifestyle factors. Early recognition and treatment can prevent more severe complications and improve patient outcomes.

病例報告 113_C67

一位肺癌患者使用化療藥物導致的皮膚病變

Gemcitabine-associated Chronic Lipodermatosclerosis

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Introduction

Gemcitabine, a frequently utilized cytotoxic antineoplastic agent, is often prescribed for solid tumors and hematologic malignancies. However, it is associated with numerous side effects, including a decrease in leukocyte and platelet counts, as well as anemia. Gastrointestinal discomfort, characterized by nausea, vomiting, and diarrhea, is reported in approximately 30%-60% of patients. Skin rash is another common side effect, occurring in about 30% of cases. {1} There are only a few documented cases of skin reactions following Gemcitabine treatment. Among cytotoxic agents, Gemcitabine was the most frequently implicated, accounting for 71% of the cases.{3} Our case involves a unique lipodermatosclerosis-like eruption in a lung cancer patient who has undergone multiple rounds of systemic Gemcitabine therapy.

Case Report

This is a case of a 38-year-old male smoker with a history of stage 4 lung adenocarcinoma, LUL, pT1aNxM1c (AJCC 8th edition), with brain and T8 bone metastasis. His lung cancer treatment included the following cytotoxic chemotherapy, including Pemetrexed, Bevacizumab, Docetaxel, Pembrolizumab and Carboplatin. However, his lung cancer progressed, and he was treated with Gemcitabine with Ramucirumab for his progressive lung cancer.

The patient has been experiencing progressive bilateral lower limb erythema, heat, and swelling without fever or pain for two months. Physical examination found red tender plaques with slight indurated sensation of bilateral shin and dorsal feet. Small vesicle-like lesions were found on the bilateral pretibial part. Dermatologic examination revealed erythema on the bilateral lower limbs. Differential diagnoses included cellulitis, panniculitis, drug hypersensitivity, and deep vein thrombosis.{2} Before the pathology report, we prescribed Oxacillin and Clindamycin for empirical cellulitis treatment. An incisional skin biopsy was performed, revealing lipodermatosclerosis. Chronic inflammatory in subcutis, focal lobular and septal involvement of the subcutaneous tissue with fat necrosis and subcutaneous fibrosis were found. No PAS stain was positive for any microorganism. After the confirmed diagnosis, we paused antibiotic, and enhanced leg elevation. The feet circumference were decreased. Topical steroid was also prescibed.

Based on the overall clinical findings, it was concluded that the skin lesion was likely associated with Gemcitabine.

Discussion

The mechanism of gemcitabine-associated lipodermatosclerosis is still unclear. Venous endothelial damage and cytokine release are likely to develop the reaction.{3} Lipodermatosclerosis is diagnosed clinically, and skin biopsy is not the routine examination. There are acute and chronic stages of the pathogenesis. Initially, increased vascular permeability

presents peripheral edema. Lately, hyperpigmentation with sclerotic dermis and subcutis is noted.

Conclusion

Gemcitabine-associated lipodermatosclerosis like eruption should be a more specific diagnosis. We could differentiate by location and distribution. Infectious cellulitis is often unilateral and warmth. But chemotherapy related pseudocelluitis usually occurs in bilateral lower extremities. We treated the patient with withdrawal Gemcitabine and antibiotics usage and topical steroids. Two weeks later, his skin lesion improved. This case report highlight the importance of recognizing and managing non-infectious dermatologic complications in cancer therapy.

病例報告 113_C68

臨床試驗的孤兒- 腎移植患者出現 IgA 腎病變的治療困境

Orphans of Clinical Trials: The Therapeutic Dilemma in Treating Post-Transplant IgA Nephropathy Patients

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Introduction

Immunoglobulin A nephropathy (IgAN) is a leading cause of primary glomerular disease. The 2024 KDIGO (Kidney Disease: Improving Global Outcomes)guidelines recommend enrolling IgAN patients with proteinuria >1g/day and GFR ≥30, controlled BP for three months, and on maximally tolerated ACEI/ARB in clinical trials. These trials focus on BAFF/APRIL inhibitors and complement blockade. We report a case of ADPKD-related ESRD post-cadaveric kidney transplant, who developed new-onset proteinuria and was diagnosed with IgAN.

Case Report

A 39-year-old man with a family history of autosomal dominant polycystic kidney disease (ADPKD) began peritoneal dialysis at 18. His family members developed end-stage renal disease (ESRD) and started dialysis between 30 and 50 years old. Before reaching ESRD, he reported foamy urine, but a kidney biopsy was not performed due to his ADPKD. At 35, he underwent a successful 6-antigen non-mismatch cadaveric kidney transplant from a 55-year-old diabetic donor (HbA1c: 7.9%, UPCR: 375 mg/g), with creatinine stabilizing at 0.8-1 mg/dL. Two and a half years later, he developed progressive proteinuria (UPCR rising from 200 to 6000 mg/g in one month). Despite this, creatinine remained stable (0.8-1 mg/dL), Tacrolimus levels were therapeutic (4-6 ng/mL), and PRA levels were unchanged (class I: 0.4%, class II: 0.6%). Both BK virus and CMV tests were negative. Elevated serum IgA (431 mg/dL) and a graft biopsy confirmed IgA nephropathy. His immunosuppressive regimen was adjusted: Tacrolimus was reduced from 2.5 mg QD to 2 mg, MPA was maintained, Prednisolone was increased from 1 tablet to 2 tablets, and Azathioprine, Hydroxychloroquine, and fish oil were added at 1 tablet QD. Proteinuria improved to 1100 mg/day after these changes.

Discussion

The patient was diagnosed with IgA nephropathy in the graft kidney. It is unclear if IgA nephropathy coexisted with ADPKD, contributing to early ESRD, indicating possible recurrence post-transplant. Earlier diagnosis and optimal management could have delayed dialysis. Recent advances in investigational agents for IgA nephropathy have shown promising outcomes, such as reduced proteinuria and slower GFR decline. However, these trials excluded post-transplant patients, preventing our patient from accessing these therapies. Although we adjusted the immunosuppressive regimen, Azathioprine is not known to improve long-term kidney survival. This raises the question: with IgA nephropathy as a major cause of ESRD, and the graft biopsy confirming recurrent or de novo IgA nephropathy, conventional immunosuppressive regimens may be insufficient in transplant recipients. These patients may benefit from clinical trials of novel therapies, like BAFF/APRIL inhibitors and complement-targeting treatments, to protect the

transplanted kidney. Concerns about overt immunosuppression are lessened, as many transplant recipients with aHUS(atypical hemolytic uremic syndrome) are successfully managed with complement-inhibiting therapies.

Conclusion

This case highlights the recurrence or de novo IgA nephropathy post-transplant in an ADPKD patient, revealing the limitations of current immunosuppressive regimens. Excluding post-transplant patients from trials of therapies like BAFF/APRIL inhibitors and complement-targeting treatments deprives them of promising options. Given the success of complement inhibitors in other conditions, future trials should include transplant patients to improve outcomes and protect graft survival in recurrent IgA nephropathy.

病例報告 113_C69

以持續嘔吐做為腎上腺結核表現之病歷報告

Persistent vomiting as a presentation of adrenal tuberculosis : a case report

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Introduction

Addison's disease was first described by Thomas Addison in 1855.The most common cause of Addison' disease in developed countries is autoimmune disease. Adrenal tuberculosis(TB) is a rare cause of Addison' disease due to the widespread use of antituberculous treatment. Adrenal TB usually presents with nonspecific symptoms. The diagnosis is often overlooked and delayed. Here, we report a rare case of adrenal TB presenting with persistent nausea and vomiting.

Case Report

A 46-year-old women with a history of myoma post operation visited our emergency department due to 14 days of perisistent nausea and vomiting. 16 months before this hospitalization, she first admitted to our hospital because of bilateral acute pyelonephritis. 1 month and 10 days before this hospitalization, she had right flank pain with fever. Abdominal computed tomography showed multiple calcified spots in the liver, spleen, peritoneum, bilateral adrenal gland and heterogeneous enhancement of right kidney with perirenal fat stranding. Gastroenterologist and oncologist considered the calcifications of liver, spleen were due to chronic infection or inflammation. She called on urologic and infectious outpatient department because of urinary tract infection. Ureteroscopy was performed. The pathology of right uretero-renal scopy biopsy was small non-caseating granuloma. Urine culture yielded mycobacteria TB complex. She was diagnosis as having urinary TB. 2 weeks before this hospitalization,she was sent to our ER due to persistent nausea and vomiting. Lab data showed hyponatremia 113 meq/L. Low cortisol 3.55μg/dL, high ACTH >1250 pg/mL, low aldosterone 2.57 ng/dL and normal plasma renin activity 1.46 ng/ml/hr. Physical examination revealed hyperpigmentation of skin and oral mucosa. Addison's disease due to adrenal TB was diagnosed based on hormone study and calcifications in both adrenal glands on CT scan. Prednisolone 10 mg twice daily was started. On the eighth day of hospitalization, her condition was stable and she was discharged.

Discussion

Treatment for adrenal TB with Addison disease was Anti-TB drug and steroid .However, determining the appropriate steroid dose and monitoring treatment outcomes for adrenal insufficiency in adrenal TB proved challenging. We adjusted the steroid dose based on ACTH levels and the patient's clinical condition during outpatient follow-up visits.

Conclusion

The clinical features of adrenal TB may be nonspecific, leading to delayed diagnosis. Clinicians should maintain a high index of suspicion for adrenal TB, especially true in patients with skin hyperpigmentation and bilateral adrenal calcifications on CT scan. Early diagnosis of adrenal TB

can prevent the severe morbidity and mortality of adrenal insufficiency.

病例報告 113_C70

急性上腸系膜動脈栓塞: 一件伴有長期新型口服抗凝血劑使用的案例

Acute superior mesenteric artery embolism (ASMAE): a case accompanied with long-term novel oral anticoagulant (NOAC) use

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Introduction

Acute superior mesenteric artery embolism (ASMAE), is a severe embolic event with high mortality if delayed diagnosis. The emboli was mostly cardiac cause, and NOAC is commonly prescribed for prevention of emboli formation. However, the NOACs may fail. Here, we present a case with regular dabigatran due to atrial fibrillation but ended up with ASMAE.

Case Report

This 71-year-old retired man has hypertension, diabetic mellitus, chronic kidney disease, atrial fibrillation with dabigatran 150mg BID for 5 years and switched to 110mg BID 7 months ago, and the past history of recurrent ischemic stroke attack 12 and 3 years ago. He had intermittent right upper quarter pain for two weeks followed by acute onset, severe, constant peri-umbilical pain for one hour. Abdominal fullness and constipation was also noticed without fever, vomiting or bloody stool. Labs showed WBC 13000 /μL, neutrophil dominance, PT 13.6 seconds, APTT 39.6 seconds, lactate 28.2 mg/dL. Computer tomography angiography revealed an embolism at the superior mesenteric artery without filling defect or pneumatosis intestinalis. Echo showed atrial fibrillation with bi-atrium enlargement. He received percutaneous mechanical thrombectomy with angiojet, heparin pump and left atrial appendage closure. After 13 days of hospitalization, he was discharged with Apixaban 2.5mg once per day.

Discussion

ASMAE presents with acute onset of severe, contact, non-localized or periumbilical, cramp-like abdominal pain, which is disproportionate to the physical examination findings, and maybe combine with vomiting or diarrhea. The laboratory tests inculde leukocytosis, metabolic acidosis, elevatied D-dimer and lactate. Both the clinical and laboratory findings are not specific for the ASMAE, which may lead to delayed diagnosis, causing irreversible intestinal obstruction or gangrene. However, ASMAE early disagnosis within 24 hours could keep the survival rate at about 89.4%, with the help of computed tomography angiography.

Due to the thrombosis is mainly from the heart, patients with atrial fibrillation have higher risk of SMA embolism. NOAC was prescribed for ischemiac events prevention. However, NOAC failure rate for atrial fibrillation ischemia events may be higher then predicted in the real world. The possible causes include the patient factors, prescription errors, other competing stroke mechanisms, or excessive cardioembolic stroke burden related to atrial fibrillation itself despite sufficient anticoagulation. Switching to another NOAC may be better then vitamin K antagonist with less complication if the NOAC failure. Though better prevention rate was not ensured.

Conclusion

If the patients with atrial fibrillation even receiving NOAC present with severe acute abdominal pain, ASMAE should be taken into consideration for high mortality rate due to delayed diagnosis and possible NOAC failure.

病例報告 113_C71

病例報告-無症狀的 IgG4 相關自體免疫性胰臟炎

A case report of asymptomatic IgG4-related autoimmune pancreatitis

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Introduction

Autoimmune pancreatitis (AIP) is a unique form of chronic pancreatitis with autoimmune characteristics, often presenting in ways that mimic pancreatic cancer, complicating accurate diagnosis. Hallmarks of the disease include pancreatic enlargement, narrowing of the main pancreatic duct, elevated serum IgG4 levels, autoantibodies, and characteristic histopathological findings. In this report, we discuss the case of a 46-year-old asymptomatic male whose AIP diagnosis was incidentally discovered during routine imaging.

Case Report

A 46-year-old male with a medical history including gout, hypertension, hyperlipidemia, and diabetes mellitus, was referred to the gastroenterology outpatient department following the incidental discovery of a pancreatic mass during a routine abdominal ultrasound. The patient had no symptoms such as fever, abdominal pain, jaundice, or weight loss. Laboratory workup revealed elevated lipase levels, though amylase, complete blood count, and liver function tests were within normal limits. Additionally, autoantibody tests including ANA and CA-19-9 were negative.

Initial abdominal ultrasound identified a 4.20 x 2.74 cm ill-defined hyperechoic lesion in the pancreas. Subsequent abdominal computed tomography showed diffuse pancreatic swelling with peripheral enhancement and enlarged lymph nodes in the left inguinal and peri-pancreatic regions. Endoscopic ultrasound (EUS) confirmed diffuse pancreatic swelling and a coarse parenchyma with a hypoechoic rim. Fine needle biopsy (FNB) from the pancreas revealed normal pancreatic morphology, with negative IgG immunostains, excluding malignancy. However, a biopsy of the ampulla of Vater showed chronic inflammation and elevated IgG4-positive plasma cells. This, combined with an elevated serum IgG4 level of 712.6 mg/dL, confirmed a diagnosis of IgG4-related AIP. The patient was prescribed 20 mg of prednisolone and scheduled for regular follow-up.

Discussion

Autoimmune pancreatitis is divided into two types: Type 1, linked with IgG4-related disease and systemic involvement, and Type 2, which is confined to the pancreas and unrelated to IgG4. This patient was diagnosed with Type 1 AIP based on elevated serum IgG4 levels, pancreatic inflammation, and IgG4-positive plasma cells in the biopsy.

Diagnosing AIP can be particularly challenging due to its similarities with pancreatic cancer, as both can present with painless jaundice, weight loss, and pancreatic masses. In this case, diagnosis was further complicated by the absence of symptoms and the incidental discovery of the pancreatic lesion. However, elevated serum IgG4 and IgG4-positive plasma cells in the biopsy were critical in differentiating AIP from malignancy.

Radiological findings in AIP typically show diffuse or segmental pancreatic enlargement with delayed enhancement. EUS plays a key role, offering high-resolution imaging and facilitating fine needle biopsy. However, in some cases, FNB may yield normal pancreatic tissue, necessitating a broader evaluation incorporating clinical, serological, and histological evidence.
The diagnostic criteria for AIP include imaging evidence of pancreatic enlargement, elevated serum IgG or IgG4 levels, histopathological evidence of fibrosis, and response to steroid therapy. The standard treatment involves corticosteroids, with a typical regimen of prednisone at 0.6-1.0 mg/kg per day. This patient, started on 20 mg of prednisolone, will require long-term monitoring due to the potential risk of relapse and the need for maintenance therapy.

Conclusion

IgG4-related autoimmune pancreatitis is frequently misdiagnosed as acute or chronic pancreatitis or even pancreatic cancer due to its overlapping clinical presentations. To ensure timely diagnosis and treatment, systematic imaging, serological testing, and biopsy when needed are crucial. Where diagnosis remains uncertain, a trial of corticosteroid therapy can help clarify the condition and guide further management.

病例報告 113_C72

法布瑞氏症病例報告

A 45-year-old man with stage 3 CKD and diagnosed with Fabry disease

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Introduction

Fabry disease is a rare X-linked lysosomal storage disorder caused by mutations in the GLA gene, leading to deficient activity of the enzyme α-galactosidase A. This deficiency results in the accumulation of globotriaosylceramide (Gb3) in various tissues, including the kidneys, heart, and nervous system. Here, we discuss a case of a 45-year-old male with Fabry disease diagnosed following an episode of unstable angina.

Case Report

This is a 45-year-old male laborer with underlying of CKD stage 3 and hypertension.
According to the patient, he was admitted on August, 2023 due to unstable angina. CKD stage 3 was diagnosis accidently. Serum creatinine was 1.5 mg/dl, GFR was 49 ml/min/1.73m^2. Urine examination revealed protein 2+. The urine total protein to creatinine ratio was 816.5 mg/g. Renal ultrasonography was arranged. The right kidney was 10.9 cm in size with 2 renal cyst (1.11cm, 0.58cm) and left kidney was 9.7 cm in size. The patient mentioned that his elder sister and sister's two sons have diagnosis of Fabry's disease before.
Therefore, Fabry disease screen test was performed. The alpha-galactosidase A showed 0.25 uM/hr, and DBS LysoGB3 showed 8.44 ng/mL. A genetic study for Fabry disease was conducted, revealing a homogeneous mutation, specifically the c.656T>C (p.Ile219Thr) mutation in exon 5 of the *GLA* gene.
Renal biopsy was perform on Aprial, 2024. The biopsy contained renal medulla and cortical parenchyma containing up to 11 viable glomeruli. Eight glomerulus showed global sclerosis. One glomerulus show segmental sclerosis. The non-sclerotic glomeruli show essentially normal in size, cellularity, with small mesangium, thin capillary walls and mild compressed lumina. There are significant foamy cell change of the podocytes, resulting in cell enlargement and mild compressed the capillary loops. Tubular atrophy and interstitial fibrosis involved about 30–40% of sampled cortex. There are also mild leukocytes, composed predominantly lymphocytes, infiltrate in the interstitium of scarred area. The arteries show moderate to severe thickening. Tissue for immunofluorescence showed 1 glomeruli were negative for IgA, IgG,IgM, C3 and C1q. The electron microscopic findings showed there are many laminated, electron-dense lipid deposits in podocytes, and distal tubules. The podocytes show partial foot processes effacement, and detachment, affecting less than 50% of the peripheral capillary loops. The morphological picture is consistent with Fabry's disease.

Discussion

Fabry disease results from the deficient activity of α-galactosidase A, leading to the accumulation of globotriaosylceramide in lysosomes. The resulting accumulation of glycosphingolipids within

cells leads to various symptoms, including heat intolerance with abnormally decreased sweat and tear production, heart disease, and cerebrovascular accident. Prevalence in white male populations has been linked to Fabry disease in a wide range, approximately 1:17,000 to 1:117,000.[1] The differential diagnosis for Fabry disease includes other lysosomal storage disorders, such as Gaucher disease and Niemann-Pick disease, as well as other causes of renal disease, heart disease, and neurological symptoms.[2] The "gold standard" to clarify if a novel mutation is pathogenic or likely benign includes in vitro GLA mutation expression assays. Evidence of lysosomal globotriaosylceramide accumulation in renal or cardiac biopsies, although invasive, may be required when interpretation of genetic GLA mutation is challenging particularly when the clinical signs are nonspecific, alternative or additional diagnoses are under consideration. Enzyme replacement therapy (ERT) with recombinant α-galactosidase A is the mainstay of treatment for Fabry disease. ERT can slow the progression of the disease and improve symptoms. Other supportive therapies may be needed to manage specific complications.[3]

Conclusion

This case highlights the importance of considering Fabry disease in the differential diagnosis of patients with unexplained CKD or heart disease, especially if there is a family history of the disease.

病例報告 113_C73

後腹腔積氣，一個少見且易跟腹腔積氣搞混的影像

Pneumoretroperitoneum, a rare and confusing image with pneumoperitoneum

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Introduction

Pneuperitoneum is an acute abdomen and usually need emergent intervention. In contrast, pneumoretroperitoneum is a rare condition which often derived from colonscopic intervention or retroperitoneal abscess. Besides, pneumoretroperitoneum is some times confusing with pneumoperitoneum in image. Herein, we presented a case of pneumoretroperitoneum secondary to pneumothorax. It underscores the importance of careful clinical evaluation and decision-making, invasive procedures may not always be necessary, and a 'wait and see' approach can sometimes be the most appropriate course of action.

Case Report

A 46-year-old female with metastatic sigmoid adenocarcinoma presented to our institution due to fluorouracil-related organizing pneumonia. She was soon admitted to the intensive care unit due to respiratory failure, and a right-sided pneumothorax developed during mechanical ventilation. We placed a chest tube in the right chest, but initially, poor drainage function was reported. Subsequently, routine X-ray screening revealed new developed left-side pneumothorax and subcutaneous emphysema (Figure 1A). Plain abdominal radiography showed radiolucency over bilateral kidneys (Figure 1B). We were ever afraied of hollow organ perforation and we arranged emergent abdominal computer tomography. Fortunately, further abdominal computer tomography not only confirmed bilateral pneumothorax and pneumomediastinum but also showed pneumoretroperitoneum around bilateral kidneys (Figure 2). All the free air in her abdomen was restricted within bilateral Gerota's fascia, with none leaking into the peritoneal space. We switched to a larger chest tube and kept her depending on ventilator, and her pneumothorax, pneumomediastinum, and pneumoretroperitoneum gradually resolved. Throughout the entire course, her abdomen remained soft, and she never exhibited signs of shock or dyspepsia.

Discussion

Previously, pneumoretroperitoneum was considered a rare clinical condition and was thought to be related to invasive procedures on retroperitoneal organs. Concomitant pneumothorax, pneumoretroperitoneum, pneumomediastinum, and subcutaneous emphysema developed after colonoscopic intervention have been reported several times so far.[1,2,3] In these cases, free air in the retroperitoneal space was directly derived from a perforated colon and then spread to the mediastinum, thorax, and subcutaneous space. Additionally, severe infection with gas-forming organisms in retroperitoneal organs like the kidney or psoas muscle could also cause pneumoretroperitoneum.[4] In other words, all of them had an index event like an invasive procedure or retroperitoneal abscess, which made it easy to correlate the origin of free air. None

of them developed in the intensive care unit.
It was challenging and confusing for us when first seeing the presenting image. Excluding acute abdomen with hollow organ perforation should always be the first priority, and we checked for this with an emergent abdominal computer tomography. In our case, we hypothesized that her right pneumothorax progressed first due to poor drainage function and then leaked into the mediastinum, left chest cavity, and finally bilateral Gerota's fascia. The absence of intraperitoneal free air made us confident about our theory. We chose the strategy of 'wait and see' rather than exploratory laparoscopy. Since no operation was done, we will never know the answer. However, the lack of shock signs and dyspepsia might indirectly support our hypothesis.

Conclusion

This case has broadened our understanding of the potential causes of pneumoretroperitoneum, highlighting that poorly drained pneumothorax can also lead to air leakage into the retroperitoneum space. Furthermore, it underscores the importance of careful clinical evaluation and decision-making, as invasive procedures may not always be necessary, and a 'wait and see' approach can sometimes be the most appropriate course of action.

病例報告 113_C74

一位雙瓣替換病史的患者感染嚴重退伍軍人症合併心臟侵犯

Severe Legionnaires' disease with cardiac involvement in a patient with a history of double valve replacement

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Introduction

Legionnaires' disease (LD) is a common cause of community-acquired pneumonia. Despite the fact that urine antigen testing (UAT) makes early detection possible, clinicians usually under-diagnose LD, which leads to delayed treatment. Besides, LD is related to various extrapulmonary presentations. Herein, we present a case of severe LD with cardiac involvement in a patient with a history of double valve replacement.

Case Report

A 54-year-old male with history of 1. permanent atrial fibrillation with biatrial anlargement 2. Barlow's mitral valve disease with chordae tendineae rupture (A1), severe mitral regurgitation, moderate aortic regurgitation, tricuspid regurgitation and LV dilation underwent aortic and mitral bioprothesis valve replacement with tricuspid ring annuloplasty on March 22, 2022, presented to our hospital due to shortness of breath for one month. After the cardiac surgery, he recovery well and could carry out daily lifes himself. Despite atrial fibrillation persisted, systolic function of his heart is acceptable and no residual mitral or aortic regurgitation was found. After admission, fever up to 39.1°C with septic shock attacked. Lab showed acute decompensated heart failure with multiple organ failure.

At first, we thought it was a worsening of valvular heart disease due to superimposed infection or prothesis failure, but subsequent echocardiograms revealed dilated left ventricle with global hypokinesia but no regurgitant flow, which overturned our assumption. We started empirical minocyclin to cover atypical pathogen and his condition improved rapidly. Subsequent chest X ray showed left lower lobar pneumonia and positive urine legionella antigen test was found. His fever subsided on day 4 and bedside echocardiogram on day 5 demostrated complete resolution of wall motion abnormality. Besides, his liver and kidney function gradually returnned to normal range within one week. He discharged after 11 days of admission. We followed up this patient for one year and no sequelae was ever found.

Discussion

Legionellosis is a waterborne disease that has two clinical presentations: Pontiac fever, a benign non-pneumonic disease, and LD, a severe pulmonary infection. Pontiac fever is a self-limited disease that often presents within 48 hours after exposure to the pathogen. In some studies, up to 40% of patients with LD require admission to intensive care units. Because Legionella is a common pathogen of community-acquired pneumonia, early diagnosis and treatment of LD are therefore important. Two methods are now commonly used to diagnose LD, including UAT and polymerase chain reaction (PCR) of respiratory specimens. UAT has a moderate sensitivity of 70–90% and a

specificity of almost 100% for the diagnosis of LD caused by serogroup 1. PCR of respiratory specimens is an emerging methods to detect Legionella spp. and have been increasingly used in recent 10 years. The only drawback of PCR is that it's too expensive and need specific equipment, which makes it unavailable in local hospital.

Severe LD might cause various extrapulmonary presentations. Among these extrapulmonary presentations, the heart is the most commonly affected organ. In those case series, the diagnosis of LD-related myocarditis was made by concomitant infection of Legionella and newly presented global left ventricular systolic dysfunction. An endomyocardial biopsy is usually not necessary for diagnosis because of the peri-procedure risk, and it may not be able to demonstrate interstitial inflammatory infiltrates. We didn't perform an endomyocardial biopsy due to concerns about procedure risk, and our patient recovered after adequate treatment. In our case, the history of cardiac surgery confused us first, but new onset global hypokinesia and concomitant infection of Legionella overturned our previous ideas.

Conclusion

LD is a potential cause of severe pneumonia and might cause extrapulmonary involvement. Early detection through UAT and prompt treatment can significantly improve patient outcomes. Besides, we should never attribute a worsening heart failure in a patient who ever underwent valve surgery to prothesis failure. A thorough and careful examination can help us better diagnose and treat patients.

病例報告 113_C75

Sulfasalazine 導致藥物反應伴隨嗜伊紅性白血球增加與全身症狀

Sulfasalazine-related drug reaction with eosinophilia and systemic symptoms (DRESS)

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Introduction

Drug Reaction with Eosinophilia and Systemic Symptoms (DRESS) is a severe, idiosyncratic drug reaction characterized by a long latency period. It presents with a variety of clinical manifestations, including an extensive skin rash accompanied by visceral organ involvement, lymphadenopathy, eosinophilia, and atypical lymphocytosis. DRESS typically occurs 2 to 8 weeks after starting medication. Even discontinuing drug, disease flares may occur.

Case Report

A 58-year-old male manager with history of ulcerative colitis, treated with mesalazine for five years, and then been switched to sulfasalazine since March 2024. Three weeks before admission, he complained bone pain and sore throat, followed by skin rash that initially noted over his trunk and spreaded to his limbs later, with fever up to 39°C. Despite receiving anti-allergy treatment, his symptoms still worsened, including generalized itching, cough, husky voice, lip swelling, and neck swelling.

Upon re-evaluation, laboratory survey reported elevated white blood cells (11,900/uL), Eosinophilia (16.6%), lactate (32.1 mg/dL), ALT (189 U/L), LDH (297 U/L), hsCRP (1.47 mg/dL), r-GT (262 U/L), alkaline phosphatase (221 U/L), ferritin (345.6 ng/mL), and D-dimer (1,323.5 ng/mL). Neck CT revealed diffuse swelling and numerous enlarged lymph nodes. Dermatological examination showed neck and facial edema, erythematous patches, and purpuric lesions, but no mucosal involvement nor bullae. Tracing related history, there was no exposure of raw food, no contact with pets and insect bites.

DRESS syndrome was diagnosed, with a RegiSCAR score of 6, favored triggered by sulfasalazine. He received Tazocin (2# Q8H, for 7 days) and methylprednisolone (0.5# Q12H for 10 days, then 0.5# QD for 2 days, and then oral form Prednisolone 1.5# BID) as treatment. Patient's condition got improved gradually, and he got discharged with follow-up instructions.

Discussion

DRESS is a rare but potentially life-threatening drug reaction that requires prompt recognition and management. Varied symptoms and delayed onset can make diagnosis challenging. Therefore, maintaining a high index of suspicion, especially for patients on high-risk medications, is crucial. For management, discontinuing the culprit drug is first step, followed by supportive treatment. Closely monitor clinical symptoms and complications is important for assess long-term sequelae. The prognosis for DRESS varies under the severity of the reaction and the extent of organ involvement. While most patients get recovered completely, some may suffered long-term complications, including autoimmune disorders and psychological impact.

Conclusion

DRESS is a severe drug reaction characterized by a delayed onset, rash, fever, eosinophilia, and multi-organs involvement. Prompt recognition, discontinuation of the offending medication, and appropriate supportive care are essential.

病例報告 113_C76

隱藏在鼻子內的魔鬼

Invasive Fungal Rhinosinusitis With Rhizopus: A Case Report

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Introduction

Fungal rhinosinusitis covers a spectrum of infections, ranging from relatively benign to rapidly fatal conditions. Fungal colonization of the upper and lower airways is common, as spores are constantly inhaled into the sinuses and lungs.

While invasive fungal rhinosinusitis can occasionally affect individuals with intact immune systems, it predominantly occurs in patients with some form of immunosuppression. This condition can be broadly categorized into acute and chronic forms.

Case Report

A 64-year-old Taiwanese housewife, with a medical history of poorly controlled type 2 diabetes mellitus (HbA1c 9.3%), end-stage renal disease on peritoneal dialysis since June 2021, and hypertension, presented with right eye swelling for two days.

Two days prior to her emergency department visit, she developed fever, right eye swelling, nasal bleeding, chills, and drowsiness. On examination, she was febrile and tachycardic. Physical examination revealed right orbital swelling, conjunctival chemosis, ptosis, eye discharge, and limited extraocular movement.

Laboratory tests showed leukocytosis with neutrophil predominance, elevated high-sensitivity C-reactive protein, elevated procalcitonin, hyperglycemia without diabetic ketoacidosis, hyponatremia, and renal impairment (BUN: 70 mg/dL, serum creatinine: 8.88 mg/dL). A head CT revealed air-fluid levels in the right paranasal sinuses (maxillary, ethmoidal, sphenoidal) with involvement of the right retrobulbar space, optic nerve, and retroantral fat plane, consistent with invasive right paranasal sinusitis with orbital extension.

Empiric antibiotic therapy with intravenous Amoxicillin-Clavulanate was initiated for sinusitis. The patient was admitted to the infectious diseases ward, where treatment was adjusted to Piperacillin-Tazobactam and Amikacin due to persistent fever. An ophthalmology consultation on day 3 identified right ocular hypertension. ENT consultation revealed bilateral nasal cavity debris, mucopus, and no visible tumors, confirming acute rhinosinusitis. Subsequent sinus CT showed bilateral frontal, ethmoidal, and sphenoidal sinus involvement with right retrobulbar infiltration, suggesting disease progression.

Emergency surgery was performed on day 5, revealing extensive black eschar and severe bone necrosis. Intraoperative specimens tested positive for Aspergillus, leading to the addition of liposomal amphotericin B for fungal sinusitis. Cultures obtained during the operation identified Klebsiella pneumoniae, Escherichia coli, and Enterobacter cloacae complex, all sensitive to Piperacillin-Tazobactam and Amikacin, so the current antibiotic regimen was continued. A second surgery on day 10 showed minimal improvement.

Further pus cultures from the second operation revealed Rhizopus spp., confirming acute invasive

fungal sinusitis with Rhizopus spp. isolation. A third surgery on day 15 showed some improvement. Unfortunately, the patient succumbed to sudden death on day 18, with aspiration or choking as the suspected cause.

Discussion

The primary pathogens responsible for acute invasive fungal sinusitis are species of Aspergillus and Mucorales. Prompt initiation of antifungal therapy is critical once clinical suspicion arises. Surgical debridement with clear margins is strongly recommended for controlling the disease and obtaining tissue samples for histopathological and microbiological diagnosis. Additionally, the removal of all necrotic tissue and the drainage of any obstructed sinuses are essential objectives in surgical management.

病例報告 113_C77

AIDS 患者驚險一線：粟粒性肺結核併發急性呼吸窘迫，VV-ECMO 成功逆轉

An Unusual Case of Miliary Tuberculosis Presenting as ARDS in a Newly Diagnosed AIDS Patient: Successful Weaning from VV-ECMO

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Introduction

Miliary tuberculosis (TB) is a serious and uncommon manifestation characterized by the hematogenous dissemination of *Mycobacterium tuberculosis*, which can present as a life-threatening condition. Cases of miliary TB complicated by acute respiratory distress syndrome (ARDS), septic shock, and multiorgan dysfunction syndrome are rare but significantly increase mortality rates, underscoring the urgent need for timely diagnosis and effective treatment. The management of miliary TB presenting as ARDS poses substantial challenges, particularly in patients with HIV infection, a recognized risk factor for extrapulmonary tuberculosis. Atypical presentations associated with HIV can complicate diagnosis and introduce difficulties in treatment.

In this report, we present a case of severe ARDS due to miliary TB in a newly diagnosed HIV patient, emphasizing the complexities of management and the successful use of anti-tuberculous therapy in conjunction with veno-venous extracorporeal membrane oxygenation (VV-ECMO), ultimately leading to successful weaning from ECMO support.

Case Report

A 31-year-old male with no prior systemic disease presented with progressive malaise over two months, accompanied by dysphagia, anorexia, fever with evening spikes, night sweats, and a 15 kg weight loss. He reported worsening exertional dyspnea and dry cough, particularly in the two days prior to admission, prompting his visit to the emergency department.

Desaturation with hypotension upon presentation. Chest X-ray revealed bilateral micronodular opacities (miliary pattern), predominantly in the right and upper lobes. Laboratory findings indicated pancytopenia, disseminated intravascular coagulation (DIC), acute kidney injury (AKI), and lactic acidosis. His condition rapidly deteriorated, with follow-up imaging showing bilateral progressive consolidation, leading to a suspicion of ARDS and septic shock with multiorgan failure. He was subsequently transferred to an isolated room in the intensive care unit (ICU) for further management.

In the ICU, the patient's PaO2/FiO2 ratio continued to decline despite lung protective strategies and prone positioning, necessitating the initiation of VV-ECMO for respiratory support. Vasopressors and continuous veno-venous hemofiltration with dialysis (CVVH-DF) were also initiated for septic shock-related lactic acidosis.

Subsequent sputum cultures yield positive result for acid-fast bacilli, and Mycobacterium tuberculosis polymerase chain reaction (PCR) testing confirmed the diagnosis. Positive acid-fast stains were also detected in gastric fluid, stool, and urine. HIV testing yielded positive results, with a CD4 count of less than 50.

Empirical broad-spectrum antibiotics, including meropenem, were administered for severe pneumonia in the immunocompromised patient at first, followed by the initiation of anti-TB therapy with amikacin and levofloxacin. Steroids were also prescribed upon ICU admission. Challenges in initiating first-line anti-TB therapy arose due to poor oral absorption and renal and hepatic dysfunction. Early consultation with an infectious disease specialist proved beneficial in guiding effective therapy. After 10 days on ECMO, the patient demonstrated a rapid response with signs of recovery, including improvement in both the PaO2/FiO2 ratio and imaging results. He was successfully weaned from ECMO on day 10. While the patient has been weaned from ECMO, he still faces challenges requiring ongoing monitoring and treatment for complete recovery.

病例報告 113_C78

非手術性老虎黴素肋膜沾黏術用於治療反覆自發性氣胸在一位全身性硬化症合併間質性肺病病患

Nonsurgical tigecycline pleurodesis for the management of recurrent spontaneous pneumothorax in a patient with systemic sclerosis and interstitial lung disease

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Introduction

Systemic sclerosis (SSc) is an autoimmune connective-tissue disease and interstitial lung disease (ILD) is the leading cause of death in patients with SSc. Spontaneous pneumothorax is an uncommon but serious complication in patients with SSc-associated ILD (SSc-ILD). The secondary spontaneous pneumothorax often presents as a difficult-to-treat condition and the recurrency rate is high. Here, we report a patient with SSc-ILD complicated with recurrent spontaneous pneumothorax which relieved by chest tube insertion and nonsurgical tigecycline pleurodesis.

Case Report

A 54-year-old woman who had a 30-year history of SSc with Raynaud's phenomenon, progressive diffuse skin tightening, cutaneous telangiectasia and positive anti-topoisomerase 1 (anti-Scl-70) Ab was hospitalized because of both feet ulcerative wounds infection for 1 month. In these 4 years, the patient was treated with low dose steroid, hydroxychloroquine and mycophenolic acid. ILD was confirmed by high-resolution computed tomography (HRCT) with usual interstitial pneumonitis (UIP) pattern that nintedanib was used in the last 2 years. The patient also took sildenafil for pulmonary hypertension. However, this time at emergency department, left large pneumothorax was accidentally found, that pig-tail catheter drainage was inserted. The follow-up chest X-ray revealed fully lung expansion and pig-tail catheter was removed 1 week later. Unfortunately, after 3 days, left large pneumothorax recurred, that chest tube drainage was performed. We consulted the thoracic surgeon for persistent air leak and chemical pleurodesis was suggested according to poor pulmonary function and multiple comorbidities. Thus, pleurodesis with tigecycline was performed twice with a 3-day interval. Supplemental oxygen was continually used. Under relative stable condition, the patient went home with chest tube drainage. One month after pleurodesis, the chest tube was removed with residual small pneumothorax.

Discussion

Spontaneous pneumothorax occasionally would be found in patients with connective tissue disease-associated ILD, including systemic sclerosis. The possible mechanism might be resulted from the rupture of acquired subpleural cysts with surrounding diffuse interstitial fibrosis. A lower body mass index and greater extent of lung reticular infiltrations were found to be risk factors of pneumothorax development. Pneumothorax was demonstrated to be associated with poor prognosis. The higher pneumothorax recurrency rate was thought to be related to diffuse interstitial pneumonitis, rigidity of lung parenchyma, or the SSc medications, including corticosteroids, which may aggravate persistent air leak. These patients commonly receive

thoracic drainage, and surgery would not be performed because of their poor general condition. Chemical pleurodesis with tetracycline or talc has been successfully used to avoid recurrent pneumothorax, but these agents can contribute to the onset of acute exacerbation of the interstitial fibrosis. Tigecycline, an antibiotic derivative of tetracycline, has been proved an effective and safe pleurodesis agent in rabbit models and in patients with liver cirrhosis and hepatic hydrothorax. In our case, the patient tolerated the tigecycline pleurodesis without worsening interstitial pneumonitis and, eventually, the chest tube was removed successfully.

Conclusion

Although an uncommon manifestation, clinical physicians should be aware of spontaneous pneumothorax management in patients with SSc-ILD. Nonsurgical tigecycline pleurodesis would be one of the treatment options. Future studies are required for the optimal treatment strategy.

病例報告 113_C79

腹膜結核偽裝成惡性腫瘤腹膜轉移：病例報告與診斷挑戰

Peritoneal Tuberculosis Mimicking Carcinomatosis in a Patient with Alcoholic Liver Cirrhosis: A Case Report and Diagnostic Challenge

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Introduction

Tuberculosis (TB), caused by Mycobacterium tuberculosis, remains a significant public health challenge due to its ease of transmission and the complexity of its diagnosis, treatment, and prevention[1]. While TB primarily affects the lungs, resulting in pulmonary TB, it can also involve other organs, a condition referred to as extrapulmonary tuberculosis (EXPTB)[2]. EXPTB is more commonly observed in immunocompromised individuals[3].

This case report details the clinical course of a 44-year-old male with a history of alcoholic liver cirrhosis who was diagnosed with peritoneal tuberculosis, mimicking carcinomatosis. Notably, in this case, diffuse invasion without a specific focal lesion was observed on imaging, which differs from previous case reports.

Case Report

A 44-year-old male with a known history of Hepatitis B virus carrier status and alcoholic liver cirrhosis complicated with ascites came to the emergency department with complaints of general weakness, abdominal distension, and bilateral lower limb edema.

Upon arrival, the patient's vital signs were stable, and there was no fever. Laboratory findings revealed pancytopenia, hypokalemia, hypoalbuminemia, conjugated hyperbilirubinemia, elevated alkaline phosphatase, and impaired liver function. Abdominal computed tomography demonstrated liver cirrhosis with splenomegaly, collateral circulation, and massive ascites.

Given the patient's decompensated liver cirrhosis with massive ascites, this patient was suggested to receive liver transplantation. A pre-transplantation workup revealed elevated tumor markers, including CA-125 (632.8 U/mL) and CA19-9 (50.7 U/mL). Positron emission tomography (PET) scan demonstrated increased FDG uptake in multiple regions of the abdomen and ascites, suggesting possible FDG-avid malignancy or carcinomatosis. Esophagogastroduodenoscopy and colonoscopy were performed but did not reveal any malignancy.

Given the findings, a laparoscopic biopsy was performed. Biopsy of the great omentum revealed necrotizing granulomatous inflammation, suggesting a diagnosis of peritoneal tuberculosis (TB). Moreover, sputum culture confirmed mycobacterium tuberculosis complex, and an acid-fast bacillus (AFB) stain was positive. The patient was started on anti-TB therapy, including Rifampin, Isoniazid, Ethambutol, and Levofloxacin for disseminated TB. Following clinical stabilization, the patient was discharged and continued on anti-TB treatment with outpatient follow-up.

Discussion

Abdominal tuberculosis (TB) accounts for approximately 5% of all TB cases and 2.7-21% of extrapulmonary TB cases[4,6]. Risk factors include immunocompromised conditions, underlying

malignancy, diabetes mellitus, HIV infection, liver cirrhosis, renal insufficiency, tobacco use, and alcoholism[6,7,8]. TB peritonitis is the second most common form of abdominal TB[8]. It is most commonly observed in individuals aged 35-45 years[9]. It presents in three distinct forms: wet, dry-plastic, and fibrotic-fixed. The wet form, characterized by loculated fluid or free ascites, accounting for approximately 90% of cases[5,10]. Diagnosis typically requires a combination of investigations, as no single test is usually sufficient[2].

Radiological investigations can be helpful in diagnosing TB peritonitis. These modalities may reveal free, loculated, or localized ascites, lymphadenopathy, and peritoneal thickening11. However, distinguishing TB peritonitis from peritoneal carcinomatosis can be challenging. Several case reports describe peritoneal tuberculosis mimicking carcinomatosis[6,12,13]. Studies have investigated the diagnostic performance of CT in differentiating TB peritonitis from peritoneal carcinomatosis, showing that smooth peritoneal thickening has reasonably good diagnostic accuracy[14]. FDG-PET-CT can also aid in the diagnosis. One study found that extensive involvement, uniform distribution, and smooth uniform thickening are suggestive of TB peritonitis[15].

For definitive diagnosis, invasive diagnostic testing may be required. Laparoscopy is considered the ideal method as it allows for magnified visualization of the peritoneal surfaces[16,17]. According to the World Health Organization (WHO) guidelines, pulmonary and extrapulmonary TB should be treated with the same treatment regimens[18,19].

Conclusion

Peritoneal TB could mimic carcinomatosis on PET scan. Tissue biopsy is essential for definite diagnosis and further treatment.

病例報告 113_C80

一位壞死性肺炎病人合併膿胸及 Takotsubo Syndrome

A case of necrotizing pneumonia complicated with empyema and Takotsubo syndrome

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Introduction

Necrotizing pneumonia is a rare complication of community-acquired bacteria pneumonia. It is characterizied by pulmonary inflammation with consolidation, peripheral necrosis and multiple small cavities[1]. Its cause is owing to the virulence factor of the microorganism and the predisposing factor of the host. We wanted to report a case with necrotizing pneumonia complicated with prolonged hospital course and morbidities.

Case Report

The patient's initial vital signs at triage were temperature: 37.4 Celsius degree, pulse rate: 121 beats per minute, respiratory rate: 26 times per minute, blood pressure: 88/57 mmHg, SpO2: 92% under ambient air. Physical examiantion revealed regular and rapid pulses, left side decreased breathsound, cold limbs and prolonged capillary refilling time. Chest percussion showed dullness over left lower chest over posterior side. Chest X-ray showed left side diffuse alveolar pattern and heart size was within normal range. EKG showed lead V1~V3 ST segment elevation but without reciprocal change. Bedside cardiac sonography showed LVEF around 20~25% but the window was poor. Emergent coronary angiography was activated. However, due to unstable blood pressure, norepinehprine pump was given and the patient still presented hypotension status. Under the impression of concomitant cardiogenic shock, venoarteial ECMO was implanted. The coronary angiography showed insignificant coronary artery disease. The patient was then sent to intensive care unit for further care. Laboratory data revealed WBC: 7500/uL, Neu.: 83.6%, Lym.: 8.9%, Mono.: 7.4%, Hb: 13.2 g/dL, Platelet: 159000/uL; BUN: 43 mg/dL, Cr: 3.35 mg/dL, Na: 132 mmol/L, K: 4.0 mmol/L, hsCRP: 34.53 mg/dL, Lactate: 41.1 mg/dL, TnI: < 0.01 ng/mL, NT-proBNP: 6070 mg/dL.
At the ICU, bedside echo showed left side pleural effusion and the chest X-ray showed white-out of left lung. The diagnosis of this patient was then switched to left lung empyema complicated with septic shock. Thoracentesis was performed and stick and brown pus was drained. Due to difficulty in aspiration, pig-tail was tried but it failed, either. Chest tube was then inserted. The blood culture later showed the growth of Pseudomonas aeruginosa which was the same as the pathogen seen in pleural fluid specimen. After the antibiotcis treatment and drainage, his vital signs were gradually stabilized. He was then discharged with left lung chest tube remained for necrotizing status of left lung.

Discussion

The most common pathogen of necrotizing pneumonia are *Staphylococcus aureus*, *Streptococcus pneumoniae*, and *Klebsiella pneumonia* .Treatment of necrotizing pneumonia consists of prolonged courses of antibiotics and possible life-saving surgery. However, massive necrotic tissue makes it difficult for the antibiotics to reach the infected areas, as well as leading to

the progressive destruction and persistent infection of the pulmonary parenchyma, possibly followed by the development of bronchopleural fistula, life-threatening hemoptysis, septicemia, and respiratory failure. Surgical treatment has been considered lifesaving in these cases.

Conclusion

Necrotizing pneumonia should be paid attention to the patient's with prolonged pneumonia disease course. Necrotizing pneumonia tends to occur in adult males with concomitant medical illness such as diabetes mellitus, alcohol abuse, and corticosteroid therapy. In our patient, the habit of smoking and use of inhaled corticosteroids may predispose the patient contratcting the Pseudomonas infection.

病例報告 113_C81

嚼檳榔與可以被預防的高血鈣–病例報告

Betel Nut Chewing and Preventable Hypercalcemia – A Case Report

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Introduction

Betel nut, the fruit of the Areca catechu palm, is extensively consumed in Southeast Asia, especially in India, Taiwan, Pakistan, and Bangladesh. Betel quid, which consists of betel nuts wrapped in betel leaves and slaked lime (calcium hydroxide or calcium carbonate), is prevalent in Taiwan. Studies have associated betel nut chewing with milk-alkali syndrome, characterized by hypercalcemia, metabolic alkalosis, and acute kidney injury. We are going to present a case of excessive betel nut consumption, complicated by severe hypercalcemia.

Case Report

The patient is a 37-year-old male admitted via the emergency department for recurrent nausea, vomiting, and abdominal pain, accompanied by acute kidney injury (AKI). Laboratory tests revealed elevated creatinine (5.64 mg/dL), severe hypercalcemia (14.4 mg/dL), and metabolic alkalosis (pH 7.54, HCO3 = 39.3 mmol/L), with severe neurological symptoms rendering him intolerable to the condition. The patient had a history of chewing approximately 150 betel nuts per day for over 10 years, and unlike most users, he swallowed the chewed quid. Over the preceding months, he experienced diffuse body pain, weakness, decreased urine output, and constipation, leading to multiple hospitalizations for renal dysfunction and severe hypercalcemia.

During his hospitalization, laboratory tests confirmed normal intact parathyroid hormone (PTH) levels at 25.5 pg/mL, and no other causes for hypercalcemia were identified, as malignancy, infection, and drug-induced etiologies were ruled out. Complete cessation of betel nut use was recommended; however, the patient refused due to his dependency on betel nuts for maintaining both his work capacity and emotional well-being. Therefore, we advised at least a reduction in the number of betel nuts consumed daily. The final acceptable dose for the patient, while maintaining calcium levels within the normal range, was 120 nuts per day, with the chewed quid being spat out. Following these adjustments, the patient's hypercalcemia-related neurological symptoms and abdominal discomfort improved significantly, and he no longer required frequent outpatient or emergency department visits for complications related to his previous hypercalcemia.

Discussion

Although only a small proportion of betel nut users develop symptomatic hypercalcemia, it is most common among those who swallow the chewed quid. In this case, the patient's estimated daily calcium intake of 210 mmol was significantly higher than the average 20 mmol seen in non-chewers. The excess calcium is likely absorbed in the lower gastrointestinal tract, impairing renal function and leading to a Bartter-like syndrome characterized by renal wasting of electrolytes and metabolic alkalosis. After reducing betel nut consumption and discontinuing quid swallowing, the patient's hypercalcemia improved significantly.

Conclusion

Betel nut chewing with slaked lime can cause hypercalcemia, metabolic alkalosis, and renal injury. While cessation is ideal, reducing intake and avoiding quid swallowing in dependent users can still lead to significant improvement. Early recognition and intervention are crucial to preventing renal damage and improving outcomes in excessive betel nut use.

病例報告 113_C82

久議仍重-腎臟移植患者因紅黴素與 Tacrolimus 藥物交互作用引起急性腎損傷的病例報告

An Ongoing Concern: Case Report of Acute Kidney Injury Due to Erythromycin and Tacrolimus Drug Interaction in a Kidney Transplant Patient

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Introduction

Kidney transplant patients require lifelong immunosuppressive therapy. Poor graft function often occurs early due to medication non-adherence and neglect of physician instructions. This can lead to rejection from low tacrolimus levels or increase the risk of infection and acute kidney injury from high levels. When treated by physicians unfamiliar with transplant medications, unconsidered drug interactions may cause unnecessary graft failure. We report a case of a patient 10 years post-transplant who developed acute kidney injury due to a drug interaction between erythromycin and tacrolimus.

Case Report

A 63-year-old female, 10 years after a living kidney transplant, presented to a local doctor with three days of upper respiratory infection symptoms without fever. She informed the physician of her kidney transplant status, and the physician prescribed empirical erythromycin, assuring her that it would not induce kidney injury and that she need not worry. After three days of erythromycin treatment, the patient noticed a decrease in urine output, bilateral pitting edema in her legs, and progressive fatigue. Approximately 12 hours later, she developed sudden and worsening right upper quadrant tenderness, prompting her to visit our emergency department (ED). On physical examination, she had a positive Murphy's sign. An abdominal computed tomography (CT) scan revealed gallbladder dilation with gallstones. Laboratory tests showed acute kidney injury (AKI) with a decline in glomerular filtration rate (GFR) to 41 mL/min/1.73m^2 from her baseline of 80-90 mL/min/1.73m^2. The patient underwent an urgent laparoscopic cholecystectomy. Tacrolimus levels were found to be elevated to 27 ng/mL, despite her self-discontinuation of the medication for one day, suggesting a drug-drug interaction between erythromycin and tacrolimus.
This interaction contributed to the patient's AKI, which was partially attributed to the acute cholecystitis and infection, and partly due to erythromycin-induced tacrolimus toxicity. After withholding tacrolimus for two days and restarting it once levels normalized, the patient's renal function recovered, with GFR returning to 101 mL/min/1.73m^2 at discharge.

Discussion

Calcineurin inhibitors (CNIs), such as cyclosporine and tacrolimus, are crucial immunosuppressants in transplantation but are susceptible to drug interactions, especially with macrolides. Tacrolimus is metabolized by CYP3A4 and P-glycoprotein, and inhibitors like erythromycin can disrupt this process, leading to complications. Erythromycin is listed in our renal transplant manual as an antibiotic to avoid, and its use requires consultation to adjust tacrolimus

dosing. In this case, the patient, reassured by her physician, overlooked this guidance, causing an avoidable complication. Fortunately, it was caught in time. This highlights the need for ongoing patient education, even years post-transplant.

Conclusion

This case underscores the importance of recognizing drug interactions in kidney transplant patients, particularly with erythromycin, which can alter tacrolimus metabolism. Even after years of transplant success, patients are at risk if immunosuppressive therapy is not properly managed, especially by non-transplant specialists. Ongoing education and reinforcement of medication guidelines are crucial to preventing avoidable complications like acute kidney injury. Clear communication between healthcare providers and transplant teams is essential for ensuring patient safety and long-term graft function.

病例報告 113_C83

肺癌免疫化療後持續性嗜中性球減少症：罕見併發症病例報告

Case report of prolong neutropenia after receiving immunochemotherapy in lung cancer: a rare complication

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Introduction

Persistent neutropenia following immunochemotherapy, accompanied the inability to recover blood cell growth, could cause severe complications. Management typically involves measures with growth factors like granulocyte colony-stimulating factors (G-CSF). Compared to cytotoxic chemotherapy, immunotherapy is believed better tolerated. However, the loss of self-tolerance can lead to immunee-related adverse events (irAE). The hematological toxicity, such as prolonged bone marrow suppression, is rare but fatal.

Case Report

We report a 68-year-old male with right lower lung squamous cell carcinoma (iT3N1M0) underwent two cycles of immunochemotherapy. The first cycle included Pembrolizumab (100 mg), Cisplatin (70 mg/m^2), and Docetaxel (60 mg/m^2), while the second cycle comprised Pembrolizumab (100 mg), Carboplatin (AUC: 5 mg/mL/min), and Docetaxel (60 mg/m^2). After the first cycle, the patient developed leukopenia, with a nadir white blood cell (WBC) count of 1,700/μL and absolute neutrophil count (ANC) of 307/μL. G-CSF with Filgrastim was administered, and the blood cell counts recovered within one week. However, following the second cycle, he presented severe neutropenia (WBC: 500/μL, ANC: 73/μL), anemia, fever, and progressive dyspnea, leading to hospitalization.

Empiric antibiotics and Filgrastim were initiated, but prolonged low blood cell count exceeds three weeks. He underwent a bone marrow biopsy, which section showed estimated marrow cellularity less than 1%. Normal trilineage hematopoietic cells nearly total absent. Also, blood smear showed decreased cellularity without tumor cells, suggesting therapy-related hypocellular marrow.

The patient further developed septic shock and respiratory failure, necessitating mechanical ventilation and stronger antibiotics. About neutropenia, Filgrastim was employed, but without improvement in WBC count. Shifting to Lenograstim, along with Danazol and Revolade for cytopenia, but remained ineffective. The patient’s condition worsened, culminating in multi-organ failure and death.

Discussion

Hematological irAE is a rare but serious side effect. This patient developed neutropenia after the first immunochemotherapy cycle, leading to a delay in the second cycle. While G-CSF treatment restored normal blood counts before the second cycle, the patient's weak bone marrow reserve remained an issue. Not only does chemotherapy affect bone marrow, but immunotherapy can worsen the condition by triggering immune responses that affect both cancer cells and healthy blood cells. In such cases, alternative treatments may be used.

Prophylactic measures, like the use of G-CSF, should be considered, particularly when low WBC counts occur without neutropenia. This approach helps prevent neutropenic fever and severe infections.

Conclusion

The case highlights the complexities of immunochemotherapy-induced neutropenia. Either immunotherapy or chemotherapy discontinuation should be considered, especially when prior neutropenia episode have occured. Close monitoring early signs of hematological irAE is crucial. The implementation of tailored strategies is essential.

病例報告 113_C84

克服免疫抑制患者中的急性呼吸窘迫症候群：葉克膜合併靜脈注射免疫球蛋白治療的病例報告

Overcoming Infection-induced ARDS in the Immunosuppressed Host by Combined ECMO and IVIG Therapy: a case report

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Introduction

Adenoviruses are non-enveloped DNA viruses most frequently associated with upper respiratory tract infections and less frequently pneumonia. Most illnesses are self-limiting, but fatal infection can occur in immunocompromised host. We report a 38-year-old male presenting with severe community-acquired pneumonia (CAP) and subsequent acute respiratory distress syndrome (ARDS). The patient had history of rheumatoid arthritis (RA), under chronic immunosuppressive therapy and was admitted with acute respiratory failure (ARF) requiring venovenous extracorporeal membrane oxygenation (VV-ECMO) support. Pathogens identified included adenovirus, coronavirus, and *Pneumocystis jirovecii* by multiplex PCR. The case highlights the complex management of infections, immunomodulation and ECMO.

Case Report

A 38-year-old male with known history of rheumatoid arthritis (on methotrexate, methylprednisolone, and leflunomide) presented to the emergency department with progressive dyspnea, fever, and hypoxia. He had productive cough for two days and dyspnea progressed. Laboratory results showed leukocytosis, elevated inflammatory markers, and severe hypoxemia. Chest x-ray showed bilateral lung opacities.

Due to ARF, he was intubated and later placed on VV-ECMO after failing maximal ventilatory support and prone positioning. PCR of respiratory aspirate with FilmArray Pneumonia Panel showed positive for adenovirus, coronavirus. Further pathogen survey also found for *P. jirovecii*. Immunosuppressive therapy was discontinued except methylprednisolone (MTP), and the patient as started on trimethoprim-sulfamethoxazole with MTP for *P. jirovecii* pneumonia (PJP), along with levofloxacin and ganciclovir. Intravenous immune globulin (IVIG) therapy of total 2g/kg separated in 5 days was given. The patient encountered VV-ECMO drainage deficiency despite adequate volume management, canulation transfer to VVV-ECMO on hospital day 5.

Complications included pneumomediastinum and liver dysfunction, likely secondary to ventilator-induced lung injury, ECMO-related perfusion issues, and prolonged antimicrobial therapy. Liver function tests showed elevated ALT and bilirubin, managed with supportive care.

After 10 days of ECMO support, ECMO was successfully weaned. In the following week, the patient was successfully extubated. He continued improving with physical rehabilitation.

Discussion

This case demonstrates the challenges of managing ARDS follow co-infection of *P. jirovecii* and viral pneumonia in an immunocompromised patient. Immunosuppression from underling disease and the therapeutic regimen likely predisposed the patient to the development of multiple

opportunistic infections. The initiation and subsequent cannulation modifying of ECMO was crucial in maintaining adequate oxygenation, serving as a bridge to infection control and lung recovery.

The use of IVIG in this case was a key adjunctive therapy, given the complexity of the patient's condition, particularly his immunosuppressed state and the presence of multiple pathogens. As cidofovir was not available, we also administered ganciclovir, which showed in vitro & in vivo activity against adenovirus, in this patient. Both IVIG and ganciclovir might be crucial in stabilizing the patient's immune response during a critical phase of ARDS and overwhelming infection.

Conclusion

This case highlights the critical role of early recognition, aggressive infection control, and the use of advanced life support measures such as ECMO in managing ARDS in immunocompromised patients. The successful outcome in this patient, despite multiple infections and organ dysfunction, emphasizes the need for a tailored and multidisciplinary approach.

病例報告 113_C85

同步發生的膽囊癌與肝細胞癌：一例罕見病例報告

Synchronous Gallbladder Carcinoma and Hepatocellular Carcinoma: A Rare Case Presentation

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Introduction

Synchronous malignancies are rare and co-occurrence of GBC and HCC is particularly uncommon. HCC typically arises in chronic liver disease settings, while GBC often presents incidentally at advanced stages. This case illustrates a case of synchronous HCC and GBC patient treated with TACE and systemic treatment, then managed surgically after successful downstaging of HCC.

Case Report

An 81-year-old male with chronic hepatitis C-related liver cirrhosis, previously treated in 2007, presented with fatigue for days. He had a past surgical history of partial gastrectomy for gastrointestinal stromal tumor (GIST) in 2009.

Imaging revealed a 6.0 cm mass in the left hepatic lobe, and a 2.2 cm polypoid gallbladder lesion. CT showed early enhancement and washout in the hepatic lesion, suggestive of HCC, with a portal vein thrombus. Laboratory tests indicated an elevated alpha-fetoprotein (AFP) level of 16,886 ng/mL and a PIVKA-II of 871.90 mAU/mL. The patient was classified as Barcelona Clinic Liver Cancer (BCLC) stage C HCC.

The patient underwent three sessions of TACE and was started on sorafenib in November 2022. Follow-up imaging showed significant downstaging, with almost complete remission of lung metastases. In April 2024, he went through a left hepatectomy and cholecystectomy. Histopathology confirmed moderately differentiated adenocarcinoma of the gallbladder and extensive necrosis in the liver with no residual HCC. The patient recovered well and remains in remission, with further adjuvant therapy for gallbladder carcinoma under consideration.

Discussion

Synchronous hepatocellular carcinoma (HCC) and gallbladder carcinoma (GBC) are exceedingly rare and present significant management challenges, necessitating a multidisciplinary approach. Literature reviews have reported only 16 cases of synchronous HCC and GBC as of 2020.

Sorafenib, a multitarget tyrosine kinase inhibitor, has shown survival benefits in advanced HCC. Further review showed particular more benefits in hepatitis C virus-related advanced HCC. In this case, the patient's positive response to sorafenib and transarterial chemoembolization (TACE) facilitated effective tumor downstaging, allowing for successful surgical resection of both tumors.

Conclusion

Synchronous GBC and HCC present significant diagnostic and therapeutic challenges. This case underscores the importance of multidisciplinary management, early diagnosis, and individualized treatment. The combination of TACE and sorafenib facilitated successful tumor downstaging, allowing for curative resection. The patient remains in remission, highlighting the value of early

intervention in managing synchronous malignancies.

病例報告 113_C86

隱藏的致命威脅：血管內動脈瘤修復術後的主動脈腸道瘻管

Silent Threat: Aortoenteric Fistula Unveiled Two Years After EVAR

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Introduction

Aortoenteric fistula (AEF) is a rare but life-threatening condition characterized by abnormal connection between the aorta and the gastrointestinal (GI) tract. Primary AEFs, which occur between the native aorta and the GI tract are rare (0.04%-0.07%). Secondary AEFs(0.77%-4.8%) develop between an aortic prosthetic graft and GI tract, following vascular reconstruction procedures. This report presents secondary AEF in patient with history of mycotic aneurysm with endovascular aneurysm repair (EVAR), developed AEF two years post-surgery.

Case Report

A 66-year-old male, history of mycotic aneurysm and rupture underwent EVAR and femoral-femoral bypass three years prior. He presented to hospital with three-month intermittent fever,anemia with hemoglobin level:11 g/dL(baseline:15). hsCRP level:11 mg/L. Blood cultures grew *Escherichia coli* (E. coli).

Abdominal computed tomography (CT) revealed ruptured abdominal aortic aneurysm following aorto-uni-iliac (AUI) procedure, with inflammation and abscess formation around bifurcation of aorta and right common iliac artery. Deep enteroscopy identified duodenal fistula with exposed stent in third portion of duodenum.Operation finding: AEF between graft and duodenum, and perforation of third and fourth portions of duodenum. Opearation method 1. resection of the AEF with direct reconstruction with Y graft 2. Resection of perforated duodenum 3. Side-to-side duodenojejunostomy 4. Feeding jejunostomy tube. Intraoperative cultures *E. coli*. Ertapenem and Teicoplanin was used, and responded well.

Discussion

AEF is a rare but severe complication following aortic reconstructive surgery. AEF can present with minor GI bleeding, signs of sepsis, abdominal pain, and hemodynamic instability. Prompt diagnosis is crucial, as delays can lead to catastrophic outcomes.

Diagnostic imaging, particularly CT-angiography is essential for identifying AEFs include loss of normal tissue planes between aorta and GI structures, presence of air or fluid in aortic sac, and active contrast extravasation in rare cases. EGD can help diagnose proximal fistulas but may exacerbate bleeding in unstable patients.

Management involves either endovascular or open surgical repair, depending on patient's condition. EVAR deploy in unstable patients, is not definitive for AEF due to association with higher rates of recurrent bleeding, infection, and re-intervention. Open surgical repair is preferred when infection or extensive graft involvement is present. In severe infection cases, extra-anatomic bypass with graft excision is recommended.

In this case, patient developed AEF associated with duodenal perforation two years after EVAR.

Surgical intervention and following antibiotic therapy, led to successful outcome.

Conclusion

Secondary AEF is a rare but life-threatening complication requiring prompt diagnosis and treatment. Physician maintain suspicion in patients with unexplained GI bleeding, anemia, or sepsis, especially with history of EVAR. Early recognition and intervention are crucial for improving outcomes. This case highlights the importance of a multidisciplinary approach to the management of secondary AEF, incorporating advanced imaging, surgical intervention, and antibiotic therapy.

病例報告 113_C87

新冠肺炎下的溶血風暴，極端數值的自體免疫性溶血

COVID-19 Triggered a Hemolytic Storm: Battling Refractory AIHA with Sky-High Bilirubin Levels

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Introduction

Warm autoimmune hemolytic anemia (AIHA) is a severe and potentially fatal condition, and COVID-19 is known to manifest with various extrapulmonary complications. When faced with a patient exhibiting extremely high bilirubin levels (78 mg/dL), it becomes essential to analyze the case comprehensively and consider all contributing factors. In this report, we present a case of refractory AIHA in a patient who initially presented with a COVID-19 infection.

Case Report

This is a 73-year-old woman with a history of autoimmune hemolytic anemia (AIHA) presented in August 2023 with general malaise, dyspnea on exertion, dark urine, and jaundice. Initial evaluations revealed anemia (hemoglobin around 7.8–8.4 g/dL), elevated bilirubin (5-9 mg/dL), low haptoglobin, positive direct antiglobulin test, and positive ANA titers (1:160 homogeneous, 1:320 speckled). Abdominal CT scans showed lymphadenopathies near the splenic hilum and para-aortic area, and a sacral osteolytic lesion; however, lymph node biopsy indicated reactive hyperplasia without malignancy. She was then treated as idiopathic warm AIHA with methylprednisolone and discharged on oral medications at outpatient department.

In June 2024, she developed fever and weakness, tested positive for COVID-19, and was readmitted with severe anemia (Hb 4.4 g/dL) and elevated bilirubin (18.7 mg/dL). Despite blood transfusions and corticosteroid therapy, her hemolysis worsened (bilirubin up to 78 mg/dL). She received intravenous immunoglobulin, plasma exchange, and rituximab for refractory AIHA and three course of remdesivir treatment for COVID-19. Imaging revealed splenomegaly and gallstones, leading to percutaneous transhepatic gallbladder drainage and later ERCP with stenting for suspected cholecystitis and cholangitis whereas high bilirubin still noted. Due to persistent fever, splenomegaly, cytopenias, hypertriglyceridemia, and elevated ferritin, hemophagocytic lymphohistiocytosis (HLH) was suspected; a bone marrow biopsy was performed and finally confirmed, and low dose etoposide (50mg weekly for 2 doses) was administered. Throughout her hospitalization, she experienced recurrent gastrointestinal bleeding; colonoscopy attempts were made, and bleeding ceased. After three dosage of rituximab, the hemolysis got more stable (higher Hb level around 7.5 and lower bilirubin around 20 mg/dL), then she was discharged and arranged surveillance at out patient department.

Discussion

Autoimmune hemolytic anemia (AIHA) involves the destruction of red blood cells by autoantibodies, leading to anemia and elevated bilirubin levels. In this patient, the onset of severe hemolysis coincided with a COVID-19 infection, suggesting a correlation between the two conditions. COVID-19 has been reported to trigger or exacerbate autoimmune responses due to

immune system dysregulation caused by the virus. Proposed mechanisms include cytokine storms inducing autoantibody production, molecular mimicry where viral antigens resemble self-antigens, and bystander activation of autoreactive B cells [1]. The significant drop in hemoglobin and rise in bilirubin levels in this patient highlight how COVID-19 can intensify hemolytic processes in AIHA patients.

Managing refractory AIHA, especially when complicated by COVID-19, requires a multimodal approach. First-line treatments like corticosteroids may be insufficient, necessitating additional therapies. In this case, the patient received intravenous immunoglobulin (IVIg) to modulate the immune response [2] and plasma exchange to remove circulating autoantibodies [2]. Rituximab, an anti-CD20 monoclonal antibody, was administered to deplete B cells responsible for producing autoantibodies [3] and, is currently the standard treatment for steroid-refractory AIHA. The development of hemophagocytic lymphohistiocytosis (HLH), a severe hyperinflammatory syndrome, further complicated the clinical picture, requiring treatment with etoposide . These interventions eventually stabilized her hemoglobin levels and reduced hemolysis.

This case underscores the complex interplay between COVID-19 and autoimmune conditions like AIHA, and, other than B cell system, HLH. It highlights the importance of recognizing potential hematological complications in COVID-19 patients and suggests that aggressive, combined therapeutic strategies may be necessary for managing refractory cases. Early identification and treatment are crucial for improving outcomes in patients where standard therapies fail.

Conclusion

In the COVID-19 era, given the diverse and significant extrapulmonary manifestations of the virus, it is crucial to adopt a multidisciplinary approach to patient care, ensuring that every critical detail is thoroughly addressed for each individual case.

病例報告 113_C88

揭密吉舒達的隱憂：甲狀腺低下誘發橫紋肌溶解的肺癌個案呈現

Unveiling Pembrolizumab's Hidden Risk: Hypothyroidism-Induced Rhabdomyolysis in a Lung Cancer Patient

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Introduction

In the past, rhabdomyolysis of pembrolizumab induced hypothyroidism was less reported. We will report a lung cancer patient who developed pembrolizumab-induced hypothyroidism leading to rhabdomyolysis and acute kidney injury, highlighting the need for vigilant thyroid monitoring during immune checkpoint inhibitor therapy.

Case Report

This is a 57-year-old male with no significant medical history presented with several months of hemoptysis. Chest CT scans revealed a large mass in the right upper lobe, and a biopsy confirmed stage IIIA adenocarcinoma. He underwent a right upper lobe (RUL) lung lobectomy with lymph node dissection, followed by adjuvant chemotherapy (cisplatin plus vinorelbine) and four cycles of adjuvant pembrolizumab. In March 2024, he developed myalgia but denied any immobility or bedridden status. Laboratory tests showed elevated creatine phosphokinase (CPK) levels at 1496 U/L and increased creatinine levels, leading to a diagnosis of rhabdomyolysis and acute kidney injury (AKI). While providing adequate fluid resuscitation, we investigated the cause of the rhabdomyolysis. Thyroid function tests revealed low free T4 (0.48 ng/dL) and high TSH (147.878 μIU/mL), indicating hypothyroidism. Levothyroxine therapy was initiated at 100 mcg per day without the use of steroids. The patient achieved euthyroid status, accompanied by decreasing CPK levels and improved renal function. His myalgia significantly improved, and he was discharged. He transitioned to alternative targeted therapy and continued regular follow-up at our outpatient department.

Discussion

Pembrolizumab, a programmed death-1 (PD-1) inhibitor, is widely used in treating advanced non-small cell lung cancer due to its efficacy in enhancing anti-tumor immune responses. However, it is associated with immune-related adverse events (irAEs), including endocrine disorders like hypothyroidism. In this case, a 57-year-old male developed rhabdomyolysis secondary to pembrolizumab-induced hypothyroidism, evidenced by elevated creatine phosphokinase (CPK) levels and acute kidney injury (AKI). Hypothyroidism can lead to metabolic disturbances in muscle tissue, resulting in muscle breakdown and subsequent rhabdomyolysis [1] [2].

Differentiating between rhabdomyolysis caused by hypothyroidism and myositis as an irAE is crucial for appropriate management. Myositis involves immune-mediated muscle inflammation, presenting with muscle weakness, elevated CPK, and sometimes respiratory muscle involvement [3]. In contrast, hypothyroid-induced rhabdomyolysis stems from metabolic muscle dysfunction without significant inflammation. The patient's lab results showing high thyroid-stimulating

hormone (TSH) and low free thyroxine (T4) levels point toward hypothyroidism rather than myositis. The improvement of symptoms and normalization of CPK levels after initiating levothyroxine therapy further support this diagnosis according the past case report. [4] [5].
Management of such cases involves prompt recognition and treatment of hypothyroidism to restore euthyroid status, which can reverse muscle symptoms and prevent renal complications. Regular monitoring of thyroid function tests in patients receiving immune checkpoint inhibitors like pembrolizumab is essential for early detection of thyroid dysfunction. A multidisciplinary approach, including oncologists and endocrinologists, enhances patient care. Educating patients about the signs and symptoms of hypothyroidism and myositis can lead to timely reporting and intervention, improving clinical outcomes.

Conclusion

Clinicians involved in administering immune checkpoint inhibitors (ICIs) need to recognize that hypothyroidism induced by these treatments can lead to rhabdomyolysis. Early detection and prompt initiation of levothyroxine therapy can reverse hypothyroidism, thereby reducing muscle breakdown and improving renal function.

病例報告 113_C89

意想不到的不典型水腦症診斷個案，以發燒為初始表現

Unmasking Hydrocephalus: A Fever, Seizure, and Gait Crisis Leads to an Unanticipated Diagnosis

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Introduction

Normal pressure hydrocephalus (NPH) is a challenging and vicious condition that can severely impact patients' quality of life. Differentiating NPH from other neurological disorders is particularly difficult when the classic symptoms do not fully present. Here, we discuss a case of fever and unsteady gait, ultimately diagnosed as NPH.

Case Report

This is a 54-year-old female with a past medical history of type 2 diabetes mellitus, hypertension, and a ruptured Acom aneurysm treated with clipping complicated with hydrocephalus status post external lumbar cisternal drainage, was admitted to the hospital due to general weakness and fever up to 39.6°C for two days. She also reported a fall at the onset of her symptoms. She denied respiratory, gastrointestinal, or urinary tract symptoms. Upon arrival, her vital signs showed hypertension (BP 165/129 mmHg), tachycardia, and a Glasgow Coma Scale score of E4V1M5. A brain CT showed post-surgical changes and encephalomalacia in the frontal lobes. Chest CT revealed mild bronchial dilation, atelectasis, and pleural thickening, while abdominal CT was unremarkable. Lab data showed hypernatremia, elevated myoglobin, and cardiac enzymes. Urine culture grew *Klebsiella pneumoniae*.

Given the fever of unknown origin, she was admitted for further evaluation. Despite initial treatment with antipyretics, fluids, and bromocriptine, persistent fever prompted further investigations, including osteomyelitis scan and brain MRI, which were non-contributory. Amantadine was added for suspected central fever. Her fever subsided on day 7 of admission after amantadine use and the lumbar puncture procedure, accompanying with the improvement of the unsteady gaits. After discussion with the neural surgeon due to the suspicion of normal pressure hydrocephalus, she underwent a ventriculoperitoneal (V-P) shunt procedure on June 15, 2024. Cerebrospinal fluid (CSF) cultures revealed Vancomycin-resistant *Enterococcus faecium* (VRE), for which she received antibiotic treatment. She experienced a focal seizure during her hospital stay, but brain imaging revealed no acute pathological change, and antiepileptic medications were started. After stabilization, she was discharged with instructions for follow-up. At discharge, her fever had resolved, her blood sugar was controlled, and she had steady gait and returned to her baseline activity.

Discussion

This case highlights the complexity of diagnosing and managing a patient with NPH and neurological symptoms alongside fever. NPH is classically characterized by the triad of gait disturbance, urinary incontinence, and cognitive impairment, often summarized as "wet, wobbly, and wacky". (1) This patient presented with unsteady gait, a key component of NPH, which

improved following lumbar puncture, supporting the diagnosis. A definitive diagnosis of NPH typically involves imaging such as brain MRI, which often reveals ventricular enlargement without corresponding cortical atrophy, and clinical response to CSF drainage, either through repeated lumbar punctures or temporary CSF shunting.

Surgical intervention with a V-P shunt is the standard treatment for NPH, aimed at reducing the excess CSF and relieving pressure on the brain. (2) However, complications such as shunt infections can occur, as evidenced by this patient's positive CSF culture for *Enterococcus faecium* (VRE). It also alerts us the importance of close monitoring following neurosurgical procedures and the sterile procedure. (3)

The co-occurrence of fever and neurological signs raises concerns for central nervous system (CNS) infection, but other diagnoses must be considered. In this case, the patient's fever persisted despite an initial course of antibiotics and antipyretics, prompting further investigation. Fever with neurological signs may arise from systemic infections, metabolic disturbances, drug reactions, or central fever caused by damage to temperature-regulating centers in the hypothalamus.

This case underscores the complexity of diagnosing and managing fever in patients with neurological conditions. While CNS infection is a critical consideration, other causes, including structural, metabolic or medication-induced fevers, must also be considered, particularly in patients with multiple comorbidities and history of brain damage with neurosurgical interventions. (4)

Conclusion

This case emphasized the importance of considering multiple diagnoses when encountering fever with neurological signs other than CNS infection.

病例報告 113_C90

病房裡的無聲殺手：小腸淋巴瘤與嚴重出血的個案呈現

A Silent Killer: Unmasking Small Bowel Lymphoma with Deadly GI Bleeding and Lethal Complications

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Introduction

This report details a 70-year-old female with end-stage renal disease, liver cirrhosis, and gastrointestinal bleeding, who was later diagnosed with diffuse large B-cell lymphoma (DLBCL) in the small bowel, highlighting the complexity and challenges of diagnosis and management

Case Report

This case involves a 70-year-old female with a medical history of end-stage renal disease (ESRD) under peritoneal dialysis (P/D) and liver cirrhosis with splenomegaly. She was admitted to our hospital through the emergency department after experiencing tarry stools for 10 days.

The patient had been discharged from our nephrology ward one month prior, where she was treated for urosepsis. However, this time, she noticed tarry stools at home, prompting her visit to the emergency department. Initial lab results revealed anemia, hyponatremia, and hypokalemia. Given these findings, she was admitted to the gastroenterology ward for further evaluation and management.

Both an esophagogastroduodenoscopy (EGD) and colonoscopy were performed, but no active bleeding was detected. An abdominal computer tomography scan revealed a mass near the para-aortic and para-jejunum regions, along with segmental bowel wall thickening and mesenteric stranding. Concerned about the possibility of small bowel malignancy, a double balloon enteroscopy was performed. This procedure identified lymphangiectasia in the proximal jejunum, and a biopsy was taken. The pathology report confirmed lymphoma. Immunohistochemistry (IHC) analysis showed CD20 (diffuse positive), Bcl-2 (positive, 20%), MUM-1 (diffuse positive), and c-Myc (positive, 20%), consistent with a diagnosis of diffuse large B-cell lymphoma (DLBCL).

After consulting with the oncology team, the patient was started on the R-CHOP chemotherapy regimen. However, due to ECOG (Eastern Cooperative Oncology Group) performance status 3 and tarry stool again, she was switched to a modified regimen, R-mini-CHOP. However, she was too weak to undergo further chemotherpay, patient and family choice to hospice care after share decision making. The patient expired in December, 2023.

Discussion

Diffuse large B-cell lymphoma (DLBCL) is the most common subtype of non-Hodgkin lymphoma, but its occurrence in the small bowel is relatively rare, accounting for approximately 10-30% of gastrointestinal (GI) lymphomas. The clinical presentation of small bowel DLBCL is often nonspecific, with symptoms such as abdominal pain, weight loss, and fatigue. However, more acute presentations, such as bowel obstruction, perforation, and gastrointestinal bleeding, are also reported. Notably, gastrointestinal bleeding is a significant complication in small bowel

DLBCL, occurring in about 10-20% of cases. This bleeding can be severe, leading to anemia and requiring urgent intervention, including endoscopic or surgical procedures. The rarity of this disease and its variable presentation often lead to delays in diagnosis, emphasizing the need for a high index of suspicion in patients with unexplained GI symptoms and anemia.

Imaging, particularly abdominal computed tomography (CT), plays a crucial role in the diagnosis and evaluation of small bowel DLBCL. The typical CT findings include segmental or diffuse bowel wall thickening, which may appear homogeneous or heterogeneous. A mass effect may also be observed, sometimes associated with mesenteric stranding and enlarged lymph nodes. These imaging characteristics can mimic other conditions, such as Crohn's disease or adenocarcinoma, necessitating careful correlation with clinical and endoscopic findings. Contrast-enhanced CT is particularly valuable in identifying complications such as perforation or active bleeding, which are critical in the acute management of these patients. Given the potential for significant overlap with other small bowel pathologies, radiologists and clinicians must collaborate closely to ensure accurate diagnosis and appropriate management.

The outcome of patients with small bowel DLBCL largely depends on the stage at diagnosis, the patient's overall health, and the treatment regimen. The standard treatment for DLBCL is the R-CHOP regimen (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone), which has been shown to be effective in achieving remission in many cases. However, in older or frail patients, just like our case, a modified regimen known as R-miniCHOP is often used to reduce toxicity while maintaining efficacy. Despite the effectiveness of these treatments, rebleeding can occur, particularly in cases where the lymphoma has caused significant mucosal disruption or ulceration. The rebleeding rate after R-CHOP or R-miniCHOP varies, with some studies reporting rates as high as 15-20%, particularly in patients who had significant bleeding at presentation. Close monitoring and supportive care, including the potential need for repeat endoscopy or even surgical intervention, are essential for managing these patients during and after treatment.

Conclusion

Small bowel DLBCL is a rare, aggressive malignancy with significant diagnostic challenges. Despite chemotherapy, frailty may limit treatment options, underscoring the importance of timely diagnosis and individualized care.

病例報告 113_C91

一位肝移植患者因播散性結核產生嚴重併發症與其治療的困境

Disseminated Tuberculosis in a Liver Transplant Patient: A Case of Severe Complications and Management Challenges

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Introduction

Disseminated tuberculosis (TB) occurs when Mycobacterium tuberculosis spreads beyond the lungs to multiple organs, posing a significant challenge, especially in immunocompromised patients. This case report presents a 63-year-old woman with a history of liver cirrhosis secondary to hepatitis C virus (HCV) infection, who developed disseminated TB after liver transplantation, resulting in severe complications.

Case Report

A 63-year-old female with a history of HCV-related liver cirrhosis post-living donor liver transplantation, on regular follow-up under immunosuppression, presented with worsening upper abdominal pain, dysphagia, mild fever, and significant weight loss (8 kg in 2 months). She also had a chronic cough for several months. Physical examination revealed a soft abdomen with a 4.6 cm protruding mass in the periumbilical region. Laboratory results showed leukocytosis (17,700/μL) with neutrophil predominance (86.6%) and elevated C-reactive protein (16.45 mg/dL). Chest CT revealed multiple nodules with cavitation in both lungs. Acid-fast bacilli were detected in sputum, stool, and urine, confirming disseminated tuberculosis (TB). Despite anti-TB therapy (HERZ), her condition worsened, complicated by respiratory distress and the development of a tracheoesophageal fistula (TEF), which required stent placement. She experienced recurrent infections, uncontrolled gastrointestinal bleeding, and required bowel resection. Tracheostomy, esophagectomy, and feeding jejunostomy were performed due to prolonged intubation and nutritional needs. Biopsies from the trachea and gastrointestinal tract confirmed TB infection. The periumbilical mass evolved into an abscess, necessitating debridement surgery, also confirming TB. After prolonged treatment, the patient was discharged in improved condition.

Discussion

Disseminated TB is a rare but serious condition in immunocompromised patients, such as organ transplant recipients. In this patient, the development of TEF and refractory gastrointestinal bleeding, a complication of TB, illustrates the destructive potential of untreated or poorly controlled disseminated infections. It can cause significant morbidity, and its management in the presence of active TB poses challenges, as surgical interventions carry increased risk in such patients. The patient's immunosuppressed state due to liver transplantation likely contributed to the widespread dissemination of TB. Early recognition and treatment are critical in preventing complications, but even with prompt intervention, the management of disseminated TB remains difficult.

Conclusion

Disseminated TB in liver transplant patients can lead to severe complications. This case underscores the importance of early diagnosis and treatment in immunocompromised patients and highlights the complexity of managing disseminated TB, which requires a coordinated multidisciplinary approach.

病例報告 113_C92

以菌血症、肺及前列腺膿瘍表現的類鼻疽 -- 一例病例報告

A case report of melioidosis, with bacteremia, lung and prostate abscess

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Introduction

Melioidosis, caused by the bacterium *Burkholderia pseudomallei*, is a zoonotic disease primarily found in tropical and subtropical regions. In Taiwan, cases mainly arise in Central and Southern areas. Classified as a notifiable disease category IV, the disease manifests in various forms, from acute to latent infections, and from sepsis to localized abscess. Here, we present a case of melioidosis characterized by a bloodstream infection, lung abscess, and prostatic abscess.

Case Report

A 73-year-old man presented to our emergency department with a four-day-long fever. His medical history included extranodal marginal zone B-cell lymphoma of the nasopharynx, treated with radiotherapy 15 years earlier, and an infrarenal abdominal aortic aneurysm with stenting two years ago.

The patient reported fever with chills, accompanied by dysuria, urinary frequency, and difficulty in bladder emptying. He denied dyspnea, chest pain, cough, diarrhea, nausea, or vomiting. Upon examination, he was alert but febrile, and with no visible open wounds. Laboratory tests showed leukocytosis with a left shift and elevated hsCRP. Urinalysis showed pyuria with positive leukocyte esterase. His chest X-ray appeared normal, but a chest CT scan revealed a 1.5 cm hypodense lesion with rim enhancement in the right middle lung, suggestive of abscess formation.

Blood cultures confirmed *Burkholderia pseudomallei*, prompting initial treatment with ceftazidime. However, due to persistent fever, meropenem was prescribed instead four days later. A PET scan indicated uptake in the prostate and right middle lung, suggesting disseminated disease. After consulting an infectious disease specialist, levofloxacin was added. A transrectal prostate ultrasound confirmed a 2.5 cm abscess at the left apex of the prostate, which was subsequently drained percutaneously, and cultures also identified *B. pseudomallei*. Following the drainage, his fever subsided, and further blood cultures returned negative. After a 29-day hospitalization, he was discharged on oral trimethoprim-sulfamethoxazole and levofloxacin.

Discussion

B. pseudomallei is a Gram-negative bacterium found in soil and surface groundwater in tropical and subtropical areas, transmitted through direct contact with contaminated sources or by inhaling contaminated dust. In Taiwan, the highest isolation rates occur in area including Ren-Der (Tainan) and Feng-Yuan (Taichung). Outbreaks typically occur after typhoon or heavy rainfalls. Risk factors for melioidosis include diabetes, alcoholism, older age, and chronic liver, lung, or renal disease. The manifestation of melioidosis varies from minimal symptoms to life-threatening sepsis and pneumonia. Diagnosis relies on culture data, and treatment involves an initial intensive phase of intravenous antibiotics, followed by long-term eradication therapy. Surgical drainage may be

necessary for some abscesses.
In this case, the patient resided in Beitun, Taichung, close to Feng-Yuan, known for high *B. pseudomallei* isolation rate. His habit of walking barefoot in parks increased his exposure risk. While the exact timing of infection was uncertain, a further survey revealed underlying diabetes mellitus, which might contribute to his susceptibility. After identification of bacteremia, assessments for disseminated disease were arranged, and surgical drainage was completed to achieve eradication.

Conclusion

Melioidosis is a serious but treatable condition when diagnosed promptly and accurately. Healthcare professionals must remain vigilant regarding the potential for *B. pseudomallei* infections, considering the disease's varied presentations.

病例報告 113_C93

Osimiertinib 在肺癌患者所引發之皮膚紅斑-病例報告及文獻回顧

Osimertinib Associated Erythema in a Lung Cancer Patient Having EGFR Mutation – Case Report and Review of Literature

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Introduction

Osimertinib, a third-generation EGFR-TKI, is used to treat non-small cell lung cancer (NSCLC) patients with specific EGFR mutations. Dermatologic adverse events (dAEs), including skin rashes, are common side effects associated with osimertinib, though most dAEs are not severe. In this report, we present a 57-year-old woman with stage IV lung adenocarcinoma who received osimertinib as first-line treatment and developed generalized erythema after one month of therapy.

Case Report

A 57-year-old woman initially presented with persistent dizziness for one month and was diagnosed with lung adenocarcinoma with brain metastasis. She underwent surgery to remove the brain tumors, and pathological examination confirmed metastatic adenocarcinoma of lung origin. Genetic testing of the tumor cells revealed an exon 19 deletion.

The patient began treatment with the EGFR-TKI osimertinib as first-line therapy. One month later, the patient developed pruritic erythema and vesicles on the trunk and all four limbs, sparing the oral mucosa, involving 10% to 30% of the body surface area (BSA). She was prescribed oral prednisolone at a dosage of 10 mg per day for two weeks, along with oral doxycycline and topical steroid cream. Despite this treatment, the skin rash progressed, affecting over 30% of the BSA, and the patient developed generalized edema and new flaccid blisters on her hands. Osimertinib was discontinued, and the prednisolone dosage was increased to 50 mg per day for seven days. A skin biopsy was performed, revealing lichenoid perivascular infiltration of lymphocytes, eosinophils, and neutrophils in the upper dermis, along with mild melanin incontinence, consistent with a lichenoid drug reaction. No definite microorganisms were identified, and there was no significant immunodeposition of IgG, IgM, IgA, or C3.

After the patient's condition improved, afatinib was introduced, and prednisolone was gradually tapered to 30 mg/day, then 15 mg/day before being discontinued. No new skin lesions developed.

Discussion

Three other cases of severe dAEs related to osimertinib (Stevens-Johnson syndrome or toxic epidermal necrolysis) have been reported previously—one from Japan[1] and two from Mainland China[2,3]. The onset of severe dAEs ranged from two to five weeks after initiating osimertinib therapy. In these studies, most patients initially received topical steroids for osimertinib-related dAEs. When the condition progressed to SJS/TEN, osimertinib was discontinued, and systemic steroids, with or without intravenous immunoglobulin (IVIG), were administered. However, these studies did not mention whether osimertinib was restarted after recovery from dAEs.

In our case, after the patient's dAEs were well controlled, we restarted EGFR-TKI therapy with afatinib for two reasons. First, the patient did not carry the T790M mutation, suggesting that afatinib could be effective. Additionally, it is believed that the likelihood of dAE recurrence is lower when restarting with a different medication.

Conclusion

The management of severe dermatologic adverse events (dAEs) associated with EGFR-TKIs is still largely approached on a case-by-case basis. We present a case of severe dAEs resulting from osimertinib, which was successfully treated with systemic steroids, followed by switching to afatinib without recurrence. Further studies are needed to assess the safety of restarting EGFR-TKI therapy after severe dAEs.

病例報告 113_C94

以嚴重貧血為初始表現的多發性骨髓瘤之偶然發現：病例報告

Incidental Discovery of Multiple Myeloma in a Patient Presenting with Severe Anemia: A Case Report

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Introduction

Anemia is a prevalent condition that necessitates thorough evaluation to determine its cause. Although anemia is common, the role of monoclonal gammopathy in its etiology is often overlooked. Monoclonal gammopathy, including conditions like monoclonal gammopathy of undetermined significance (MGUS) and multiple myeloma, involves the proliferation of abnormal plasma cells producing monoclonal proteins. While MGUS is generally benign, it can progress to more serious conditions such as multiple myeloma, which is often associated with anemia. Despite advancements in understanding these conditions, challenges remain in early detection and differentiation of monoclonal gammopathy-related anemia from other types of anemia. Recent studies emphasize the need for improved screening strategies to identify patients at risk for progression from MGUS to MM. However, gaps persist in the field regarding the optimal diagnosing strategy of anemia specifically caused by monoclonal gammopathy. This case report presents a 68-year-old female with severe anemia as an initial presentation, leading to the incidental discovery of MM.

Case Report

A 68-year-old retired caterer with no significant medical history presented to the emergency department with shortness of breath lasting one week. Her symptoms did not improve with rest, and she denied chest pain, decreased urine output, or other systemic symptoms. Upon evaluation, she had severe anemia and elevated NT-proBNP levels. After initial treatment and discharge, she returned with worsening symptoms, including orthopnea and reduced urine output. A chest radiograph showed cardiomegaly with bilateral hilar infiltration. Physical examination revealed pale conjunctiva, crackles, and lower limb edema. Laboratory tests indicated macrocytic anemia with vitamin B12 deficiency and indirect hyperbilirubinemia. Hemolytic anemia was less likely through related negative lab tests. Endoscopy revealed gastric ulcers without active bleeding. Echocardiography showed preserved cardiac function. Due to suspected renal involvement and fair cardiac systolic function, serum immunoglobulins were tested, revealing elevated IgG and an abnormal kappa/Lambda ratio. We consulted an oncologist, and a subsequent bone marrow biopsy confirmed MM.

Discussion

This case report describes a 68-year-old female whose severe anemia led to the incidental diagnosis of MM. Key findings include her initial symptoms of shortness of breath and anemia, elevated IgG levels, an abnormal kappa/lambda ratio, and confirmation of MM via bone marrow

biopsy. However, we seldom investigate hematologic malignancy as an initial anemia workup. Anemia in MM is caused by malignant plasma cell infiltration in the bone marrow and depletion in EPO synthesis and is related to elevated cytokines, including IL-1, TNF-alpha, and IFN-gamma, resulting in the reduction of erythropoiesis. In addition, anemia is common in MM, affecting over two-thirds of patients. This case highlights how severe anemia can be an initial presentation of multiple myeloma, which is often overlooked in differential diagnoses. Our case report is limited because it is based on a single case observation, which may not be representative of all patients.

Conclusion

MM can present with normocytic anemia due to bone marrow infiltration by malignant plasma cells and cytokine-mediated erythropoiesis suppression. This case underscores the need for clinicians to consider hematological malignancies in atypical presentations of anemia for timely diagnosis and management.

病例報告 113_C95

29 歲女性因腹痛和反覆手部水泡-一罕見的病例報告

A case report of a 29-year-old female presented with abdominal pain and recurrence of hand blisters

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Introduction

Porphyrias are a group of diseases caused by inherited enzyme dysfunctions in heme biosynthesis. They can be classified as hepatic or erythropoietic, depending on whether the liver or bone marrow is primarily affected by the initial accumulation of pathway intermediates. Clinically, they can also be categorized into acute porphyrias, which present with abdominal pain, neuropathy, and neuropsychiatric changes; blistering cutaneous porphyrias, characterized by chronic skin blistering and scarring; and non-blistering cutaneous porphyrias, which cause recurrent, painful, but mostly non-blistering photosensitivity. Clinically, acute porphyria attacks often lead patients to seek medical treatment repeatedly and seek emergency help. Acute intermittent porphyria is the most common, followed by variegate porphyria (VP), hereditary coproporphyria, and aminolevulinic aciddehydratase deficient porphyria (ALADP)[1]. The diagnosis is often difficult and confusing to physicians.

Case Report

This 29-years-old female with history of unknown skin lesion on bilateral hands was admitted to emergency department (ED) due to persistent epigastric cramping pain for 5 days, accompanying with nausea, vomiting and watery diarrhea. At ED, laboratory data showed white blood cell was 4200 per microliter, with a segmented neutrophil percentage of 48.6%, hemoglobin was 13.7 g per deciliter, alanine transaminase was 27 internal units per liter; lipase was 24 units per liter, creatinine was 0.93 mg per deciliter, total bilirubin was 1.3 mg per deciliter, and sodium was 135 mmol per liter. Esophagogastroduodenoscopy revealed gastroesophageal reflux disease and hemorrhagic gastritis, while an abdominal ultrasound showed gallbladder sludges. However, four days later, the patient returned to the ED due to recurrent abdominal pain. Abdominal computed tomography showed fatty liver and liver cyst. Besides, recurrence vesicles formation at bilateral hands were developed. The patient was admitted for further survey, which showed positive of urine porphobilinogen test. The confirm test revealed increasing urine Uroporphyrin, increasing urine Coproporphyrin I + III, normal Porphobilinogen deaminase (PBGD). The lead poisoning was excluded. Plasma fluorescence scanning showed a peak at 626 nm. The patient was diagnosed as VP, and started to receive hemin supplement. Her abdominal pain and skin lesions improved significantly after treatment. Further gene test showed PPOX gene, exon 13, find c1.330_1331del mutation. Thereafter, preventive supplementation with Normosang 1 amp of heme every 3 weeks, adjusting dose based on symptoms.

Discussion

Variegate porphyria (VP) is an inherited porphyria characterized by both cutaneous porphyria and

acute porphyria. The most common manifestation of VP is adult-onset cutaneous blistering lesions (subepidermal vesicles, bullae, and erosions that crust over and heal slowly) on sun-exposed skin, especially on the backs of hands and face. Acute neurovisceral symptoms can occur at any time after puberty, but less frequently in the elderly. The most common manifestations include severe, steady abdominal pain that is diffuse rather than localized. Other symptoms may include constipation, back pain, chest pain, anxiety, seizures, hyponatremia, and a primarily motor neuropathy that may progress to quadriparesis and respiratory paralysis[1,2].

The biochemical diagnosis of VP is established in an individual with elevated urine porphobilinogen (PBG) or porphyrins and a fluorescence peak at approximately 626 nm on plasma fluorescence scanning. It is generally preferred to perform confirmatory single-gene (PPOX) testing after establishing the biochemical diagnosis of VP[3].

As for management, the cornerstones of management include discontinuation of porphyrinogenic drugs and chemicals, administration of oral or intravenous dextrose and intravenous hemin, and the use of analgesics and antiemetics.

Conclusion

Although rare, acute hepatic porphyria should be considered in the evaluation of all patients, particularly women aged between 15 and 50 years who experience recurrent severe abdominal pain that cannot be attributed to common causes[2].

病例報告 113_C96

庫欣氏症候群引起之低血鉀與嚴重感染

Cushing's syndrome with hypokalemia and severe infection: a case report

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Introduction

A female patient presented with limb weakness over several months. The CT scan revealed bilateral adrenal masses and lung nodules. Hypokalemic metabolic alkalosis and elevated cortisol levels lead to a diagnosis of Cushing's syndrome. This case study will discuss the clinical manifestations of CS and its potential association with severe infectious events.

Case Report

A 67-year-old woman with hypertension presented to our hospital with diarrhea for two days. She also suffered from back pain and progressive leg weakness, resulting in frequent falls over a two-month period. Significant muscle atrophy and multiple ecchymosis were observed. Crusted lesions on the chest indicated the recovery stage of herpes zoster. CT scan revealed bilateral adrenal masses with a maximum diameter of 4 cm, and two nodules in the right middle lobe of the lung. A spine MRI showed L3-L5 HIVD.

After admission, multiple episodes of hypokalemia occurred, with the lowest potassium level 2.3 mmol/L, accompanied by metabolic alkalosis. The high urine K:Cr ratio of 431.1 mmol/g (>22) suggested urinary potassium loss. Hypokalemic metabolic alkalosis led to the consideration of activation in the renin-angiotensin-aldosterone system. Her aldosterone-renin ratio and aldosterone were normal. Notably, she exhibited elevated serum cortisol levels, with morning cortisol at 57.49 ug/dL (6.7-22.6) and ACTH 10.6 pg/mL. The 24-hour urinary cortisol level was markedly elevated over 60 ug/dL. These findings led to a suspicion of Cushing's syndrome, which was later confirmed by dexamethasone overnight test. In general, plasma ACTH levels are typically suppressed in instances of adrenal cortisol excess. The normal ACTH levels suggested ACTH-dependent hypercortisolism, indicating a loss of negative feedback. A high-dose dexamethasone suppression test was conducted to determine whether the ACTH originated from the pituitary. Most pituitary corticotroph adenomas still display regulatory features, while ectopic ACTH sources typically resist dexamethasone suppression. In this case, cortisol levels only decreased from 26.64 to 22.17 ug/dL after the test (positive if suppression > 50%), indicating ectopic ACTH production. However, a bronchoscopic biopsy of the lesion showed negative of malignancy. Her family declined further tissue sampling.

During the hospital days, four limb weakness intensified, with dysarthria and dysphagia. A brain MRI revealed no metastases or stroke. A lumbar puncture found positive for *Varicella Zoster Virus*. She was then intubated due to development of drowsiness and bradypnea. During the ICU stay, she suffered from frequent infections including carbapenem-resistant *Acinetobacter baumannii* pneumonia and candidemia.

Discussion

Cushing's syndrome is an uncommon and potentially life-threatening disorder resulting from prolonged hypercortisolism. Early detection of hypercortisolism is crucial for diagnosis. The primary exclusion should be the exogenous use of glucocorticoids. Endogenous CS includes both ACTH-dependent and independent causes. Cushing's disease, which represents a pituitary origin, is the most common form of ACTH-dependent causes (80-90%). Non-pituitary ACTH-secreting tumors (ectopic) account for 10–20% of ACTH-dependent forms, with the lung being the most frequent site. Adrenal tumors or hypertrophy are common ACTH-independent causes.
The manifestations of hypercortisolism are largely nonspecific, often resulting in diagnostic delay. Infection has been reported as the main cause of death within 90 days of diagnosis. The susceptibility to severe bacterial and opportunistic infections appears to be positively correlated with cortisol levels. Routine screening for pathogens responsible for disseminated infections, such as *Candida*, *Pneumocystis*, *Histoplasma*, *Cryptococcus*, *Pseudomonas aeruginosa*, *Nocardia*, and *Klebsiella pneumoniae*, may be considered.

Conclusion

Hypokalemia and metabolic alkalosis suggest that hypercortisolism is activating mineralocorticoid receptors, with frequent infections indicating a compromised immune system. This combination should prompt consideration of hypercortisolism and initiation of the diagnostic process.

病例報告 113_C97

乾燥症相關肺部結節性澱粉樣病變的案例報告

A case report of nodular pulmonary amyloidosis associated with Sjögren's syndrome

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Introduction

Amyloidosis is a heterogeneous disease characterized by the deposition of toxic, insoluble beta-sheet fibrillar protein aggregates in various tissues. Typically, it is a systemic disease that impacts multiple organ systems, resulting in organ failure and significant mortality. It can also be localized to a single organ, commonly affecting the orbit, pharyngeal-laryngeal region, tracheobronchial tree, pulmonary parenchyma, and urinary bladder [1]. In this report, we present a case of nodular pulmonary amyloidosis associated with Sjögren's syndrome.

Case Report

A 43-year-old male non-smoker with no known systemic diseases presented to our clinic with intermittent cough for several months. He worked in a chemical factory and denied any family history of lung cancer. There were no signs of fever, weight loss, or night sweats. His physical examination was unremarkable. Chest radiography revealed a nodule in the right lower lobe (Figure 1). Computed tomography showed multiple calcified pulmonary nodules in both lungs, measuring between 2 and 6.5 mm (Figure 2), and a lobulated soft tissue mass in the anterior mediastinum measuring 51 x 22 x 56 mm. Thoracoscopic wedge resection of the right upper and middle lobes, along with excision of the nodule and biopsy of the mediastinal tumor, were performed. The pathological report indicated nodular pulmonary amyloidosis and thymic hyperplasia, respectively. Further evaluation of the etiology of the amyloidosis was conducted. Serum and urine immunofixation electrophoresis ruled out multiple myeloma and monoclonal gammopathy. Autoimmune profiles were tested, revealing an antinuclear antibody (ANA) titer of 1:640 with a speckled nuclear pattern, anti-Sjögren's syndrome-related antigen A (anti-SSA) ≧240 U/mL, and anti-Sjögren's syndrome-related antigen B (anti-SSB) ≧320 U/mL. A diagnosis of Sjögren's syndrome with nodular pulmonary amyloidosis was subsequently established.

Discussion

Pulmonary amyloidosis can be either localized or part of systemic amyloidosis and is classified into three subtypes: nodular, diffuse alveolar-septal, and tracheobronchial amyloidosis [2]. Previous studies indicate that nodular pulmonary amyloidosis may result from mucosa-associated lymphoid tissue (MALT) lymphoma or autoimmune diseases such as rheumatoid arthritis or Sjögren's syndrome [3]. It is suggested that Sjögren's syndrome may lead to the infiltration of lymphocytes and plasma cells in specific organs, resulting in the local production of immunoglobulin light chains that deposit as amyloid fibrils [4]. Nodular pulmonary amyloidosis typically presents as solitary nodules, whereas amyloidosis associated with Sjögren's syndrome often manifests as multiple calcified nodules [5]. Chest radiography and computed tomography have limited ability to differentiate amyloidosis from other diseases. Therefore, diagnosis relies on

a comprehensive evaluation that includes symptoms, imaging studies, electrophoresis, autoimmune markers, tissue biopsy, and, in some cases, bone marrow examinations. For the management of nodular pulmonary amyloidosis, conservative excision is usually sufficient, and the long-term prognosis is favorable. Systemic treatment may be necessary if there is an underlying lymphoproliferative disorder.

Conclusion

In summary, we presented a rare case of nodular pulmonary amyloidosis associated with Sjögren's syndrome, which is difficult to diagnosis and necessitates a comprehensive evaluation for potential underlying systemic diseases.

病例報告 113_C98

肺蛋白沉積症肺灌洗的創新方式：一個成功的個案報告

An Innovative and Effective Approach to Whole Lung Lavage for Pulmonary Alveolar Proteinosis - A Case Report of Successful Treatment

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Introduction

Pulmonary alveolar proteinosis (PAP) is a rare lung disease characterized by impaired alveolar macrophage function, leading to the abnormal accumulation of surfactant protein within the alveoli. Whole lung lavage (WLL), a procedure aimed at physically removing the proteinaceous material from the alveoli, has been the cornerstone of PAP treatment since its first description in 1964. WLL has demonstrated long-term efficacy for most patients and is considered both safe and effective. However, standardized guidelines for the WLL procedure remain absent. This article presents a case of PAP successfully treated with WLL, where the procedure was performed bedside in the Intensive Care Unit (ICU), deviating from the traditional approach, which typically requires an operating room. By conducting the procedure bedside, we demonstrate its feasibility and potential to reduce overall costs. A detailed description of the procedure, precautions, and monitoring is provided to emphasize its practicality and safety.

Case Report

A 30-year-old female presented with progressive exertional dyspnea and chronic cough. Diagnostic workup confirmed PAP, and she underwent WLL, which resulted in improved oxygenation and exercise capacity. Follow-up assessments, including the 6-Minute Walk Test (6MWT) and Pulmonary function tests (PFTs) demonstrated progressive recovery after multiple WLL procedures.

Discussion

We provided a detailed step-by-step protocol for WLL, supplemented with images for visual guidance. In addition, we compared our protocol with previously published methods from other institutions, highlighting the potential advantages and disadvantages of different approaches.

Conclusion

By sharing this successful treated case, we aim to present a new approach to WLL that potentially balances cost, safety, and efficacy. However, determining the most appropriate WLL method will require more comprehensive future studies.

病例報告 113_C99

疑似纖維性縱隔腔炎引發的肺部高血壓-案例報告

A Case Report of Suspected Fibrosing Mediastinitis-Induced Pulmonary Hypertension

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Introduction

Fibrosing mediastinitis is a rare disorder characterized by fibrotic sclerosis that encases and impairs mediastinal structures. It is a rare cause of Group 5 pulmonary hypertension, often misdiagnosed or underdiagnosed.

Case Report

We report the case of a 21-year-old woman who initially presented with upper abdominal pain. An upper gastrointestinal endoscopy revealed a single esophageal varix. She later developed progressive shortness of breath, and imaging showed massive pericardial effusion with a D-shaped ventricle. Pericardiocentesis was performed, but she subsequently developed cardiogenic shock, requiring temporary veno-arterial (V-A) extracorporeal membrane oxygenation (ECMO) support. After successful weaning from V-A ECMO, right heart catheterization confirmed pulmonary hypertension with pre-capillary hemodynamic characteristics, leading to an initial suspicion of idiopathic pulmonary arterial hypertension. Despite outpatient treatment with the phosphodiesterase-5 inhibitor sildenafil and the prostacyclin analogue iloprost, she remained symptomatic, with marked fluid retention and recurrent pleural effusions. She was also referred to the thoracic medicine outpatient department for suspicion of extrapulmonary tuberculosis (positive right pleural effusion TB-PCR, but negative acid-fast stain and culture). She was later admitted to National Taiwan University Hospital for acute heart failure. Repeated right heart catheterization revealed combined post- and pre-capillary pulmonary hypertension, raising suspicion of constrictive pericarditis. Surgical intervention included bilateral pulmonary vein enlargement with a bovine pericardium patch, partial pericardiectomy, and right pulmonary artery reconstruction. Histological examination supported the diagnosis of fibrosing mediastinitis.

Discussion

Symptoms of fibrosing mediastinitis arise from compression of mediastinal structures. In our case, pulmonary hypertension arose from pulmonary artery compression, while pulmonary vein compression caused congestion that contributed to refractory pleural effusion. Pericardial compression causes constrictive pericarditis. Congestive liver, portal hypertension, and pitting edema were caused by right heart dysfunction.

Potential triggers of fibrosing mediastinitis include infections (such as histoplasmosis and tuberculosis), sarcoidosis, malignancies, and autoimmune processes. In our case, extrapulmonary tuberculosis can't be ruled out as a potential triggering factor for fibrosing mediastinitis; however, false positives need to be considered due to negative culture results and no further positive evidence in repeat sampling.

Chest radiograph findings, although non-specific, can aid in diagnosis. Our patient had the triad of

atelectasis, pulmonary artery congestion, and pleural effusion described by Cao YS et al. (2020). Four-chamber equalization noted during the first right heart catheterization exam, with similar pressure levels between left and right ventricular diastolic pressures, right atrial pressure, and pulmonary wedge pressure, could also be a sign of constrictive pericarditis.

Conclusion

Fibrosing mediastinitis is a rare cause of pulmonary hypertension. Careful interpretation of imaging findings is essential, as it can provide diagnostic clues for early identification.

病例報告 113_C100

侵襲性異乳鏈球菌感染相關之中毒性休克症候群的案例報告

A case report of toxic shock syndrome associated with invasive Streptococcus dysgalactiae infection

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Introduction

Streptococcal toxic shock-like syndrome (STSS) is a severe and life-threatening condition, commonly associated with Streptococcus pyogenes (group A streptococcus, GAS). It is characterized by rapid onset of shock, multiorgan failure, and a high mortality rate. However, cases involving other streptococcal groups have been increasing. This report presents a case of toxic shock syndrome caused by Streptococcus dysgalactiae.

Case Report

A 48-year-old male cement worker with hypertension and alcoholic liver cirrhosis was transferred to the emergency department (ED) due to altered mental status. He was previously independent in daily activities. Initial evaluation at a local hospital showed hypoglycemia and hypotension, and he was subsequently intubated. Full-body CT imaging showed no intracranial abnormalities. Based on a tentative diagnosis of sepsis due to urinary tract infection, the patient was started on piperacillin/tazobactam before being transferred to our ED.

On examination, swelling and redness of his right lower leg, coupled with his deteriorating condition, raised strong suspicion of necrotizing fasciitis. Emergency fasciotomy was performed, and broad-spectrum antibiotics were continued. Despite aggressive fluid resuscitation, the patient developed profound septic shock, acute kidney injury (AKI), and metabolic acidosis. He was admitted to the medical intensive care unit (MICU) where continuous renal replacement therapy was initiated.

Blood and wound cultures grew *Streptococcus dysgalactiae*, raising suspicion for STSS. The antibiotic regimen was broadened to include teicoplanin and clindamycin. Despite these measures, necrosis of the right leg progressed and amputation was recommended, but the family declined surgery due to the poor prognosis and high surgical risk. The patient died on the fourth day of admission.

Discussion

Streptococcus dysgalactiae is a Gram-positive bacterium capable of causing a wide range of diseases from mild skin infections to severe conditions like necrotizing fasciitis and bacteremia. Invasive *S. dysgalactiae* infections typically occur in older adults or individuals with significant comorbidities. The rising incidence of these conditions poses a growing public health concern, especially in aging populations.

According to the Centers for Disease Control and Prevention (CDC), STSS is diagnosed when group A streptococci are isolated, and clinical criteria such as hypotension and multi-organ involvement are met. In this case, although *S. dysgalactiae*, a group G streptococcus, was isolated, the patient

met the clinical criteria for STSS.
Several studies, particularly from Japan, have reported an increasing prevalence of invasive *S. dysgalactiae* infections in patients over 50 years old with underlying conditions. Risk factors for poor outcomes include liver disease, shock, and elevated serum creatinine and creatine kinase levels, all of which were present in this case.

Conclusion

This case highlights the severe and rapidly progressing nature of *Streptococcus dysgalactiae* infection, which can lead to STSS. The increasing incidence of this invasive infection, particularly in developed countries with aging populations, emphasizes the need for early diagnosis and aggressive treatment to improve outcomes.

病例報告 113_C101

感染性心內膜炎引起的急性冠狀動脈栓塞

Coronary artery embolism resulted from septic emboli of infective endocarditis

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Introduction

Septic embolism is a well-known complication of infective endocarditis, but septic embolism related ST-elevation myocardial infarction (STEMI) is rare and life-threatening. Coronary artery embolism (CE) is considered a cause of nonatherosclerotic STEMI, including embolism caused by atrial fibrillation, cardiac tumor, infective endocarditis and malignancy. We reported a case of infective endocarditis presented with anterior STEMI and subarachnoid hemorrhage (SAH).

Case Report

A 59-year-old man had fever accompanied by chill, dyspnea, chest pain and chest tightness for 3 days. Laboratory data showed leukocytosis with elevated infectious profile and cardiac enzyme. An electrocardiogram revealed ST-segment elevation in leads V1-V4. He underwent emergent coronary angiography (CAG), which revealed left anterior descending artery (LAD) segment 7 was 99% obstructed by thrombus and TIMI 0-1 flow. After aspiration thrombectomy with some whitish thrombi was aspirated, final report showed LAD TIMI 3 flow. Intravascular ultrasound (IVUS) during CAG showed diffuse plaque over LAD Segment 6-7 without significant stenosis. In addition, brain computed tomography (CT) showed left frontal minimal SAH, with perifocal edema.
Transthoracic echocardiography (TTE) showed left ventricular ejection fraction (LVEF) 47% with severe aortic regurgitation (AR), suspected infective endocarditis (IE), or sinus of Valsalva rupture. Transesophageal echocardiography (TTE) was therefore performed, and the results showed severe AR with multiple vegetations on the aortic valve, involving right coronary cusp and left coronary cusp, with a maximum size of 1.5x0.4 cm. After discussion with the cardiovascular surgeon, aortic valve replacement surgery was planned after a 28-day course of antibiotics. Apixaban was also added after a subsequent brain CT showed no progression of SAH but some hypodense content, indicating the presence of septic emboli. The patient received an operation of aortic valve replacement with Extra-Corporeal Membrane Oxygenation (ECMO) and intra-aortic balloon pump (IABP) support. The surgery was successful, and the ECMO and IABP were removed a few days later. However, new infectious events and severe left ventricle hypokinesis with cardiogenic shock occurred. The patient died due to ventricular tachyarrhythmia.

Discussion

STEMI is a well-known life-threatening event caused by atherosclerotic plaque rupture with complete occlusion of the coronary arteries. In some cases, clinical evidence of myocardial infarction may manifest as normal and near-normal coronary arteries on angiography, such as coronary embolism (CE). The definition of CE was established by Shibata et al, including three major and three minor criteria [1].

The most common cause of CE is atrial fibrillation, followed by dilated cardiomyopathy, infective endocarditis, and intracardiac tumor. Other less common etiologies included valvular prosthesis, deep vein thrombosis and hypercoagulable states, such as malignancy and antiphospholipid syndrome [2]. In the absence of coronary artery stenosis, traditional treatments for STEMI, including percutaneous coronary intervention with stent placement and dual antiplatelet therapy (DAPT), may not be needed. Treatment strategy should be tailored to the cause of coronary embolism. Complications of the etiology should also be considered. In addition, CE has been reported to be associated with higher mortality compared with STEMI associated with atherosclerotic plaque rupture.

Previous studies have presented cases of coronary embolism [3]. In our case, the patient had infective endocarditis complicated with septic embolism-related STEMI and severe AR caused by large vegetation. Apixaban was prior to DAPT for septic embolism. Aortic valve replacement should also be performed for severe AR.

Conclusion

Coronary embolism is a rare cause that may lead to acute coronary syndrome. The definition of coronary embolism has been established. The most common cause of CE included atrial fibrillation, cardiomyopathy and infective endocarditis. Previous studies reported a higher mortality rate of CE. In this case, we would like to highlight the rare complication of infective endocarditis and the different treatment strategies compared with atherosclerotic plaque rupture related ACS.

病例報告 113_C102

單側腎上腺切除術治療雙側腎上腺增生合併原發性醛固酮增多症的療效

The Treatment Efficacy Of Unilateral Adrenalectomy in Bilateral Adrenal Hyperplasia with Primary Aldosteronism

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Introduction

Primary aldosteronism is an endocrine disorder where the adrenal glands produce excessive aldosterone, leading to an imbalance in sodium and potassium levels in the body, which can cause hypertension and hypokalemia. It is estimated to be responsible for 5 to 20 percent of hypertension in humans. The treatment options include mineralocorticoid receptor antagonists and laparoscopic adrenalectomy, depending on the subtype of the disease. Subtypes include bilateral adrenal hyperplasia(BAH), aldosterone-producing adenoma(APA), idiopathic hyperaldosteronism(IHA), familial hyperaldosteronism(FH), adrenocortical carcinoma(ACC), and ectopic aldosterone-producing adenoma.

Case Report

A 52-year-old man with a history of type 2 diabetes mellitus, hypertension, hyperlipidemia and insomnia, was on Bisoprolol 5 mg, indapamide 1.5 mg and sevikar 5/20 mg (amlodipine+olmesartan) once daily respectively for hypertension control. He presented with bilateral leg weakness for 3 days. His laboratory data showed severe hypokalemia (Potassium level: 1.6 mmol/L). No intracranial hemorrhage or acute stroke was detected after brain CT scans. After admission, urine potassium-to-creatinine ratio (14.37) and transtubular potassium gradient (4.71) were checked, indicating renal potassium loss. Subsequently, a high aldosterone renin ratio (45) and saline infusion test (post infusion aldosterone value: 22.64 ng/dL) confirmed primary aldosteronism.

Abdominal CT scan and Adrenal Sampling

Abdominal CT scan revealed bilateral adrenal nodular thickenings, especially on the left side (Fig. 1). Adrenal sampling was then arranged (Fig. 2.3). The result was showed a left aldosterone-producing adenoma (Table.1 and Table.2), although sampling seemed to have failed on the right

side.

I-131 NP-59 SPECT/CT and Left Laparoscopic Adrenalectomy

Due to the failed adrenal sampling, I-131 NP-59 (131 6-beta-iodomethyl-19norcholesterol) SPECT/CT was further arranged, which revealed an aldosterone-producing adenoma on the left side (Fig 4) .Left laparoscopic adrenalectomy was then performed (Fig.5).

Pathology Report and Follow-up

The pathology report confirmed adrenal cortical adenoma. Postoperative laboratory data showed resolution of hypokalemia(potassium levels increased to 4 mmol/L)and improvement in plasma renin activity(PRA increased from 0.3 to 0.54). The patient's blood pressure stabilized, and his antihypertensive medications were reduced gradually as well.

Discussion

The primary goal in treating patients with primary aldosteronism is to reduce the morbidity and mortality associated with hypertension, hypokalemia, kidney toxicity, and cardiovascular damage. In this case, we demonstrated the treatment efficacy of unilateral adrenalectomy on a patient with bilateral adrenal hyperplasia and unilateral aldosterone- producing adenoma. The management of bilateral adrenal hyperplasia remains controversial. While medical therapy is commonly recommended, it often provides with insufficient blood pressure control effect. Subtotal adrenalectomy may not achieve adequate blood pressure regulation, and bilateral adrenalectomy carries significant risks, including the need for life-long glucocorticoid and mineralocorticoid supplementation.

However, in this case, a left aldosterone-producing adenoma was detected through adrenal venous sampling and I-131 NP-59 SPECT/ CT. Thus, unilateral Laparoscopic adrenalectomy was performed, resulting in satisfactory biochemical outcomes and improved quality of life.

Conclusion

Unilateral adrenalectomy can be an effective treatment option for patients with bilateral adrenal hyperplasia. This case highlights the importance of accurate diagnosis and appropriate treatment of primary aldosteronism to prevent cardiovascular complications.

病例報告 113_C103

轉移性胰腺癌患者放置金屬支架後出現的總膽管十二指腸廔管

Choledochoduodenal fistula after placement of metal stents for metastatic pancreatic cancer

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Introduction

Self-expandable metallic stents (SEMS) is the mainstay of treatment for unresectable malignant biliary strictures. With the increased use of SEMS, a number of serious complications have been reported. Choledochoduodenal fistula (CDF) is one of the extremely rare, delayed adverse event. Here, we present a case of CDF after SEMS placement for distal common bile duct obstruction in a patient with metastatic pancreatic cancer.

Case Report

This 68-year-old woman presented with progressive jaundice for one month. Abdomen computed tomography (CT) showed suspicious pancreatic head cancer with obstruction of biliary tract caused by tumor compression. Endoscopic retrograde cholangiopancreatography (ERCP) was performed for placement of endoscopic retrograde biliary drainage (ERBD) and trans-papillary biopsy. We diagnosed the patient with pancreatic ductal adenocarcinoma (PDAC), cT2N0M0, stage IB. Neoadjuvant chemotherapy with SLOG (TS-1, leucovorin, oxaliplatin, gemcitabine) was prescribed for resectable PDAC with high risk feature and 3 times of ERBD revision was performed due to cholangitis. However, 6 months later, Positron Emission Tomography-Computed tomography (PET-CT) for pre-operation survey disclosed thoracic spine metastasis. We performed ERBD revision with uncovered SEMS (Taewoong Niti-S 10mm x 60mm). We changed chemotherapy regimen to NALIRIFOX (Onivyde, leucovorin, 5-Fluorouracil, oxaliplatin) and to AGSL (Abraxane, gemcitabine, Ts-1, leucovorin) and the patient underwent palliative radiation therapy 35Gy in 10 fractions to bone metastasis and external beam radiation therapy (EBRT) 45Gy in 5 fractions to pancreatic cancer. One year after SEMS insertion, the follow-up CT revealed persistence of PDAC but stent occlusion without stent migration or deformity and wall thickening of the duodenum with distended stomach. The upper endoscopy revealed that the mesh of the lateral aspect of SEMS was clearly visible eroding through the large circumferential ulcer base in the duodenal bulb. To maintain the patency of the SEMS, we attempted to perform biliary drainage through the cell of the SEMS but failed. The fluoroscopy revealed CDF with patent SEMS and no evidence of contrast leakage into the extraduodenal abdominal cavity. The patient received Roux-en-Y gastric bypass and feeding jejunostomy and was discharged home following palliative cancer treatment.

Discussion

We reported a case of asymptomatic CDF after placement of SEMS for biliary stricture caused by metastatic PDAC. The patient had good performance status and quality of life after Roux-en-Y gastric bypass. CDF is a rare complication of SEMS related to bile duct wall injury caused by stent migration, sharp end of the metal stent, chemoradiation or tumor invasion. The mechanism of forming a CDF could not be clearly verified, however, vulnerable bile duct wall due to

chemoradiation was likely involved in the present case. Given the efficacy and safety of novel chemotherapy, the overall survival rates of PDAC have gradually improved over the years. CDF, a rare and delayed complication of SEMS was sporadic reported. We present this case and share our experience with physicians in Taiwan.

病例報告 113_C104

晚期尿路上皮癌患者的遲發性 Pembrolizumab 誘發胰臟炎：案例報告與臨床觀察

Late-Onset Pembrolizumab-Induced Pancreatitis in Advanced Urothelial Carcinoma: A Case Report and Clinical Observations

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Case Report

A 68-year-old male patient, diagnosed with high-grade left renal pelvis urothelial carcinoma, underwent 14 cycles of Pembrolizumab immunotherapy (200 mg/dose every three weeks), with the most recent administration on 2024/05/03. His medical history includes several comorbidities, such as myelodysplastic syndrome, systemic lupus erythematosus, inflammatory bowel disease, and hepatitis B, all managed with regular follow-ups.

The patient arrived at the emergency department after experiencing several days of vomiting and diarrhea, though without symptoms of abdominal pain or fever. Laboratory tests revealed elevated lipase levels of 1615 U/L (normal range: 13–55 U/L), prompting a Computed Tomography scan of the abdomen. The scan confirmed acute pancreatitis, classified as Balthazar score grade E, and identified the presence of retroperitoneal fluid. Given the absence of common etiologies such as alcohol use or gallstones, the differential diagnosis included autoimmune pancreatitis and Pembrolizumab-related pancreatitis. The patient underwent blood tests for Antinuclear antibodies (ANA), Immunoglobulin G4 (IgG4), C3, and C4, all of which returned normal results. An ultrasound-guided pancreas biopsy was performed and revealed predominant plasma cell infiltration, and IgG4 immunostaining was negative.

Despite the lack of clinical symptoms, follow-up blood tests showed a persistent elevation in lipase levels. Due to highly suspect Pembrolizumab-induced pancreatitis, the patient was started on intravenous methylprednisolone 40 mg per day. The steroid therapy led to improvements in blood test results and a reduction in pancreatic swelling. Subsequently, the patient's oral intake was gradually resumed, and the steroid dosage was tapered to 15 mg/day of prednisolone. The patient was hospitalized for a total of 15 days. Ongoing observation is planned to monitor his condition.

Discussion

With the increasing use of Immune Checkpoint Inhibitors (ICIs), various side effects have gradually emerged, one of which is ICI-related pancreatitis (IRP). Currently, the incidence of IRP is low, approximately 0.5–1.6% for anti-PD-1. The onset of pancreatitis after the initiation of ICIs ranged from 18 days to 16 months, adding another layer of difficulty to the diagnostic process.

In the presented case, pancreatitis symptoms appeared approximately 11 months after the start of Pembrolizumab immunotherapy. Meanwhile, the patient also developed inflammatory bowel disease (IBD) after 8 months of Pembrolizumab treatment, which may also be related to immunotherapy. Notably, our patient underwent a pancreatic biopsy, which revealed predominant plasma cell infiltration, and negative IgG4 immunostaining. Histologic evaluation has been performed in only a limited number of cases. A literature review of twelve patients with IRP found considerable variability, with five patients exhibited predominantly neutrophilic

infiltrates, five had predominantly lymphocytic infiltrates, and two showed a mix of both. Despite these varied findings, pancreatic biopsy remains a crucial consideration when IRP is suspected. This procedure plays a vital role in differentiating IRP from other conditions, including alcoholic pancreatitis, gallstone pancreatitis, and pancreatic tumor induced pancreatitis.

We present a case of pancreatitis likely induced by pembrolizumab. While IRP is not a frequent adverse event, its potential to cause significant morbidity and mortality makes it a clinically relevant concern. Clinicians should consider the possibility of IRP when patients on these therapies present with classical symptoms of pancreatitis. In these situations, a prompt diagnostic workup, including appropriate imaging and histological examinations, is essential for accurate diagnosis and timely intervention.

病例報告 113_C105

經典案例分享：年輕運動員迷走神經過激引發之心臟房室干擾

Vagotonia Related Atrioventricular Interference in A Young Adult Athlete-a classic case sharing

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Introduction

Bradycardia is not uncommon in healthy young people. The autonomic nervous system plays an important role in cardiac rhythm modulation, especially in young adults. In athletes, high-intensity exercise could lead to structural and functional changes, including increased left ventricular volume, thicker ventricular wall, increased stroke volume, altered cardiac-neural modulation, and lower heart rate. It's important to differentiate the etiology and diagnosis of bradycardia in athletes, because although most bradycardia is physiologically benign, it can also be fatal, such as high-degree AV block or sinus node dysfunction, which can be seen in overtrained athletes.

Case Report

Here we report a case of a 19-year-old female athlete without underlying disease who presented to our outpatient clinic with intermittent chest tightness for almost 2 years after COVID-19 vaccination. During an outpatient visit, the ECG revealed AV dissociation and the patient was subsequently admitted for further evaluation. Cardiac echo showed normal cardiac structure with trivial pericardial effusion. Given the above, along with her history of mRNA COVID-19 vaccination and self-reported occasional chest tightness, myocarditis-related AV block had to be ruled out. Therefore, an electrophysiology study (EPS) was ordered. In the EP laboratory, the baseline intracardiac electrogram (IEGM) showed AV dissociation with low atrial rate associated with junctional escape and occasional normal conduction. High right atrium (HRA) pacing showed loss of 1:1 AV conduction at cycle lengths below 900 ms, with intermittent AV block occurring as supra-Hisian block. Atropine 0.5 mg injection was given for vagolysis and her heart rate increased. IEGM showed normal AV 1:1 conduction. HRA pacing showed AV 1:1 conduction to a cycle length of 350ms without AV block. This young adult athlete's AV dissociation was confirmed as vagotonia related atrioventricular interference, which does not require any intervention or treatment.

Discussion

Bradycardia is mostly benign in young adults, but serious conditions such as AV block or sinus node dysfunction must be recognized. Once an AV conduction abnormality is identified, a detailed history and a thorough examination to determine the underlying etiology are critical. The physiologic state of athletes differs from that of the general population due to their high-intensity training. The autonomic nervous system change, especially vagotonia, is often closely associated with bradyarrhythmias. In most young athletes, extreme sinus bradycardia is the predominant manifestation; however, AV interference is a less common presentation. We performed a comprehensive EPS study in this case to confirm that it was a classic vagotonia-related atrioventricular interference in a young adult athlete.

Conclusion

AV dissociation is a serious medical problem that can be life-threatening or require permanent pacing. However, young athletes exhibit distinct cardiac physiological changes compared to the general population, and bradyarrhythmia induced by vagal tone and high-intensity exercise may be normal in this group. Accurate diagnosis is critical. Here, we report a fully confirmed and pedagogically significant case of vagotonia-related atrioventricular interference.

病例報告 113_C106

一則潛伏梅毒的病例報告：如何避免不典型的臨床表現導致漫長診斷路

A Case Report of Latent Syphilis: How to Avoid Diagnostic Odyssey in Elusive Mimicry

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Introduction

Syphilis, a sexually transmitted infection caused by *Treponema pallidum*, can manifest in various ways, ranging from characteristic skin lesions to more subtle symptoms in its later stages. Latent syphilis, a phase without clinical symptoms, can present with inflammatory changes, including vasculitis. Diagnosing vasculitis, an inflammatory process affecting blood vessels, can be challenging, particularly when it occurs atypically, leading to diagnostic delays and potential complications. We describes a 39-year-old male initially presenting with signs mimicking cellulitis, eventually leading to the diagnosis of latent syphilis.

Case Report

A 39-year-old male chicken processor presented to the emergency department (ED) with acute, painful swelling of his right hand and forearm that developed within hours. He had mild discomfort in his left hand and forearm but no other concerning symptoms. His laboratory work-up showed leukocytosis and neutrophilia, suggesting an infectious process (WBC = 11,600/uL, PMN = 78.1%). His systemic inflammatory markers were elevated (CRP = 7.79 mg/dL, MDW = 25.26). Imaging showed no fractures, and CT of his chest and abdomen reported inconclusive results. Despite being advised of further treatment, he refused hospitalization.

The swelling in his right arm resolved in two days, but a similar discomfort appeared on his left arm. During follow-up at the rheumatology clinic, concerns of autoimmune disease arose due to elevation in erythrocyte sedimentation rate (ESR = 59 mm/1h), antinuclear antibody (ANA = 1:80), and anti-cardiolipin IgM (55 MPL-U/mL). The initial RPR test seemed positive (1:64), but he reported no recent sexual contacts. Ultrasound of the joints revealed subcutaneous tissue edema but no synovitis. Empiric treatment with aspirin and hydroxychloroquine showed partial improvement of symptoms, but no definitive diagnosis was reached.

Three weeks after his ED visit, the patient developed palpable purpura, characteristic of leukocytoclastic vasculitis (LCV), on his lower extremities (Figure 1). Further investigations showed a rising RPR titer (1:32) and positive TPPA (1:2560) test results suggested syphilis. Upon further inquiry, he reported unprotected sexual encounters five months prior to visiting the ED. The diagnosis of latent syphilis was established; a single intramuscular dose of 2.4 million units of benzathine penicillin G, along with doxycycline for a week were prescribed. This treatment repeated for 3 weeks and his symptoms effectively resolved (Figure 2).

Discussion and Conclusion

This case demonstrates the difficulty in diagnosing latent syphilis when patients present with atypical inflammatory manifestations. While syphilis may often present with distinctive lesions on

the palms, soles, or in the oral cavity, this patient exhibited confusing signs like migrating joint pain, reminiscent of other inflammatory conditions like cellulitis or autoimmune disorders. The coexisting systemic inflammatory changes may have further confounded initial diagnosis, necessitating meticulous investigation and consideration of syphilis in the differential diagnosis.
Despite an initial concern of autoimmune disease due to positive ANA and anti-cardiolipin IgM levels, it was the distinctive, palpable purpura (LCV), accompanied by a positive history of unprotected sexual activity, that triggered suspicion of syphilis as the culprit. This case reinforces the crucial role of comprehensive evaluation, considering a broad differential diagnosis, and thoroughly investigating potential underlying causes. This case highlights the significance of recognizing and considering syphilis as a cause of inflammatory conditions beyond its classic clinical presentation, leading to accurate and timely diagnosis and treatment.

病例報告 113_C107

第四期 ALK 陽性肺腺癌病患產生新的惡性肋膜積液：產生抗藥性嗎？NGS 的關鍵作用

Development of a New Malignant Pleural Effusion in a Stage IV ALK-Positive Lung Adenocarcinoma Patient: Treatment Resistance? The Critical Role of NGS

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Introduction

Anaplastic lymphoma kinase (ALK)-positive lung adenocarcinoma accounts for approximately 4-6% of all non-small cell lung cancers (NSCLC) in Taiwan. With the advent of personalized therapy and precision medicine, ALK-tyrosine kinase inhibitors (ALKi) have become a cornerstone in the treatment of these patients, showing better progression-free survival (PFS) and overall survival (OS) compared to standard chemotherapy. However, the development of new malignant pleural effusions under ALK inhibitor treatment often indicates disease progression, prompting a reassessment of the treatment strategy. Next-generation sequencing (NGS) can be a valuable tool to detect resistance mutations, whether on-target or off-target, which would signal treatment failure and the need for next-line therapies. Conversely, if NGS results show no new mutations, poor drug compliance should be considered as a cause of limited therapeutic response.

Case Report

We present a case of a 68-year-old ex-smoker who initially presented with an 8-month history of chronic cough. He was diagnosed in December 2020 with right lower lobe adenocarcinoma and malignant pleural effusion (cT4N2M1a, stage IVA). Genetic testing revealed negative results for EGFR, ROS1, and PD-L1 mutations, but positive for ALK. Due to personal reasons, the patient did not initiate ALKi therapy and was lost to follow-up. He returned in November 2022 with worsening dyspnea. A chest CT revealed a right lung tumor with multiple metastases, including pleural, liver, spleen, and bone involvement. Brigatinib was initiated on November 26, 2022. His condition improved, and he underwent VATS-based decortication, though the surgical wound did not heal well.

After nearly one year of brigatinib treatment, a new left-sided pleural effusion was incidentally discovered on November 8, 2023. Cytology confirmed malignant cells, raising suspicion of resistance to brigatinib. Pleural fluid was collected and sent for NGS, which surprisingly revealed the presence of the same ALK V3b mutation—a variant of the ALK mutation found previously. Upon further discussion, the patient admitted to poor adherence to brigatinib due to concerns that the drug caused bleeding in his unhealed surgical wound. The poor compliance likely resulted in suboptimal plasma drug levels, contributing to the disease's poor control. After being counseled on the importance of medication adherence, the patient resumed regular brigatinib use. The malignant pleural effusion gradually resolved, and his cancer remained stable without further progression.

Discussion

For ALK-positive lung adenocarcinoma patients, second-generation ALK inhibitors—such as

alectinib, ceritinib, and brigatinib—demonstrate superior efficacy compared to chemotherapy or first-generation crizotinib. However, resistance to first-line ALKis is inevitable, frequently manifesting as local or distant metastasis, including malignant pleural effusion. Tissue analysis and distinguishing between on-target and off-target resistance mechanisms are crucial, with lorlatinib serving as a potential salvage ALKi. However, lorlatinib's duration of efficacy in cases of resistance to first-line ALKi typically ranges from 2 to 6 months.

NGS is an essential tool for identifying resistance mutations. In this case, NGS confirmed the presence of the ALK V3b variant, suggesting that brigatinib remained effective and that non-compliance was likely responsible for lower serum drug concentrations and disease progression. Instead of switching therapies prematurely, addressing the underlying causes of poor drug adherence was key to restoring treatment efficacy.

Conclusion

While resistance to current effective drugs is inevitable, accurately identifying the cause of resistance is crucial. NGS plays a pivotal role in uncovering the mechanisms of resistance and guiding further treatment decisions. In this case, ensuring medication adherence was critical to maintaining disease control, avoiding unnecessary changes to an otherwise effective therapy.

病例報告 113_C108

由血管肉瘤引起的成人腸套疊病例

A case of adult intussusception caused by angiosarcoma

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Introduction

Intussusception is a condition in which one segment of the gastrointestinal tract telescopes into another segment of the bowel. Intestinal intussusception most commonly occurs in pediatric patients and is rare in adults. We present a case of jejunojejunal intussusception caused by angiosarcoma of the jejunum.

Case Report

A 67-year-old male with underlying diseases of hypertension, chronic kidney disease stage 3, and abdominal aortic aneurysm post-stent indwell. He suffered from tarry stool starting on April 1, 2024, associated with epigastric pain and dyspnea on exertion. Laboratory tests showed leukocytosis and severe anemia. Esophagogastroduodenoscopy revealed unremarkable findings. Colonoscopy showed: 'No active bleeding identified, but tarry contents was noted through the whole colon'. Enhanced abdominal computed tomography was performed to exclude small bowel bleeding, which showed 'suspect small bowel intussusception with prominent soft tissue, probably tumor'. Oral route enteroscopy revealed 'suspected proximal jejunal tumor'.

Surgery was performed involving reduction of jejunojejunal intussusception and segmental resection of jejunum with stapled side-to-side anastomosis, along with mesenteric lymph node excision. Intraoperatively, a 20 cm jejunojejunal intussusception was found, located 10 cm from the ligament of Treitz, along with mesenteric lymphadenopathy. Pathology confirmed jejunal angiosarcoma. Positron emission tomography-computed tomography (PET-CT) showed multiple metastasis in the lungs, adrenal glands, lymph nodes, with staging classified as T2aN0M1, stage IV. Port implantation and chemotherapy with paclitaxel and Avastin were initiated. Follow-up enhanced abdominal CT revealed 'local recurrent tumor'. Due to intermittent tarry stools, he underwent palliative surgery for bleeding control. After the surgery, he requested hospice care due to the overwhelming burden of his condition and his inability to tolerate further treatment, and he was subsequently transferred to the hospice ward.

Discussion

Intussusception in adults is an extremely rare condition, occurring in fewer than 0.1% of abdominal surgeries. Diagnosing intussusception in adults is challenging, because it mimics many other diagnoses. Abdominal computed tomography is the most sensitive tool for preoperative diagnosis of adult intussusception.

The most common cause of adult intussusception is neoplasm. Intussusception can be classified either by etiology or location. Etiologically, it can be divided into benign, malignant, or idiopathic. Based on location, it is divided into three types: enteric, confined to the small bowel; ileocolic, where the ileum protrudes into the colon through the ileocecal valve; and colocolic, limited to the

large bowel. Most cases of intussusception in children are idiopathic, though viral infections may play a role in some instances.
Angiosarcoma is a rare malignant mesenchymal sarcoma that arises from vascular or lymphatic endothelial cells. Angiosarcoma of the small intestine has a low incidence and is more commonly seen in elderly male patients. Risk factors include exposure to toxins and radiation. It most frequently occurs in the jejunum, followed by the ileum and duodenum. The diagnosis of small intestinal angiosarcoma is challenging, contributing to its poor prognosis.

Conclusion

Although intestinal intussusception is rare in adults, it should still be considered as a differential diagnosis in cases of unexplained abdominal pain or gastrointestinal bleeding.

病例報告 113_C109

使用 TC-325 止血粉噴霧劑治療巨細胞病毒引起的瀰漫性食道潰瘍出血

Using TC-325 hemostatic powder spray (Hemospray) to stop bleeding from diffuse esophageal ulcer caused by cytomegalovirus infection

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Introduction

TC-325 hemostatic powder spray (Hemospray; Cook Medical, Winston-Salem, North Carolina, USA) is an inert mineral powder that was created for endoscopic hemostasis. Hemospray has demonstrated its capacity to achieve rapid hemostasis, outstanding practicality, and a favorable safety profile for the treatment of different gastrointestinal bleeding (GIB) etiologies. We presented the use of Hemospray to stop the bleeding in a 70-year-old woman with numerous comorbidities who presented with a widespread esophageal ulcer caused by cytomegalovirus (CMV) infection. This is the first time that Hemospray has been used to control GIB in Asia.

Case Report

A 70-year-old woman was admitted to the neurosurgery ward due to lumbar spine osteomyelitis and related complications of paraspinal abscess and compression fracture. She had underlying diseases of end-stage renal disease (ESRD) with long-term hemodialysis, asthma, and chronic inferior vena cava (IVC) thrombosis with long-term warfarin treatment. Corynebacterium jeikeium bacteremia was discovered after admission, and treatment with antibiotics was then administered. Approximately two weeks following her hospitalization for antibiotic therapy, we were consulted regarding hematemesis. To evaluate the cause of upper gastrointestinal bleeding (UGIB), we performed an esophagogastroduodenoscopy (EGD) examination for the patient initially. EGD revealed circumferential and widespread esophageal ulcers with active oozing from the middle to the lower esophagus. Given the limitations of traditional hemostatic methods for such extensive lesions, we used Hemospray for hemostasis, and the bleeders immediately ceased. The patient underwent a very smooth and well-tolerated procedure. Despite the pathologic test later demonstrating cytomegalovirus (CMV) infection, no antiviral medication was administered to the patient. However, another tarry stool episode appeared about nine days later. To check rebleeding, we arranged another EGD examination. Surprisingly, the outcome demonstrated the full recovery of previous esophageal ulcer. As a result, the patient received further work-up for lower GIB.

Discussion

Despite management with the current hemostatic techniques (e.g., injection therapy, Argon plasma coagulation, endoscopic clips), there is still a rebleeding rate of 8-25% of all nonvariceal UGIB cases, especially those with high-risk stigmata or multiple comorbidities.

TC-325 hemostatic powder spray (Hemospray) seems to be a safe and useful treatment. The powder is completely eliminated from the gastrointestinal tract as soon as 24 hours after use,

showing its brief duration in the digestive system. Hemospray demonstrated more than 95% immediate hemostasis efficacy in different studies. Besides, in cases of malignant nonvariceal UGIB treated with Hemospray, the rate of rebleeding was significantly lower than patients treated with conventional endoscopic procedures. Given that certain traditional endoscopic techniques can be challenging to implement in malignant GIB patients, these results suggest that the use of Hemospray is a very promising strategy for treatment. This year, the Taiwan Food and Drug Administration (TFDA) approved the license for Hemospray. Since our case is the first in Asia to employ Hemospray in GIB treatment, we anticipate further experiences with its potential to treat challenging GIB cases.

Conclusion

Hemospray is a fast and safe way to manage some difficult nonvariceal GIB cases that were difficult to treat with conventional endoscopic hemostatic methods.

病例報告 113_C110

惡性胰臟癌以表皮轉移為疾病第一表現的病例報告

Cutaneous metastasis as the first sign of pancreatic cancer

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Introduction

Sister Mary Joseph's nodule (SMJN) is an umbilical metastasis from a visceral malignancy and considered an indicator of poor prognosis. We describe an interesting case of pancreatic cancer with SMJN to highlight its significance as a diagnostic and prognostic indicator for clinicians.

Case Report

We reported a 67-year-old male suffered from periumbilical dull pain since March 2024. Decrease in body weight of 6 kg over the past six and a firm, non-movable, non-tenderness nodule has been noted at umbilicus by himself since July 2024. A slight serosanguinous discharge has been noted since it appearance. The patient visited a local hospital, where an abdominal CT scan revealed a pancreatic tail tumor with another tumor found in the anterior abdominal wall.

The patient was referred to the medical center for MRCP to determine the TNM staging, with an impression of T3N0M1, stage IV. ERCP with EUS-FNB was performed for tissue confirmation. The pathology report confirmed the diagnosis of grade 2 pancreatic adenocarcinoma. Surgery for SMJN removal was performed, and the pathology report revealed metastatic carcinoma, consistent with the diagnosis.

Discussion

Umbilical tumors, mostly considered benign (60%), with 30% of metastatic malignancies, known as Sister Mary Joseph's nodule (SMJN), named after Sister Mary Joseph, a nurse and surgical assistant who observed that cancer patients with umbilical nodules tended to have more advanced disease and a poorer prognosis compared to those without the nodule.

SMJN usually presents as a painful, indurated, irregular protrusion in the umbilical region. The incidence of SMJN is low, only 1%–3% of all intra-abdominal or pelvic malignancies. Histologically, SMJN is most commonly diagnosed as adenocarcinoma, with the majority metastasizing from gastrointestinal malignancies.

Pancreatic cancer accounts for approximately 10% of cases of umbilical metastasis and tends to originate from the body or tail of the pancreas. For the diagnostic approach to pancreatic cancer, EUS-FNB is a safe, rapid, and reliable method for the differential diagnosis of such lesions. The prognosis remains poor for pancreatic cancer patients, regardless of whether they present with SMJN.

Conclusion

In summary, SMJN is a rare subcutaneous manifestation that suggests an underlying advanced

malignancy with an ominous prognosis regardless of the cancer type. The physician must be aware of such lesions, as malignancy should be thoroughly investigated.

病例報告 113_C111

冷凍治療應用於近希斯氏束的局部心房頻脈: 一例病例報告

Management of Parahisian Focal Atrial Tachycardia in a 47-Year-Old Woman: A Case Report on Cryoablation Efficacy

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Introduction

Para-Hisian atrial tachycardia poses a significant challenge due to its proximity to the AV node and His bundle, risking high-degree AV block. We present a successful case of cryoablation in a 47-year-old woman with this condition.

Case Report

The patient, with no significant chronic diseases, sought treatment for frequent palpitations. Initial ECG showed sinus rhythm, and subsequent tests ruled out ischemic heart disease. Despite propranolol treatment, her symptoms worsened, and supraventricular tachycardia (SVT) was later documented. An ECG revealed a narrow complex tachycardia at 147 bpm with a short PR interval and specific P wave morphology indicative of atrial tachycardia originating from the anterior septum or perinodal area.

Following a 14-day Holter monitor showing frequent SVT episodes, the patient underwent an electrophysiologic study (EPS) and potential ablation. Burst pacing in the right atrium triggered SVT. Ventricular overdriving pacing was able to entrain the SVT and showed typical VAAV pattern with apparent high low direction after termination, confirming atrial tachycardia. Electroanatomical mapping revealed the earliest activation site near the para-Hisian area, just 1.5 cm from the His bundle.

Given the risk of complete AV block, cryoablation was discussed, but the patient initially opted for supportive treatment. After two months, persistent symptoms led her to choose cryoablation. A second EPS confirmed the previous findings, with mapping showing a focal atrial tachycardia activation site only 0.6 cm from the His bundle.

During the cryoablation procedure, the tachycardia was triggered but terminated after 20-30 seconds of ablation. Post-procedure, SVT could not be induced. The patient recovered without complications and reported no further episodes at her three-month follow-up.

Discussion

Atrial ectopy and non-sustained atrial tachycardia are common but often asymptomatic. Focal atrial tachycardia is recognized in 15-25% of SVT cases during EPS, and its treatment presents unique challenges, especially in para-Hisian areas due to the risk of AV block.

Traditional approaches to treating para-Hisian tachycardia involve careful radiofrequency (RF) ablation, but this method carries a risk of high-degree AV block in up to 14% of patients. Cryoablation offers a safer alternative, minimizing the risk of permanent damage. This technique effectively adheres the tissue to the catheter as it cools, reducing the likelihood of unintended catheter movement and allowing for reversible effects during the procedure.

Recent studies support the safety of cryoablation for para-Hisian and mid-septal accessory pathways, showing no instances of complete heart block. This case illustrates that cryoablation can be effectively and safely performed close to the His bundle, providing a viable treatment option for para-Hisian focal atrial tachycardia.

Conclusion

Cryoablation, combined with precise electroanatomic mapping and careful monitoring, represents a promising therapeutic strategy for managing para-Hisian focal atrial tachycardia, warranting further research to validate its long-term effectiveness.

病例報告 113_C112

游離脾臟 – 一個無症狀案例的探討

Wandering Spleen - a Brief Overview in an Asymptomatic Case

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Introduction

Wandering spleen is a rare condition in which a spleen is located outside of its normal location. It may lead to severe complications and is underdiagnosed in half of the cases. Here we reported an asymptomatic case and perform a brief overview on wandering spleen.

Case Report

A 29-year-old woman who visited a local gynecologist for abdominal pain initially was transferred to our hematology clinic since splenomegaly was noticed on the sonography. We found iron deficiency anemia (IDA), positive anti-double stranded deoxyribonucleic acid (anti-dsDNA) antibody, and positive anti-connective tissue disease (anti-CTD) antibodies screening on the laboratory test. The patient was referred to the rheumatology clinic, and had been followed up for 2 years, during which she did not receive any treatment owing to absence of symptoms and unfulfilled diagnosis of an autoimmune disease.

However, the patient came back to our hematology clinic 3 years later as abnormal location of spleen in addition to splenomegaly was found on sonography by a local physician. We arranged an abdomen computed tomography (CT) scan for her, which showed a wandering spleen lying in the pelvic cavity and compressing the bladder. There were no other abnormalities on clinical symptoms and laboratory test. The patient was referred to a hepatobiliary surgeon who suggested surgical intervention to her, but she seemed to be undecided.

Discussion

Wandering spleen results from laxity of the ligaments that normally fix the spleen, and can be attributed to congenital or acquired causes. Besides children, it is mainly found in women at reproductive ages, which suggests the effect of hormonal changes and multiparity on the elasticity of ligaments. Notably, splenomegaly arising from any medical condition is another acquired factor, while it may also be a presentation caused by parenchymal congestion in a wandering spleen. However, it is difficult to determine a precise etiology in our case. It may not be a congenital abnormality judging from the age of the patient. Additionally, we could not find her obstetric history on the medical record. As for the splenomegaly, we assumed it is a resultant sign of a wandering spleen rather than a cause as we could not perfectly refer splenomegaly to any known clinical finding.

Regardless of the etiology, we believed that surgery is the optimal option to the patient. The long vascular pedicle of wandering spleen is predisposed to splenic torsion, while repeated torsion may cause recurrent abdominal pain. Furthermore, splenic infarction, splenic vein thrombosis and pancreatitis are all reported to be potential complications. These conditions can only be prevented by the surgical process, and splenopexy may be favorable for the patient since it

preserved the spleen and is the first-line treatment for asymptomatic cases. The patient should realize that she might take a risk of splenectomy once infarction or acute abdomen occurs in the future.

Conclusion

Being a rare condition, wandering spleen can result from several acquired etiologies. Surgery seemed to be the only way to prevent associated complications, and preservation of spleen is available if the condition managed early.

病例報告 113_C113

服用滴雞精後上肢和頸部出現不尋常硬塊：一個罕見的腫瘤性鈣化症病例報告

Unusual hard masses in upper limbs and neck after chicken essence consumption: A rare case of tumoral calcinosis

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Introduction

Tumoral calcinosis is an uncommon complication associated with end-stage kidney disease(ESKD) and chronic kidney disease(CKD) that affects bone and mineral metabolism. It is characterized by the deposition of calcium phosphate crystals in soft tissue, bone, and periarticular spaces[1]. The incidence of tumoral calcinosis is rare and estimated between 0.5 and 7% of patients in hemodialysis patients[2].

We report a 48 years old female with ESKD who developed tumoral calcinosis following kidney transplant graft failure and back to long-term hemodialysis.

Case Report

A 48 years old female developed several painful and firm mass over her left elbow and wrist area after hemodialysis for 18 months. Over time, the mass gradually progressed to her left neck. Laboratory analysis showed serum calcium of 5.35mg/dL(4.6~5.32mg/dL), serum phosphorus of 6.61mg/dL(2.5~4.5mg/dL), and parathyroidism hormone of 598.7pg/mL(15~65pg/mL). Her medications include Calcium Carbonate, Aluminum Hydroxide, and intravenous Calcitriol during every hemodialysis session. At first, she was diagnosed with gouty arthritis with tophus formation, and oral febuxostat 40mg daily was prescribed, but the treatment proved unsuccessful. Further computed tomography scans revealed multiple amorphous calcification at subcutaneous layer abutting left medial aspect of wrist, elbow joint, and left transverse process around cervical spine. Surgical excision for the mass confirmed the lesions to be pathologic calcification.

Her medical history revealed she was a recipient of cadaveric kidney transplantation for 13 years prior to hemodialysis. Before transplantation, she received peritoneal dialysis for 2 years. A dietary review indicated that the patient consumed chicken essence after each hemodialysis session in recent one year, which contributed to her refractory hyperphosphatemia.

As a result of dietary modifications and oral phosphate binder prescription, her hyperphosphatemia was under control. We discontinued Calcitriol and calcium-containing medications. There was no further growth of existing lesions nor was there any further development of new lesions. Currently, the patient is preparing for a parathyroidectomy in order to address her persistent hyperparathyroidism.

Discussion

Tumoral calcinosis is classified into primary, caused by genetic mutations in phosphate metabolism genes, and secondary, resulting from conditions like chronic kidney disease or hyperparathyroidism[3]. Diagnosis involves clinical evaluation, imaging, and blood tests[4]. Treatment varies based on type and may include dietary phosphate restriction, phosphate-binding

medications, or surgical excision[4].
Our patient performed genetic analysis with no specific gene mutation detected. Chicken essence, rich in phosphate, is commonly used in Asia to combat fatigue. The existing tertiary hyperparathyroidism due to chronic dialysis and CKD coupled with a high phosphate diet and hyperparathyroidism management promotes tumor calcinosis in this case.

Conclusion

In patients with ESKD undergoing hemodialysis, consumption of chicken essence may lead to refractory hyperphosphatemia. When combined with Calcitriol use for hyperparathyroidism can potentially result in severe tumoral calcinosis. Careful medication management and dietary control are crucial for managing this condition. Surgical excision is an effective means of treating symptoms while also providing specimens that can be used for conclusive tissue analysis.

病例報告 113_C114

年輕女性罕見的肺動脈肉瘤 — 一個需要和急性肺栓塞鑑別的疾病

Intimal Sarcoma in a Young Female Patient – A Disease that Needs to be Differentiated from Acute Pulmonary Embolism

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Introduction

Intimal sarcoma (IS) is a rare mediastinal tumor with poor prognosis. The clinical presentation of IS is unspecific. Patients may present with dyspnea, cough, hemoptysis, or chest pain, which are all classic symptoms associated with acute or chronic pulmonary thromboembolism. We present a unique case of a 26-year-old woman with unspecific symptoms and a CT scan showing a large mediastinal tumor involving the right pulmonary trunk.

Case Report

A previously healthy 26-year-old woman presented with mild chest tightness and blood-tinged sputum for days. Chest X-ray revealed a large mass at the right hilar region. Chest CT with enhancement disclosed a 6.5 cm solid mass in the central zone of the right lung. The lesion extended to the adjacent hilum and mediastinum, occluding the segmental bronchus of the right middle lobe. It also caused thromboembolism at the right main pulmonary artery and segmental arteries. EBUS-TBNA of 10R lymph node and RML tumor with transbronchial biopsy were both performed. Microscopic examination of the specimen showed hypercellular and poorly differentiated spindle cells. These cells exhibited marked nuclear atypia and increased mitotic figures in fascicular pattern. The immunohistochemistry test was notable for positive staining for MDM2, CDK4 (focal reactivity), FLI-1, and negative for CK, TTF-1, p40, S100, SS18-SSX, synaptophysin, CD31, desmin, and myoD1 stains. A diagnosis of sarcoma was made. Differential diagnosis included intimal sarcoma (IS) and dedifferentiated liposarcoma (DD-LPS). Since a surgical biopsy could not be carried out in this case, a clinical diagnosis favoring intimal sarcoma was made based on the location of the tumor, which was the pulmonary artery, whereas DD-LPS more commonly arises from the retroperitoneum. Subsequent PET/CT scan also showed no extrapulmonary malignancy suggesting other primary tumor, which further strengthened the diagnosis of intimal sarcoma.

Discussion

Intimal sarcoma (IS) is a rare mediastinal tumor that often obstructs the vascular lumens, resembling acute or chronic thromboembolism, just like the images presented in this case. It usually occurs in patients with a mean age of 40-50 years, and has a slight female predominance. The tumor commonly develops from the pulmonary artery, and can also arise from the aorta, the right ventricular outflow tract, and the heart valves. The reported incidence of IS was very low, around 0.001-0.03%, possibly owing to the major limitations on histopathology and radiology in the past. IS should also be differentiated from the hematogenous spread of metastatic sarcoma in other parts of the body. In this case, based on the whole-body PET/CT scan and the patient's

history, no evidence suggested another primary site of sarcoma, which further solidified the diagnosis of primary pulmonary sarcoma.

Conclusion

Even though intimal sarcoma usually emerges in middle-aged patients, it can still occur in younger patients, as seen in the presented case. Given the aggressiveness of the disease, a misdiagnosis with pulmonary vascular disease and treated as such may not only delay the proper treatment, but also cause detrimental effects on the patient. With the current advancements in diagnostic tools, clinicians should raise more awareness on correctly identifying intimal sarcoma.

病例報告 113_C115

腎臟移植患者罹患擴散性帶狀疱疹後出現嚴重皮膚壞死與結痂：一例罕見且令人警惕的病例

Severe skin necrosis and crusting following disseminated herpes in a kidney transplant patient: A rare and alarming case

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Introduction

Kidney transplantation significantly improves life quality and survival rates for patients with end-stage kidney disease. However, life-long immunosuppression increases the risk of infections. It leads to decreased cellular immunity and increased risk of developing atypical infection, including herpes zoster from reactivation of latent varicella zoster virus. Disseminated herpes zoster in transplant recipients can be fatal. This report discusses a rare and severe case of disseminated herpes zoster in a kidney transplant recipient.

Case Report

A 57-years-old man suffered from generalized vesicles and erythematous papules on the trunk and limbs, which were diagnosed as disseminated herpes zoster. Prior to this, he had received a cadaveric kidney transplant ten months ago and was maintained on tacrolimus, everolimus, and mycophenolate acid for immunosuppression at the time. Before kidney transplantation, he has been receiving hemodialysis treatment for eleven years. Additionally, he received Dupilumab 300mg every 2 weeks for refractory atopic dermatitis at the same time. Despite initial antiviral treatment, his skin condition worsened, leading to necrotic ulcers with superimposed bacterial infection. In addition to broad-spectrum antibiotics, the patient's immunosuppression was minimized to low doses of tacrolimus and prednisolone. After aggressive wound care, his clinical condition improved, and he was discharged as stable.

Discussion

In kidney transplant recipients, the incidence of herpes zoster is higher than in immunocompetent individuals. Therefore, kidney transplant recipients are at an increased risk of serious herpes zoster complications. Considering the patient's immunosuppression and previous history of dialysis, he was at risk for virus infection. The prescription of Dupilumab aggravated his condition and resulted in disseminated herpes zoster. Everolimus, with its antiproliferative nature, prevented wound healing of his rupture vesicles, contributed to the severity of the patient's skin condition. As a result, it is imperative that immunosuppressive therapy be adjusted carefully in transplant patients to balance infection risks with graft rejection prevention. Dupilumab is beneficial for the treatment of atopic dermatitis, but its immunomodulatory effects should be taken into consideration when using it in kidney transplant recipients.

Conclusion

In this case, disseminated herpes zoster manifests severe consequences in a kidney transplant

patient, emphasizing the importance of vigilant monitoring and the careful evaluation of additional immunomodulatory agent's therapy. For complex cases such as this, the interplay between immunosuppression and infection control is crucial, underscoring the need for a multidisciplinary approach to ensure the best possible outcome for the patient.

病例報告 113_C116

擬似闌尾腫瘤的原發性盲腸瀰漫性大型 B 細胞淋巴癌

A case report of primary diffuse large B-cell lymphoma of the cecum mimicking appendiceal neoplasm

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Introduction

Extranodal lymphoma refers to lymphoma originating outside the lymph nodes. The gastrointestinal (GI) tract is the most common site of secondary extranodal non-Hodgkin lymphoma (NHL), though primary GI lymphomas are rare, comprising only 1-4% of malignancies in the stomach, small intestine, and colon. Among these, primary colorectal lymphoma is particularly uncommon, making up just 0.2%-1% of all colonic cancers. In this report, we present a 64-year-old male patient who was initially suspected to have an appendiceal neoplasm, but was ultimately diagnosed with diffuse large B-cell lymphoma (DLBCL). He was successfully treated with chemotherapy.

Case Report

A 64-year-old man with no underlying health conditions experienced right lower abdominal pain for three months. However, after visiting National Taiwan University Hospital Hsin-Chu for symptoms control, his pain persisted, and he developed right thigh numbness. He then visited the emergency department at Taitung Hospital. A computed tomography (CT) scan performed on December 17, 2023, revealed a large mass (6.9 × 5.7 × 3.5 cm) in the right lower quadrant, adjacent to the cecum, with irregularities in the appendix, fluid accumulation, and a thickened wall. Metastatic lymph nodes in the right common iliac region suggested appendiceal neoplasm. Laboratory tests indicated elevated C-reactive protein and white blood cell levels. A diagnosis of appendiceal adenocarcinoma presenting as acute appendicitis was considered. The patient underwent a laparoscopic right hemicolectomy with ileo-colic anastomosis on December 21, 2023. However, histopathology revealed a high-grade B-cell lymphoma, favoring DLBCL of the germinal center B-cell subtype. Immunohistochemical results showed positive markers for CD20, BCL-6, and a Ki-67 index of >95%, confirming the diagnosis. He was treated with intravenous methylprednisolone and underwent a bone marrow biopsy, which showed no lymphomatous involvement. Postoperatively, he developed intra-abdominal infection and septic shock, which improved after treatment. He completed six cycles of R-CHOP chemotherapy (rituximab, cyclophosphamide, doxorubicin, vincristine, prednisolone) in the next visits.

Discussion

Primary colorectal lymphomas are rare, representing 0.2%-1% of colonic malignancies. Although extranodal NHL frequently involves the GI tract, it most often affects the stomach, followed by the small intestine and, rarely, the colon. DLBCL is the most common subtype of extranodal NHL, characterized by its aggressive nature and rapid proliferation.

Symptoms of GI lymphoma are often nonspecific and may mimic other conditions, as in this case,

where the mass initially suggested an appendiceal neoplasm. A definitive diagnosis requires histopathological examination and immunohistochemical analysis. In this patient, the tumor cells were positive for CD20 and BCL-6, with a high Ki-67 index, indicating a highly proliferative DLBCL. The standard treatment for DLBCL is the R-CHOP chemotherapy regimen, which was administered to our patient following surgery. Recent studies suggest that combining surgical resection with chemotherapy can improve outcomes in localized intestinal DLBCL, as was demonstrated in this case.

Conclusion

Primary non-Hodgkin lymphoma of the colon is rare, with nonspecific symptoms that complicate early diagnosis. Histopathology and immunohistochemistry are crucial for diagnosis, and the combination of surgery and chemotherapy appears to provide favorable outcomes in localized cases.

病例報告 113_C117

臺灣成人罕見全身性破傷風病例報告

A Rare Case Report of Generalized Tetanus in an Adult Patient in Taiwan

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Introduction

Tetanus is rare in resource-rich countries like Taiwan due to effective vaccination programs. However, life-threatening generalized tetanus can still occur in adults, with early symptoms such as sweating, tachycardia, and localized muscle spasms frequently being overlooked. We present a case of generalized tetanus to raise awareness and share our clinical management experience. This report also underscores the crucial importance of adhering to scheduled tetanus toxoid booster vaccinations following the primary immunization series.

Case Report

We report the case of a 76-year-old Taiwanese male who presented to the emergency department with painful jaw spasm, dysphagia, and worsening of a left ankle avulsion wound sustained in a motor vehicle accident 4 days prior. On admission, he tested positive for SARS-CoV-2 via PCR and was placed in isolation during the COVID-19 pandemic. Despite prompt initiation of metronidazole and benzodiazepines for suspected tetanus, the patient rapidly progressed to trismus, opisthotonos, and acute respiratory failure by hospital day three. Due to a difficult airway, an urgent cricothyrotomy was performed, followed by mechanical ventilation. After emergent wound debridement, the patient was transferred to the intensive care unit (ICU). Continuous infusions of metronidazole, atracurium, and oral clonazepam provided partial relief, but persistent chest wall rigidity and muscle spasms remained. Notably, patient-ventilator dyssynchrony was significantly improved with the initiation of continuous magnesium sulfate infusion. The patient was eventually discharged after a 64-day hospital course, including 39 days in the ICU.

Discussion

Victims of tetanus typically present with trismus, generalized hypertonia, painful muscle spasms, and autonomic dysfunction. Acute respiratory failure is a leading cause of mortality in tetanus cases. In the case we presented, a timely emergency cricothyrotomy was fortunately performed, preventing a fatal outcome. Despite tetanus being rare in resource-rich countries like Taiwan, clinicians must remain vigilant for tetanus-related symptoms in individuals with recent contaminated wounds, particularly those who have not received scheduled tetanus toxoid booster vaccinations following their primary immunization. Close monitoring of respiratory patterns and early consideration of intubation are crucial in generalized tetanus due to the high risk of a difficult airway. Based on our experience, continuous magnesium sulfate infusion plays a significant role in alleviating muscle spasms and managing autonomic dysfunction.

Conclusion

Tetanus is rare in resource-rich countries such as Taiwan, but when it does occur, it can be life-

threatening. This case of generalized tetanus underscores the need for heightened clinical awareness and emphasizes the importance of early intubation and continuous magnesium sulfate infusion in management. It also highlights the critical importance of adhering to scheduled tetanus toxoid booster vaccinations to prevent such cases.

病例報告 113_C118

一位發燒合併心包膜積液的罕見個案: 自體免疫性疾病相關的心包膜炎

A rare case with fever and pericardial effusion: autoimmune diseases associated pericarditis

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Introduction

Pericardial effusion may arise from infection, such as viral diseases, HIV (human immunodeficiency virus) or tuberculosis, inflammation, trauma leading to blood fulfilling into pericardial space, or malignancies. Autoimmune diseases are an uncommon but important cause of pericardial effusion. We reported a rare case manifesting fever with incidentally found pericardial effusion, and diagnosed as an autoimmune diseases-associated pericarditis.

Case Report

A 77-year-old female with hypertension and thyroid goiter presented with intermittent fever, general weakness, decreased appetite and body weight, and mild dyspnea on exertion for 2 weeks. She also mentioned right lower leg reddish swelling and being treated as cellulitis at local medical department. She denied special occupational or exposure history. Physical examination showed body temperature 37.8°C, heart rate 97/bpm, respiratory rate 18/cpm, blood pressure 101/62 mmHg, and saturation 99% under room air. The electrocardiography was sinus rhythm with paroxysmal atrial fibrillation. The computer tomography revealed pericardial effusion with bilateral pleural effusion. The echocardiography showed moderate amount of pericardial effusion. Laboratory test showed elevated level of C-reactive protein (69.6 mg/L). The surgery of pericardial window, pericardiectomy and pericardial effusion drainage was performed. The analysis of pericardial effusion demonstrated pleocytosis with monocyte predominant (96%), and elevated level of lactate dehydrogenase (958 IU/L). The nucleic acid amplification test of mycobacterium tuberculosis was not detected. No special microorganism was isolated. The cytology of effusion was negative for malignancy. The pathology of pericardium was granulomatous inflammation. Considering inflammatory diseases, we investigated the antinuclear antibodies and scleroderma panel. The autoimmune profiles revealed positive of antinuclear antibody (1:320, nucleolar pattern), anti-Ro>240, anti-La 30, anti-Histone strong positive, anti-Scl-70(2+), anti-Nor90(3+), and anti-Ro-52(3+). Therefore, hydroxychloroquine and prednisolone 15mg daily were prescribed. Fever subsided, and both the pericardial effusion and pleural effusion reduced in the followed chest x-ray. The patient discharged under relative stable condition.

Discussion

Autoimmune diseases which may involve the pericardium include systemic lupus erythematosus, systemic sclerosis, dermatomyositis, polymyositis, Sjögren's disease, rheumatic fever and the vasculitis. The most common manifestations are acute or recurrent pericarditis with asymptomatic pericardial effusion. Massive pericardial effusion with cardiac tamponade may be less common but should be noted in late-diagnosed cases. Our case has an exudative effusion,

and positive in anti-Scl-70, anti-Nor 90, and anti-Ro 52 antibodies. The probable diagnoses included systemic sclerosis (SSc) and secondary Sjögren's disease. However, further examination including nailfold capillaroscopy were required for confirmed diagnosis. Although pericardial effusion is not uncommon in systemic sclerosis, cardiac involvement in systemic sclerosis may be associated with poor prognosis and increased mortality.
We report this case to emphasizes the prompt diagnosis is crucial for these autoimmune diseases associated pericarditis.

Conclusion

For patients presenting with fever and pericardial effusion, accurate diagnosis should be made, and autoimmune diseases should be considered in the differential diagnosis, especially when negative cultures or malignancies.

病例報告 113_C119

雙側肺動脈栓塞置放超音波震碎血栓導管治療(EKOS) 後發生大量血性心包積液：疑因新診斷的巨大縱隔腫瘤侵犯心包膜，經抗凝後溶栓引起之大量出血

Massive Bloody Pericardial Effusion Following EKOS (Ekosonic Endovascular System Thrombolysis) Placement for Bilateral Pulmonary Artery Embolism: A Case of Suspected Urokinase and Heparin-Induced Hemorrhage due to Newly Diagnosed Huge Mediastinal Tumor Involvement of the Pericardium

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Introduction

Pulmonary embolism is known as a silent but deadly killer. In recent years, several innovative catheter-based interventional therapies have been developed. An advanced version of the catheter-directed thrombolysis is the use of ultrasound-assisted thrombolysis (EKOS), which enhances the traditional thrombolysis treatment.

However, EKOS thrombolysis still carries bleeding risk. We present a case with massive bloody pericardial effusion after EKOS thrombolysis placement.

Case Report

A 55 years old man denied any systemic disease and was totally independent on daily activities. This time, he had dyspnea, especially on exertion for 2 weeks. He denied peripheral limbs edema, fever, abdomen discomfort or chest tightness. He came to our emergency department for help. At emergency department, tachypnea with mild desaturation was noticed and improved after oxygen used. CTA (computed tomography angiography) showed huge submucosal mass over lower thoracic esophagus and esophago-cardiac junction. Also, thromboembolism over bilateral main pulmonary arteries and segmental arteries of both lungs. Right ventricle engorgement was noticed at bedside echo and elevated troponin I was found through lab examination. Due to intermediate high risk of adverse outcome, we started EKOS thrombolysis.

However, sudden onset air hunger, profound shock was noticed on following morning. We intubated and kept ventilator, ECMO (Extra-Corporeal Membrane Oxygenation) support. Lab examination showed no obvious infection sign or anemia. However, bedside echo showed newly developed massive pericardial effusion. Bedside pericardial drainage was performed immediately. Pericardial drain showed persisted bloody effusion. Currently we kept transfusion and pericardial drainage. He was relatively stable.

Discussion

Pulmonary embolism is known as a silent but deadly killer. Mild cases may present with no significant symptoms, while severe cases can lead to shock or even sudden death. In recent years, several innovative catheter-based interventional therapies have been developed, primarily categorized into two main types: one involves local catheter-directed thrombolysis, and the other uses mechanical thrombectomy. An advanced version of the catheter-directed thrombolysis is the use of ultrasound-assisted thrombolysis (EKOS), which enhances the traditional thrombolysis

treatment.
EKOS thrombolysis uses ultrasound to aid the process by oscillating the blood and thrombolytic agents, improving their mixing and breaking up the thrombus, making it easier and quicker to dissolve while reducing the risk of bleeding. The dosage of thrombolytic agents used is about one-fifth of that in traditional thrombolysis, with a thrombus clearance rate exceeding 90%. This makes it an ideal option for patients with a lower tolerance for bleeding risks.
In our case, after reviewing patient's computed tomography, tumor with micro pericardium invasion was highly suspected, leading to massive pericardium bleeding after EKOS placement.

Conclusion

Though considered relatively lower bleeding risk therapy, EKOS placement still needed to be concern in high bleeding risk circumstance such as tumor with big vessel, pericardium effusion.

病例報告 113_C120

一個車禍鈍挫傷導致的腎動脈栓塞個案成功的藉由導管介入來完成治療

A case of Blunt trauma Related Renal Artery Thrombosis successfully treated by Percutaneous Endovascular Therapy

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Introduction

Renal vascular injury, a rare complication of blunt trauma, is often caused by renal artery thrombosis. Approximately 40% of such cases was caused by thrombosis.[1] The effectiveness of revascularization in improving renal function has been a topic of debate.[2] Percutaneous endovascular therapy (EVT) has gained popularity since 1995 and has shown promising results.[3, 4] In this case, a young adolescent with blunt trauma related renal artery thrombosis underwent percutaneous EVT, which greatly improved the renal function after the intervention.

Case Report

A 15-year-old male adolescent had a traffic accident on 2023/2/2. He was riding a motorcycle and collided with a car, experiencing temporary loss of consciousness. He was taken to Ministry of Health and Welfare Pingtung hospital, where initial tests showed multiple fractures and injuries including rib fracture, facial fracture, and arm fracture with dislocation. He was then transferred to our hospital for further treatment.

At our hospital, he had mild hypotension, tachycardia, and complained of abdominal and low back pain. Lab tests showed elevated liver enzymes and creatinine. CT scans revealed liver laceration, splenic laceration, and bilateral renal infarction. (Figure 1) We performed angiography and EVT with stenting to treat the renal artery occlusion. (Figure 2-6) After the procedures, renal blood flow improved and the patient was kept on dual-antiplatelet medication. (Figure 7)

He was monitored in the pediatric intensive care unit and had good urine output without electrolyte imbalance. His creatinine levels initially rose but eventually returned to normal. He was discharged with improved kidney blood flow and prescribed aspirin for continued treatment.

Discussion

Our case of blunt trauma related renal artery thrombosis was successfully treated with endovascular intervention and stent placement. Prompt intervention before tissue necrosis is believed to play a significant role in improving renal function.

Conclusion

We demonstrated a young adolescent with blunt trauma related renal artery thrombosis, whose renal function improved very well after percutaneous endovascular angioplasty and bare metal stent placement. Though the role of revascularization for such cases still needs further larger study for illustration, endovascular intervention may play an important role in cases of blunt trauma-related renal artery thrombosis.

病例報告 113_C121

以慢性腹瀉表現的嗜伊紅性腸胃道疾病

A case report of eosinophilic gastrointestinal diseases presented with chronic diarrhea

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Introduction

Eosinophilic gastrointestinal diseases are chronic, immune-mediated disorders characterized by eosinophil-predominant tissue inflammation and gastrointestinal symptoms. The disease refers to a group of conditions including eosinophilic esophagitis (EoE), eosinophilic gastritis (EoG), eosinophilic enteritis (EoN), and eosinophilic colitis (EoC), with 50-70% of patients having an allergic disease including asthma, defined food sensitivities, eczema, or rhinitis. Clinical manifestation varies and includes abdominal pain, diarrhea, nausea, vomiting, and weight loss. Diagnosis typically involves endoscopic evaluation and eosinophilic ascitic fluid, with eosinophilic infiltration of the gastrointestinal tract on biopsy, and exclusion of other causes of intestinal eosinophilia.

Case Report

A-64-year-old female previously diagnosed with stage I colorectal cancer post low anterior resection, presented to the emergency department with complaints of chronic diarrhea for months, associated with epigastric pain for one month, nausea, vomiting, and poor appetite. Peripheral parenteral nutrition was initiated for nutrition support. Abdominal computed tomography showed suspect mucosal thickening in the colon. Gastric emptying test demonstrated significant gastric emptying prolong, suggestive of gastroparesis. For positive fecal occult blood test, EGD (esophagogastroduodenoscopy) was performed and revealed hemorrhagic and nodulated gastritis in body and antrum and gastric shallow ulcers. Pantoprazole was prescribed to manage the gastric ulcers, and Otilonium Bromide was given to control diarrhea. Rheumatologic lab data survey revealed negative findings. After treatment, she had better appetite and smooth defecation, without nausea, vomiting, or abdominal pain. She was then discharged with oral medications control.

However, one month after discharge, she experienced a recurrence of aggravated diarrhea, with mucus and bloody content. Follow-up EGD (esophagogastroduodenoscopy) showed hemorrhagic and nodulated gastritis over low body and antrum. Pathology revealed chronic gastritis with eosinophilic infiltration. Colonoscopy revealed pancolitis with pseudopolyps and shallow ulcers, sparing the terminal ileum, and pathology revealed eosinophilic infiltration in terminal ileum, colon, and rectum. These findings led to a diagnosis of eosinophilic gastritis (EoG) and eosinophilic colitis (EoC). Therefore, we added oral prednisolone for control, with gradual titration. Laboratory data showed normal IgE level, normal eosinophil count, and positive allergy test especially with egg products. The diarrhea was improving following the initiation of steroid therapy, and the prednisolone dose was gradually tapered and discontinued in three months. We also added Montelukast, a leukotriene antagonist, to her treatment regimen. EGD (esophagogastroduodenoscopy) for post-treatment follow-up showed multiple gastric polyps with shallow ulcers and duodenitis

with nodularity, and the eosinophilic infiltration was improving. However, two months after discontinuing steroids, the patient experienced recurrent diarrhea. As a result, we added back low dose prednisolone (5mg/day) and kept Montelukast for eosinophilic gastritis and colitis. Her diarrhea was better controlled under current medical treatment.

Conclusion

The case demonstrated the diagnosis and management of eosinophilic gastritis (EoG) and eosinophilic colitis (EoC), with presentation of chronic gastrointestinal symptoms, that led to further investigation, ultimately revealing significant eosinophilic infiltration. Treatment for these conditions generally involves dietary modifications, corticosteroids, and in some cases, leukotriene inhibitors like Montelukast or biologic therapies such as Lirentelimab, Cromolyn, or Omalizumab, aimed at controlling inflammation and preventing recurrence. Our patient initially responded well to steroid therapy, but symptoms recurred following steroid discontinuation. The reintroduction of low-dose steroids alongside Montelukast resulted in a favorable treatment response. Such experience underscores the early recognition and tailored treatment for eosinophilic gastrointestinal disorders, as well as the need for long-term treatment strategies to manage recurrent symptoms effectively.

病例報告 113_C122

以吐血表現的食道靜脈瘺管

A rare case presented with hematemesis diagnosed as aortoesophageal fistula (AEF)

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Introduction

Aortoesophageal fistula (AEF) is a rare but fatal cause of upper gastrointestinal bleeding. In this case, the patient's fistula was likely secondary to her mycotic aneurysm, which caused esophageal erosion and rupture. AEF has a mortality rate exceeding 90% without timely intervention. We report a case of a 74-year-old female with an underlying disease of chronic kidney disease (CKD) stage 5, coronary artery disease (CAD) post-coronary artery bypass grafting (CABG), heart failure with reduced ejection fraction (HFrEF), and type 2 diabetes mellitus (T2DM). She presented with hematemesis and then finally diagnosed as ortoesophageal fistula (AEF), likely secondary to mycotic aneurysm.

Case Report

A 74-year-old female with an underlying disease of chronic kidney disease (CKD) stage 5, coronary artery disease (CAD) post-coronary artery bypass grafting (CABG), heart failure with reduced ejection fraction (HFrEF), and type 2 diabetes mellitus (T2DM). Her activities of daily living are totally independent.

She presented with dyspnea, chest pain, back pain, poor appetite, and progressive dysphagia over the past 10 days. There was no allergy to NSAID (nonsteroidal anti-inflammatory drug). She denied any alcohol, betel nut, cigarette use. She also denied cancer history in her family.

Initially, Physical examination revealed an elevated heart rate (95 beats per minute), low blood pressure (98/54 mmHg), and reduced oxygen saturation (SpO2 88%). No signs of fever were noted, and respiratory and heart sounds were normal upon auscultation. Initial blood tests revealed elevated white blood cell (WBC) count, anemia (hemoglobin 8 g/dL), elevated blood urea nitrogen (BUN) at 123.7 mg/dL, and hyperkalemia (potassium 5.8 mmol/L). The arterial blood gas analysis indicated metabolic acidosis with a pH of 7.13 and a bicarbonate level of 10.7 mmol/L. A posteroanterior chest radiograph showed no signs of consolidation, lobar collapse, or masses. Esophagogastroduodenoscopy(EGD) was arranged, revealing a gastric ulcer (Forrest IIc classification), a common finding in patients with upper gastrointestinal bleeding. Conservative management was initiated, including the administration of proton pump inhibitors (PPI), empiric antibiotics, and supportive care for her kidney failure. The clinical status got worse after two days after admission. A chest computed tomography angiogram (CTA) was ordered on November 9 due to worsening clinical status, revealing a mycotic aneurysm at the descending aorta with esophageal rupture into the mediastinum and abscess formation. This life-threatening condition, known as an aortoesophageal fistula (AEF), was responsible for the recurrent episodes of hematemesis and severe blood loss.

The patient was deemed a candidate for thoracic endovascular aortic repair (TEVAR), a minimally invasive procedure that has replaced open surgery as the first-line treatment for AEF.

Unfortunately, despite the successful control of the hemorrhage and stabilization with TEVAR, the patient experienced a rapid clinical decline. On the same day, following a massive oral and nasal hemorrhage, she suffered cardiac arrest. Resuscitation efforts were unsuccessful, and the patient expired after 30 minutes of cardiopulmonary resuscitation.

Discussion

Aortoesophageal fistula (AEF) is a rare but fatal cause of upper gastrointestinal bleeding. In this case, the patient's fistula was likely secondary to her mycotic aneurysm, which caused esophageal erosion and rupture. AEF has a mortality rate exceeding 90% without timely intervention. The hallmark of AEF is Chiari's triad, which includes mid-thoracic pain, sentinel hemorrhage, and exsanguination after a symptom-free interval. In this patient, sentinel hemorrhage occurred multiple times before the final, massive bleeding episode.

Conclusion

This case illustrates the complexity and high mortality associated with aortoesophageal fistula in the context of gastrointestinal bleeding. The diagnosis of AEF is often delayed, and early intervention with procedures like TEVAR can be life-saving. However, the prognosis remains poor, particularly in patients with advanced comorbid conditions like CKD and CAD. This case highlights the importance of early recognition and multidisciplinary management in patients with gastrointestinal hemorrhage, especially when rare conditions like AEF are involved.

病例報告 113_C123

在 EGFR TKI 標靶治療後，使用次世代基因分析找到罕見且可治療的 FRK-ROS1 融合基因為抗藥機轉

A rare but druggable FRK-ROS1 fusion gene as a resistance mechanism under EGFR TKI treatment identified through next-generation sequencing

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Introduction

ROS proto-oncogene 1 (ROS1) gene rearrangements occur in approximately 1% to 2% of patients with non-small cell lung cancer (NSCLC). Among these, fyn-related kinase (FRK) is an extremely rare fusion partner with ROS1. While ROS1 is typically considered a primary driver mutation, it is uncommon for ROS1 fusion genes to serve as a resistance mechanism. Using next-generation sequencing (NGS), we identified a novel FRK-ROS1 rearrangement in a stage IIIB NSCLC patient after developing resistance to afatinib monotherapy. Transitioning to a combination of crizotinib and gefitinib yielded an excellent response.

Case Report

A 56-year-old nonsmoking woman presented with a 3-month history of chronic dry cough. Chest radiography revealed opacities in the right upper and lower lobes. A contrast-enhanced computed tomography (CT) scan of the chest showed a spiculated nodule (3.9 cm in diameter) in the right lower lobe, with ipsilateral hilar and mediastinal lymphadenopathy. Positron emission tomography-CT (PET-CT) revealed 18F-fluorodeoxyglucose (FDG) avidity in the right lower lobe, as well as in the left hilar and mediastinal lymph nodes. Magnetic resonance imaging (MRI) of the brain showed no evidence of metastases. A transbronchoscopic biopsy of the lung mass confirmed lung adenocarcinoma. Genetic testing revealed an EGFR mutation (L858R) and ROS1 rearrangement, and the clinical stage was determined to be cIIIB (cT3N2M0). She was initially treated with afatinib, an EGFR TKI, and tolerated the treatment well.

After 7 months, however, a follow-up CT scan revealed disease progression, including tumor enlargement and new metastases within the ipsilateral lobe. A repeat transbronchoscopic biopsy of the lung tumor was performed, and tissue NGS analysis identified a rare FRK-ROS1 rearrangement, in addition to the coexisting EGFR L858R and TP53 mutations. Her treatment was then switched to a combination of crizotinib and gefitinib. A follow-up chest radiograph three weeks later showed partial tumor resolution.

Discussion

FRK-ROS1 is a novel ROS1 fusion variant first reported in advanced NSCLC with brain metastases in 2021. In that case, treatment with crizotinib resulted in a partial response within 1 month, which was maintained for almost 8 months without severe adverse effects.

ROS1 rearrangements lead to dysregulated ROS1 kinase activity and inappropriate downstream signaling. They have been observed predominantly in female patients, non-smokers, and in advanced stages of NSCLC. The oncogenic properties of different ROS1 fusion kinases remain

unclear. Crizotinib, a first-generation tyrosine kinase inhibitor (TKI), is approved for use in ROS1-positive and anaplastic lymphoma kinase (ALK)-positive metastatic NSCLC, with a median progression-free survival (PFS) of 15 to 20 months.
The growing use of NGS in NSCLC has revealed an increasing number of co-mutations involving genes such as EGFR, ALK, and ROS1. The optimal treatment strategy for patients with multiple actionable mutations remains a subject of ongoing debate, but combination therapy targeting co-mutations offers a promising approach.

Conclusion

We report a case of advanced NSCLC with coexisting EGFR mutation and FRK-ROS1 gene fusion. NGS played a pivotal role in identifying the complex molecular landscape of the tumor upon progression, guiding subsequent targeted therapy and leading to a favorable clinical response.

病例報告 113_C124

使用 PET/CT 輔助不明熱之診斷—在一個以發燒與骨髓抑制作為表現的瀰漫性結核菌感染病例之應用

PET/CT in the Diagnosis of Fever of Unknown Origin: A Case of Disseminated Tuberculosis with Fever and Bone Marrow Suppression

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Introduction

The diagnosis of fever of unknown origin to infectious specialist is a big challenge. Evaluation of FUO should begin with a thorough history taking, examination, and the initial diagnostic testing. Positron emission tomography–computed tomography (PET-CT) apply in FUO survey and even treatment response of tuberculosis has more and more studies.

Case Report

A 81-year-old man with a history of hypertension, liver cirrhosis, COPD, Alzheimer's disease and benign prostatic hypertrophy was admitted because of fever for 5 days. He suffered from persistent fever and malaise without obvious discomfort. Laboratory studies revealed leukopenia (1500/μl; neutrophils :62.9% and lymphocytes: 20.5%), anemia (Hemoglobin: 12.4 gm/dL) and thrombocytopenia (Platelet: 117000/ul). After admission, bone marrow biopsy was performed for pancytopenia and fever of unknown origin. Pathology of bone marrow of iliac bone documented noncaseating granuloma. Chest X ray disclose bilateral lower lung ill-defined opacity. Self-paid F-18 FDG PET/CT whole body scan showed suspected pulmonary, splenic, lower thoracic vertebral lesions.

The patient's sputum acid fast stain discloses negative for 3 sets. Bone marrow tissue MTB-PCR disclose Non-detected of MTBC DNA. We highly suspicious of tuberculosis infection according to bone marrow pathology report and PET-CT multiple abnormal lesions, HERZ was administered. Sputum TB Culture growth Mycobacterium tuberculosis 12 days after started HERZ. After Anti-TB Drugs treatment, the patient's fever subsided and pancytopenia recovery.

Discussion

The diagnostic of FUO yield of FDG PET-CT appears to be more than 50%, and the yield is at least 30% greater than that of conventional CT. However, FDG PET-CT cannot reliably differentiate active TB lesion from malignant lesions and false positives can also be due to other infective or inflammatory conditions. Diagnosis tuberculosis still need microbiology proof and pathology document. According to abnormal lesion from PET-CT to get microbiology culture and tissue pathogen can have more disease information to help diagnosis.

Conclusion

PET-CT can give more information and help diagnosis to fever of unknown origin.

病例報告 113_C125

甲狀腺亢進以急性肝炎表現: 個案報告

Hyperthyroidism present as acute hepatitis: Case report

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Introduction

Hyperthyroidism is a common disease but present with acute hepatitis as first clinical manifestation is uncommon. Hyperthyroidism impacts multiple system. Liver dysfunctions are found in between 15% and 79% of untreated hyperthyroidism patients. About 1% to 2% have fulminant hepatitis. The prevalence of liver dysfunction with respect to alanine transaminase (ALT), aspartate transaminase (AST), alkaline phosphatase (ALP), total bilirubin (BIL), and γ-glutamyl transferase (GGT) among the hyperthyroid patients were 33%, 23%, 44%, 12%, and 24% respectively. Here we report a case of hyperthyroidism prevent with acute hepatitis and anemia.

Case Report

A 36y/o female suffered from general malaise, nausea, vomiting, dull upper abdomen pain and mild fever for 3 days. She is well before, no herbs or private medication, no systemic disease, and denied blood or sex exposure and travel recently. She called on emergent department and was admitted due to acute hepatitis. Liver function showed AST: 212 U/L (reference range: 15–39). ALT: 242 (7– 45). Total bilirubin: 1.21mg/dL (0.3-1.2); Direct bilirubin: 0.46 mg/dL (0.0-0.4). RBC: 4.87 x10^6/ul. Hb: 8.7 gm/Dl. Hct: 29.2 % Platelet: 260x10^3/ul. Physical examination reveal Gr 2 goiter but no apparent proptosis. Thyroid function showed TSH<0.005 uIU/mL (>18 歲:0.55-4.78) Serum T3: 245.47ng/dL (60-181) Free T4: 2.12 ng/dL (0.89-1.76). Thyroid echography showed the typical feature of Graves' disease. Gastroendoscopy found gastric ulcer without bleeding. Abdomen echography found: marked hepatosplenomegaly. After 3 months of antithyroid medications and attainment of mild hyperthyroidism, liver function was in normal range, include AST, ALT, bilirubin, CBC data. Hepatosplenomegaly also reduced size.

Discussion

Putative Mechanisms for Liver Dysfunction in Hyperthyroidism include direct toxicity from excessive thyroxin, free-radical damage, liver cell degeneration, autoimmune-related liver injury, congestive hepatopathy, previous liver disease and antithyroid medication-related toxicity. So far, studies have not demonstrated a correlation between abnormal liver biochemical tests and thyroid hormone levels. Recent review study showed that following the initiation of antithyroid medications and attainment of euthyroidism, there was normalization abnormalities in ALT, AST, ALP, BIL, and GGT in 83%, 87%, 53%, 50%, and 70% respectively.

Conclusion

Hepatic dysfunction associated with thyrotoxicosis is common finding in clinical practice. The exact mechanisms are still unknown. Examination of liver function in the setting of

hyperthyroidism is important to identify any abnormalities; timely initiation of antithyroid medication generally results in improvement of complications and prevent or minimize multi-organ dysfunction. Clinicians should maintain a high index of suspicion for underlying hyperthyroidism in patients presenting with unexplained liver dysfunction or unexplained jaundice.

病例報告 113_C126

老年人特發性血色素沉著症以低血糖呈現：病例報告

Idiopathic hemochromatosis present as hypoglycemia in the aged: a case report

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Introduction

Hemochromatosis is a genetic disorder characterized by increased iron storage in various organs with progressive multisystemic damage. The diagnosis of hemochromatosis poses a challenge to clinicians due to its non-specific symptoms and indolent course causing significant delay in disease recognition. The key organ that is affected by iron overload is the liver, suffering from fibrosis, cirrhosis or hepatocellular carcinoma, complications that can be prevented via early diagnosis and treatment.

Case Report

A 71-year-old male with Hypertension and hepatitis C carrier state in stable condition without treatment. He was sent to ED due to hypoglycemia (68 mg/dL) at home and admitted due to recurrent hypoglycemia (60 mg/dl) after intravenous glucose supplement. Associated symptoms with nausea, weakness, cold sweating, but he denied abdominal pain or diarrhea. Physical examination revealed no apparent finding. The laboratory data revealed postprandial glucose 76 mg/dl, RBC 5.5 x 106/Ul, hemoglobin 16.6 g/d/, MCV 94.2fL platelet 151 x103/ul, ALT 134 U/L, AST 106 U/L, Ferritin 681 ng/mL (refer range 24-330), iron 214 ug/dL (50-212), transferrin 201 mg/Dl (200-360), transferrin saturation 82.6% 20-50). TIBC 259 ug/Dl (200-560), Insulin level > 300 uIU/mL (1.9-23), C-peptide: 14.2 (0.9-4.0), A1c: 5.8 %. Abdominal image with echography, CT and MRI found no apparent lesion. Condition improved after one week treatment and he was discharged.

Discussion

This case suffered from recurrent postprandial hypoglycemia and laboratory test revealed postprandial hyperinsulinemia, hepatitis and hemochromatosis. Insulin resistance related with hemochromatosis was suspected. Liver biopsy may help diagnosis but the patient refuse. Hemochromatosis less presented with hypoglycemia as first clinical manifestation and this manuscript depicts our personal experience with a clinical case that presented with nonclassical symptoms that we had to match to obtain the correct diagnosis. The disease brings into the spotlight the liver involvement, which may be present prior to the hemochromatosis diagnosis and can be misleading as an etiology. Thus, we would like to emphasize the importance of a correct and prompt diagnosis leaning on current tools such as iron serum markers and imaging. Since liver injury is the most problematic organ involvement in hemochromatosis and may progress to fibrosis, cirrhosis, or hepatocellular carcinoma, special attention should be given to this organ's involvement.

Conclusion

Hemochromatosis is an iron storage disease that can be challenging to diagnose in the presence of unspecific symptoms and multiorgan involvement. The multidisciplinary cooperation and early intervention may reverse the course of multiorgan damage and prognosis.

病例報告 113_C127

抗丙型干擾素抗體相關成人免疫缺乏症候群：病例報告及文獻回顧

Anti-interferon Gamma Antibody-Associated Adult Onset Immunodeficiency : A Case Report and Literature Review

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Introduction

Anti-interferon Gamma Antibody (AIGA)-Associated Adult Onset Immunodeficiency is a rare condition. Interferon gamma (IFNN) is crucial for immunity against intracellular bacteria. Genetic defects in its signaling pathway can lead to increased susceptibility to opportunistic infections, including those caused by mycobacteria, Talaromyces marneffei, and Salmonella. Herein, we report a rare case of anti-interferon gamma antibody-associated adult-onset immunodeficiency with disseminated non-tuberculous mycobacterial (NTM) infection.

Case Report

We reported a 49-year-old Taiwanese man with a history of heavy smoking and disseminated non-tuberculous mycobacterial infection, treated in 2021, presented in 2024 with right-sided chest pain, cough, and shortness of breath. Chest CT revealed rapid progression of right lung consolidation and pleural effusion, leading to a diagnosis of right empyema. He underwent Video-Assisted Thoracic Surgery (VATS) and was treated with antibiotics. Post-surgery, he developed septic shock, managed with Levofloxacin and Ceftriaxone, and right pneumothorax requiring pigtail drainage. Cultures showed Candida albicans, leading to antifungal treatment with fluconazole for invasive candidiasis. The patient was successfully extubated and discharged after completing his treatment.

Given his history of severe and recurrent infections, along with his non-tuberculous mycobacterial infection, an anti-interferon gamma antibody test was performed and returned positive.

Discussion

IFN-ussionositive.of severe and recurrent infections, along with his non-tuberculous mycobacterial infection, an anti-interferon gamma antibody test was performed andith right-sided chest pain, cough, and shortness of breath. Chest CT revealed rapid progretivity, which can be done through various methods like ELISA or flow cytometry but can be time-consuming and costly. Despite intensive antimicrobial treatment, AIGA patients often experience disease recurrence. Thus, treatment may also include immunosuppressive therapy and potentially IFN-γ supplementation.

Conclusion

Past research and case reports suggest that AIGA-associated adult-onset immunodeficiency should be considered in cases of chronic or recurrent mycobacterial infections often disseminated infections in severe cases.

病例報告 113_C128

以膽總管假性腫瘤表現的 IgG4 自體免疫胰臟炎：一例診斷困難的病例

IgG4-Related Pancreatitis Presenting as a Pseudotumor in the Common Bile Duct: A Diagnostic Challenge

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Introduction

Autoimmune pancreatitis (AIP) is a rare disease with challenging diagnosis, often mistaken as malignant pancreatic cancer. The patient presented here nearly underwent surgery, only to confirm with AIP instead after a successful treatment with corticosteroids.

Case Report

A 60-year-old man first presented with a significant weight loss (10kg) in a month, with no other remarkable clinical presentations or physical findings. Abnormal liver enzymes and elevated total bilirubin levels were observed. Abdominal ultrasound examination showed the presence of gallbladder stones and intrahepatic duct dilatation. CT scan showed diffusely enlarged pancreas with delayed enhancement, swelling of pancreatic head and dilatation of distal common bile duct (CBD) and intrahepatic ducts. In suspect of autoimmune pancreatitis, biopsies were obtained from ampulla vater during the esophagogastroduodenoscopy. Abdominal MRI revealed annular wall thickening (2.4mm) of the distal CBD with proximal dilatation of the biliary tract, raising suspicion of malignancy. An endoscopic retrograde cholangiopancreatography (ERCP) was performed for biliary tract biopsy, followed by biliary drainage and pancreatic stent placement. The biopsies were negative for malignancy. His IgG4 (411 mg/dL; normal range: 3-201) and cancer marker CA19-9 (145.25 U/mL; normal range: 0-37) were found elevated. He received a trial of steroids therapy instead of surgery after a shared decision. Follow up imaging revealed regressed tumor after a month of treatment, along with improving liver enzymes, bilirubin and IgG4 levels. He was diagnosed with definitive AIP type 1. Four months into treatment, the pancreatic and biliary stents were removed, and steroids were tapered. He remains in good health during clinic follow up.

Discussion

First proposed by Yoshida in 1995, AIP is now categorized into two types: AIP type 1 (IgG4-related disease) and AIP type 2 (pancreas-specific disease unrelated to IgG4). According to ICDC 2010, AIP is a specific form of pancreatitis presenting with obstructive jaundice with or without pancreatic masses, with lymphoplasmacytic infiltrate and fibrosis, often highly responsive to steroids therapy. Diagnosing AIP remained challenging for several reasons. AIP can closely resemble pancreatic malignancy, leading to unnecessary surgery. In patients with obstructive jaundice or pancreatic mass, having a marked elevation of IgG4 (>2 times ULN) is strongly suggestive of AIP, but false positive of IgG4 has been seen in pancreatic cancer. Conversely, a report of nearly 30% of histologically confirmed AIP patients were found with normal serum IgG4 levels. Fortunately, AIP is highly responsive to corticosteroid therapy, with rapid improvement expected after 2-4 weeks of treatment. This strategy can confirm a strong suspicion of AIP but should not substitute the need

for further investigation. Maintenance therapy may be required if complete remission is not achieved. Although patients with AIP-1 were found to have a higher relapse rate compared to AIP-2, the disease does not affect long term survival.

Conclusion

AIP should be one of the differential diagnoses considered when a pancreatic mass was identified. Some AIP patients with atypical presentations may be more challenging to diagnose despite having a set of diagnostic criteria. A rapid response to corticosteroid therapy is expected in AIP and prognosis is usually good.

病例報告 113_C129

經皮內視鏡胃造口術後意外出現門靜脈氣體：病例報告

Unexpected Portal Venous Gas Following Percutaneous Endoscopic Gastrostomy: A Case Report

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Introduction

Portal venous gas (PVG) is a rare radiological sign that can indicate serious issues, such as bowel ischemia, with mortality rates between 39% and 75%. PVG has also been observed after endoscopic procedures like percutaneous endoscopic gastrostomy (PEG) placement. Due to the infrequency of this condition, there are no established management guidelines. This case report demonstrates successful conservative management of PVG post-PEG insertion, underscoring the need for careful evaluation to rule out serious causes.

Case Report

An 87-year-old male with left upper gum cancer underwent PEG tube insertion to improve nutrition. Enteral feeding began the day after without significant discomfort. On day 3, he developed abdominal pain and lost consciousness after vomiting. Lab tests showed leukocytosis (WBC: 10790/μL), metabolic acidosis (pH: 7.26, HCO3: 18.8), and elevated C-reactive protein (35.78 mg/dL). A CT scan revealed pneumatosis gastritis and extensive PVG. With adequate bowel perfusion and no ascites or other infection sites observed on CT, the PVG was suspected to be related to PEG placement. Since the patient showed no signs of peritonitis or shock, a conservative approach was administered with fasting, proton pump inhibitors, and Piperacillin/Tazobactam. By day 8, a follow-up CT showed no further gas accumulation. PEG feeding was resumed on day 14 with good tolerance, and the patient was stable upon discharge.

Discussion

PVG is associated with several conditions, including bowel ischemia, elevated digestive tract pressure, abdominal infections, endoscopic procedures, and emphysematous gastritis. Although PVG is a rare complication of PEG tube insertion, it may result from increased abdominal pressure. Contributing factors include advanced age, excessive air during the procedure, sepsis, severe malnutrition, and rapid or high-pressure feeding, leading to gastrointestinal distension and mucosal injury.

PVG can develop at any time after PEG tube insertion, from hours to months, though it typically occurs soon after the procedure. Symptoms may include abdominal pain, nausea, vomiting, shock, or altered mental status. In some cases, patients may remain asymptomatic despite the presence of PVG, especially when there are no signs of bowel ischemia or other serious complications. Post-PEG PVG typically has a lower mortality rate compared to other causes. CT scans are essential in detecting the fatal cause of PVG and guiding the timing of surgical intervention.

Initial management for patients without peritonitis or ischemic bowel typically involves conservative measures. Surgical intervention may be needed if symptoms persist or if peritonitis

develops. If gas does not resolve within 5-7 days, a surgical consultation is advised. Surgery, including exploratory laparotomy, bowel resection, or PEG tube replacement, is crucial when bowel ischemia or significant gas threatens obstruction or perforation.

Conclusion

The management of PVG following PEG insertion depends on the patient's clinical presentation. For stable or asymptomatic patients, conservative management may be sufficient, avoiding unnecessary surgical interventions. However, careful monitoring is crucial to rule out any serious complications.

病例報告 113_C130

大腸鏡下的無症狀類腫瘤性子宮內膜異位症

An asymptomatic recto-sigmoid junction tumor-like endometriosis under colonoscopy

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Introduction

Bowel endometriosis is the most common form of severe deep endometriosis, often necessitating surgery for infertility, chronic pain, or stenosis. Here, we presented a case of incidentally found asymptomatic rectosigmoid junction tumor-like endometriosis noted under colonoscopy, sequentially laparoscopic partial colectomy was then performed.

Case Report

This 51-year-old female patient, with a history of hypertension and right ovarian endometrioid carcinoma (pT1c3N0, AJCC 8th edition), underwent debulking surgery. Followed by systemic chemotherapy with paclitaxel and carboplatin. During a routine follow-up colonoscopy for ovarian cancer, a sigmoid colon tumor was incidentally discovered. The last colonoscopy involved a polypectomy, which revealed negative findings for malignancy, but showed focal glandular hyperplasia (Fig 1) and further pathology revealed endometriosis. The patient was subsequently referred for surgical intervention. She denied symptoms such as constipation, diarrhea, tenesmus, dyschezia, and rectal bleeding.

Based on the findings of endometriosis and an adenomatous polyp in the sigmoid colon, treatment options include endoscopic submucosal dissection (ESD), and direct surgical intervention has been provided. After shared decision making (SDM), the patient preferred surgery. Laparoscopic partial colectomy was scheduled. During surgery, a firm 2.5 cm tumor was found in the sigmoid colon, along with pelvic adhesions from her previous ovarian surgery. Postoperative pathology confirmed sigmoid colon endometriosis (Fig 2), with no evidence of malignancy and clear resection margins. Additionally, four mesocolonic lymph nodes were negative for tumor involvement. Postoperatively, she has shown no signs of complications, and follow-up care includes wound management and routine monitoring with colonoscopy to ensure there is no recurrence of disease.

Discussion

Bowel endometriosis is the most prevalent form of severe deep endometriosis, affecting 3.8% to 37% of individuals with the condition. It often leads to debilitating symptoms, including dysmenorrhea, dyspareunia, dyschezia, constipation, rectorrhagia, and tenesmus, significantly impacting quality of life and potentially affecting fertility rates.

Surgery becomes a consideration in cases of chronic pain, infertility, or stenotic lesions. A thorough clinical examination and preoperative imaging are essential to accurately characterize the lesions, enabling customized surgical strategies that prioritize organ preservation. Surgeons typically choose among three surgical approaches: shaving, discoid resection, and segmental resection. While shaving is suitable for non-transfixing lesions with minimal digestive symptoms, discoid

excision can be applied to lesions up to 3 cm, and even 5 cm with robotic assistance. Segmental resection is necessary for stenotic lesions in the sigmoid and small bowel but carries a higher risk of complications with more extensive resections.

Despite advancements in imaging, there remains a lack of standardized surgical guidelines, leading to variability in treatment approaches across different practitioners and regions. Consequently, patients with similar lesions may receive either conservative or radical treatments based on their specific contexts.

In addition to surgical options, medical treatments play a crucial role. Hormonal therapies, such as combined hormonal contraceptives, progestogens, and GnRH agonists or antagonists, aim to manage symptoms by suppressing menstruation. Pain relief can be achieved with NSAIDs, complemented by a multidisciplinary approach that includes osteopathic, psychological, and nutritional support. These treatments can alleviate pain and enhance quality of life for many patients.

When medical management proves insufficient or when fertility is desired, surgical intervention can improve spontaneous and assisted pregnancy rates while also reducing pain and digestive issues.

Overall, the management of bowel endometriosis requires a tailored approach that considers individual symptoms, lesion characteristics, and patient goals to optimize outcomes and enhance quality of life.

病例報告 113_C131

接受 Atezolizumab 與 Bevacizumab 治療後出現血小板減少症的肝細胞癌病例報告

Thrombocytopenia in a Hepatocellular Carcinoma Patient Following Atezolizumab and Bevacizumab Treatment: A Case Report

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Introduction

A 54-year-old male with advanced hepatocellular carcinoma (HCC) developed thrombocytopenia following treatment with atezolizumab and bevacizumab, presenting a challenging case of potential immune-related adverse events.

Case Report

The patient, diagnosed with HCC (BCLC stage C), began atezolizumab and bevacizumab treatment on October 9, 2023, and received his 12th cycle on June 13, 2024. His medical history included Klebsiella pneumoniae bacteremia, for which he was hospitalized from July 12 to July 18, 2024. Thrombocytopenia was noted during treatment, with platelet count declining, necessitating daily platelet transfusions from July 3 to July 9, 2024. Concurrently, the patient developed pulmonary embolism and was started on enoxaparin, later transitioned to edoxaban, which was discontinued due to epistaxis. On July 16, 2024, the patient began eltrombopag, leading to improved thrombocytopenia. Currently, the patient is under Lenvatinib treatment following the 12th cycle of immunotherapy.

Discussion

The thrombocytopenia could be attributed to immune-related thrombocytopenia (ir-TCP) from checkpoint inhibitor therapy, bevacizumab-induced thrombocytopenia, or other factors such as cirrhosis and infection. Immune-related thrombocytopenia typically develops within the first 12 weeks of therapy, with a median onset of 40-44 days, potentially involving autoimmune destruction of hematopoietic stem cells. Bevacizumab-induced thrombocytopenia, though rare, may cause platelet dysfunction and consumption. Standard ir-TCP management involves corticosteroids, but in this case, eltrombopag was recommended due to the patient's complex condition.

Conclusion

This case illustrates the complexities of managing thrombocytopenia in advanced HCC patients receiving immunotherapy. Multiple potential etiologies, including immune-related events, drug-induced effects, and underlying conditions, must be considered, with close monitoring and tailored management strategies being critical.

病例報告 113_C132

一位以胃食道逆流為初始表現的 HSV 食道炎患者

HSV esophagitis in an immunocompetent host with initial presentation of gastroesophageal reflux

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Introduction

Esophagitis is most often caused by noninfectious conditions, whereas esophageal infections mainly occur in individuals with compromised immune systems from chemotherapy, transplantation, or HIV infection. The most common causes of infectious esophagitis include Candida, cytomegalovirus (CMV), and herpes simplex virus (HSV).

Case Report

A 43-year-old male without any systemic underlying diseases. He is an active alcoholic and smoker, and has recently quit chewing betel nuts. He visited the gastroenterology outpatient department primarily due to marked acid regurgitation after meals for several days. He was found to have a temperature of 38.4°C during the outpatient department evaluation. He reported general malaise but denied having symptoms such as chills, cough, dysuria, or epigastric pain. Esophagogastroduodenoscopy (EGD)suggests that the patient may have a reactivation of a latent HSV infection. The patient was treated with Lansoprazole and sucralfate for two weeks and received a 5-day course of Acyclovir.

Discussion & Conclusion

The average age of incidence for HSV esophagitis is 35 years, with a predominance in males. While most cases are caused by HSV-1, there have been a few reported cases of HSV-2 esophagitis. Patients typically have a prodromal systemic manifestations of fever, nausea, or vomiting before the onset of esophageal symptoms.

病例報告 113_C133

以 Infliximab 治療貝塞特氏病後發生線性 IgA 皮膚病的罕見案例

A rare case of linear IgA dermatosis after Infliximab as treatment for Behcet disease

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Introduction

Linear IgA dermatosis (LABD) is a rare subepidermal vesiculobullous disease that affects both adults and children. In children, the prognosis is often favorable, but managing the condition in adults can be more complex. The causes and mechanisms behind LABD remain unclear, though it has been associated with malignancies, inflammatory diseases, and certain medications. In adults, drug-induced LABD must be carefully considered, especially since there are few reports linking the condition to infliximab. Here, we discuss a rare case of LABD triggered by infliximab, administered to a patient with Behçet's disease, with a notable response to dapsone treatment.

Case Report

The patient, a 48-year-old woman, had a history of Behçet's disease, characterized by recurrent painful oral ulcers, colonic ulcers, and a positive pathergy test since 2019. Her condition worsened in January 2023, with increased oral ulcers, fatigue, and severe anemia. Despite evaluations, no clear cause for her anemia was found. In August 2023, a second colonoscopy revealed colitis, and she was hospitalized in September for further treatment. Her management included hydroxychloroquine, colchicine, levamisole, and increased prednisolone. Infliximab was added to address her persistent oral ulcers, which gradually improved.

However, one week after starting infliximab, the patient developed painful, itchy erythematous plaques on her palms and soles. A skin biopsy revealed linear IgA immunoreactivity, confirming LABD. The lesions responded only partially to steroids and recurred with dosage reductions. Dapsone was introduced, and after increasing the dose, her condition improved, with no new lesions appearing. Steroids were gradually tapered, and her improvement continued at follow-up.

Conclusion

LABD is a rare autoimmune disease, with reported incidences ranging from less than 0.5 to 2.3 cases per million annually. It is characterized by the linear deposition of IgA at the basement membrane zone. Although the disease is understood as autoimmune, the specific mechanisms behind it are unclear. It has been linked to infections, malignancies, inflammatory diseases, and medications, especially vancomycin. In this case, infliximab triggered the condition, though reports of this association are scarce.

LABD can present with various skin manifestations, including erythematous papules, plaques, and vesiculobullous lesions, often accompanied by pruritus. Diagnosis relies on pathology, and the primary treatment is dapsone, with notable improvements typically seen within days. Other treatments, including steroids and antibiotics, have been used, but relapse is common with medication tapering. In this patient, infliximab triggered the development of LABD, which responded poorly to steroids but showed a dramatic improvement with dapsone. Close

monitoring for side effects, particularly on liver function and blood counts, is essential during dapsone treatment. This case underscores the importance of considering drug-induced LABD in adults and the effectiveness of dapsone in managing this rare condition.

病例報告 113_C134

小腸基質瘤造成的消化道出血病例報告

A case report of obscure gastrointestinal bleeding caused by jejunal gastrointestinal stromal tumor

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Introduction

Bleeding from the small bowel is uncommon, but it is responsible for the majority of patients with gastrointestinal bleeding that recurs without an obvious etiology after upper endoscopy, colonoscopy, and, possibly, radiologic evaluation of the small bowel. Gastrointestinal stromal tumors (GISTs) are rare tumors of the gastrointestinal (GI) tract, which can occur in majorly stomach, and rarely in the small intestine, rectum, and large intestine. We report a jejunal GIST presented with melena.

Case Report

A 66-year-old male patient with a past history of hypertension was admitted via the emergency department due to tarry stool lasting for 5 days.

The patient reported experiencing tarry stool for 5 days. He had previously sought help from a local medical doctor (LMD) without improvement. He also noted general malaise, dyspnea on exertion, and dizziness while working over the past 5 days. He denied nausea, vomiting, chest pain, abdominal pain, dysuria, and watery diarrhea. He had been hospitalized for a similar episode last year and reported intermittent tarry stools during the year, especially after consuming alcohol. At the emergency department, laboratory data revealed no leukocytosis, normocytic anemia (Hb: 5.2 g/dL), no thrombocytopenia, and normal coagulation function. Blood chemistry showed mild hypokalemia (K: 3.3 mmol/L), normal liver enzyme levels, and an elevated creatinine level (1.43 mg/dL). With a presumptive diagnosis of gastrointestinal bleeding, the patient was admitted for further evaluation and management.

Following admission, upper endoscopy revealed erosive esophagitis, LA Grade A, gastric ulcers (status post biopsy), and a gastric polyp. Colonoscopy revealed colonic and rectal polyps (status post removal). Abdominal CT indicated a lobulated neoplasm in the proximal jejunum. A general surgeon was consulted, and the patient underwent laparoscopic surgery with proximal jejunal segmental resection and anastomosis on 2024/05/22. The final pathology report confirmed a gastrointestinal stromal tumor of the gastric body, pT2N0M0, stage I.

Discussion

Jejunal GIST usually manifests as asymptomatic subepithelial mass and is associated with abdominal discomfort or GI bleeding. Sudden unset bleeding is a rare manifestation of jejunal GIST.

Conclusion

Recurrent GIB and unusual imaging findings should raise clinical suspicion for alternative causes for GIB, including tumors such as GIST.

病例報告 113_C135

新型隱球菌引起之軟組織感染: 一例台灣罕見的病例報告

A case report of soft tissue infection caused by Cryptococcus neoformans: an unusual pathogen in Taiwan

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Introduction

Cryptococcus is a yeast found in the environment, transmitted to humans via inhalation. Immunocompromised individuals are at risk for severe infections like meningitis. Skin infections are rare and usually occur through direct inoculation in both healthy and immunocompromised people.

Case Report

This case involves a 62-year-old male with a medical history of alcoholic hepatitis, hypertension, chronic kidney disease, and benign prostatic hyperplasia, who presented with left lower limb erythema and swelling for one month. He had previously been treated for left leg cellulitis with Piperacillin + Tazobactam, which initially improved his condition. However, after discharge, his symptoms recurred, including left leg swelling and erythema, prompting further evaluation.

Physical examination revealed redness, induration, heat, and swelling extending from the dorsum of the foot to the medial leg. Despite no fever or systemic signs of infection, empirical antibiotics were administered again for presumed cellulitis. The patient's symptoms persisted, particularly pain and tenderness in the calf, leading to a consultation with an infectious disease specialist. A skin biopsy and blood tests revealed cryptococcal infection, confirmed by the presence of Cryptococcus neoformans in both cerebrospinal fluid and skin samples. The patient was diagnosed with cryptococcal meningoencephalitis and cellulitis.

Treatment initially involved Flucytosine + Amphotericin B Liposome, but was later switched to Flucytosine + Fluconazole due to worsening renal function. An MRI revealed multiple brain lesions consistent with cryptococcomas. Although his cellulitis improved, he experienced severe side effects, including hepatic encephalopathy, necessitating a switch to oral fluconazole. He was discharged in stable condition on August 6 and continues with outpatient follow-up.

Discussion

The skin is the third most common site of cryptococcal infection, typically associated with disseminated disease in immunocompromised patients. It can mimic bacterial infections like cellulitis or abscesses and present as papules, maculopapules, or draining sinuses. A full workup is needed to assess for systemic disease.

Conclusion

Cryptococcal soft tissue infections are rare in immunocompetent or mildly immunosuppressed patients and may be overlooked without suspicion. This report recommends early tissue sampling, special staining, and cultures for atypical skin lesions unresponsive to antibiotics, regardless of immune status.

病例報告 113_C136

一位患有嗜酸性食道炎的病人：個案報告

Eosinophilic esophagitis in a young male patient: A Case Report

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Introduction

Eosinophilic esophagitis (EoE) is a chronic, immune-mediated esophageal disorder characterized by eosinophilic infiltration of the esophageal mucosa. It is increasingly recognized as a significant cause of upper gastrointestinal symptoms in both adults and children, but remains a relatively rare disease in Taiwan. This case report presents a 31-year-old male with eosinophilic esophagitis, highlighting the clinical presentation, diagnostic workup, and management approach.

Case Report

A 31-year-old male with no known underlying systemic diseases and a history of heavy smoking (3 packs per day) presented to our hospital with progressive dysphagia. He did not have any history of asthma or allergic disease.

The patient initially experienced a sore throat and a sensation of a foreign body in his throat 5 days prior to admission. These symptoms were accompanied by a poor appetite and a rash that appeared over his trunk and bilateral thighs. Over the following days, his symptoms worsened, developing into dysphagia and chest tightness. He sought treatment from a local medical doctor but did not experience any relief. One day before admission, the patient developed a fever, prompting his referral to our hospital for further evaluation and management.

On admission, laboratory studies revealed significant peripheral eosinophilia with an absolute eosinophil count of 3016/μL, and a total IgE level of 76200 IU/L. An esophagogastroduodenoscopy showed a small mucosal break (< 5 mm in length) and circumferential fold thickening with a slightly whitish appearance. A biopsy was taken. Oral lansoprazole and anti-histamine, levocetirizine and glycyrrhizinate, were prescribed for esophagitis. Later, histopathological examination of the biopsy revealed parakeratosis, squamous hyperplasia, chronic inflammation, and a marked increase in eosinophils (25/HPF), findings consistent with eosinophilic esophagitis. Consequently, the patient was diagnosed with eosinophilic esophagitis. His symptoms were gradually relieved after treatments.

Discussion

Eosinophilic esophagitis (EoE) is a chronic inflammatory condition of the esophagus characterized by symptoms of esophageal dysfunction and an esophageal epithelial eosinophilic infiltrate. The incidence of EoE varies significantly by region, with estimates ranging from 1.30 to 12.8 cases per 100,000 individuals. Higher prevalence was observed in males, Whites, and with certain environmental factors, including antibiotic use in infancy, caesarean delivery, preterm birth, and low birth weight.

EoE presents with a range of symptoms primarily related to esophageal dysfunction. Dysphagia (difficulty swallowing) is the most common symptom. Other manifestations include central chest

pain, gastroesophageal reflux disease (GERD)-like symptoms or refractory heartburn, and upper abdominal pain. These symptoms are often chronic and may be mistaken for other esophageal conditions, such as GERD, leading to delays in diagnosis.

The diagnosis of EoE is based on clinical, endoscopic, and histological findings. About 50-60% of patients exhibit elevated serum IgE levels, often above 114,000 units/L. Peripheral eosinophilia is also common, seen in 40-50% of patients. Endoscopy frequently reveals characteristic features such as esophageal edema, stacked circular rings (also known as “trachealization”), strictures, whitish specks indicative of eosinophilic abscesses, linear furrows, and a narrow-caliber esophagus. Histologically, it is characterized by eosinophil-predominant inflammation in the esophageal mucosa, predominantly mediated by CD4+ T helper type 2 (Th2) cells.

The treatment of EoE focuses on reducing inflammation and preventing complications such as esophageal stricture formation. Several therapeutic options included proton pump inhibitors (PPIs), elimination diets, swallowed corticosteroids, esophageal dilation and biologics. Dupilumab, a monoclonal antibody targeting the IL-4 receptor alpha subunit, has emerged as a promising biologic therapy for EoE. It works by inhibiting key inflammatory pathways involved in the Th2 response, thus reducing eosinophilic inflammation.

Conclusion

Eosinophilic esophagitis is a chronic, immune-mediated disease that significantly impacts patients' quality of life due to its debilitating symptoms and long-term complications. Early diagnosis and appropriate management are essential to prevent esophageal damage and improve outcomes. Advances in biologic therapies, combined with established treatments like dietary modification and corticosteroids, offer hope for better control of the disease in affected individuals.

病例報告 113_C137

原發性胃淋巴癌:內視胃鏡下的模仿者

Primary gastric lymphoma: a mimicker in upper gastrointestinal endoscopy

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Introduction

Gastric neoplasms frequently exhibit overlapping endoscopic characteristics, including similarities in shape, border definition, location, central features, and overall morphology. We present a primary gastric lymphoma that mimicked both gastric adenocarcinoma and gastrointestinal stromal tumor (GIST) on endoscopy, highlighting the diagnostic challenge posed by these shared features.

Case Report

An 80-year-old man with hypertension was admitted after an episode of massive hematemesis. On arrival, he presented with systolic blood pressure of 116 mmHg and heart rate of 77 bpm. Nasogastric tube decompression yielded over 500 mL dark, bloody content. Blood test showed anemia (Hb 11.8g/dL). Upper gastrointestinal endoscopy visualized two 2.0cm round ulcers at greater curvature of the upper body of the stomach. It presented with central indentation, elevated borders with sharp angles transitioning to the adjacent mucosa. Upon applying pressure with forceps, the lesions were immobilewithout pillow sign.

(Figure 1) Biopsy was obtained. Microscopically, there was diffuse infiltration of atypical large lymphoid cells within the gastric mucosa, without evidence of Helicobacter pylori infection. The pathology diagnosis is diffuse large B cell lymphoma(DLBCL), germinal center type. Abdominal CT showed no evident lesion in stomach. Whole body positron emission tomography (PET) confirmed the diagnosis as Ann-Arbor stage II, involving the cardia, fundus, and body of the stomach, along with lymph nodes surrounding the celiac trunk.

Discussion

Gastric neoplasms often exhibit similar characteristics during endoscopic examination. Gastric lymphoma represents 1–5% of malignant tumours of the stomach. Primary gastric lymphomas are confined to the stomach and regional lymph nodes. They are predominantly non-Hodgkin lymphomas of B-cell origin.[1] Most subtypes share same endoscopic feature. There is no universal classification criteria at the macroscopic level currently.[2]

A systemic review analyzed 2000 cases of primary gastric lymphoma, mostly MALT lymphoma and DLBCL, according to their initial clinical and endoscopic presentations. They classified the endoscopic findings into 3 major types: exophytic type; ulcerative type; hypertrophic type, or mixed type.[3]

Gastric adenocarcinoma represents over 95% of malignant gastric tumours.[4] A study was conducted to evaluate the challenges of discriminating advanced gastric cancer from gastric lymphoma during endoscopic examination. The overall diagnostic accuracy by experts was only

60.4%.[5]

Primary GISTs typically presents as large masses with irregular lobulated margins, mucosal ulceration, central necrosis, hemorrhage, and heterogeneous enhancement. [6] Mucosal ulceration may occur in 15–50% cases.

Reviewing our case and its endoscopic findings, these ulcers had a central indentation, elevated borders with sharp angles transitioning to the adjacent mucosa. Upon applying pressure with forceps, the lesions were immobile and lacked the pillow sign. The differential diagnosis included benign gastric ulcer, gastric cancer, GIST, or lymphoma. Benign gastric ulcer was excluded due to elevated borders with central depression. Gastric cancer was also excluded, as it is rare to present with two distinct lesions. GIST was suspected, but reconsidered owing to its rare presentation with two simultaneous lesions. Besides, the lesion's immobility without blunt interface excluded possible submucosal tumor. These characteristics led us to conclude it as an ulcerative type of gastric lymphoma.

病例報告 113_C138

PTEN 基因突變相關之縱膈腔動靜脈畸形併發嚴重肺高壓

A case report of PTEN gene mutation with mediastinal arteriovenous malformation causing left to right shunting and severe pulmonary hypertension

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Case Report

A 39-year-old man with history of T2 spinal dural arteriovenous fistula status post transarterial embolization and complicated with vertebral fracture causing paraplegia, and chronic respiratory failure under tracheostomy with intermittent mechanical ventilation support, was referred to our hospital due to progressive abdominal distension and shortness of breath for several weeks. Physical examination revealed increased abdominal girth with shifting dullness, and limbs pitting edema. Ascites and hepatic congestion was confirmed through abdominal computed tomography, and peritoneocentesis was done. After analyzing ascites data, cardiogenic ascites was favored. His chest plain film illustrated widened upper mediastinum and pulmonary congestion. A dilated right atrium (RA) and D-shaped left ventricle were identified through echocardiography. Cardiac catheterization was performed under the suspicion of severe pulmonary hypertension, which found elevated pressure of RA (33/33 mmHg), right ventricle (RV) (70/20 mmHg) and pulmonary artery (PA) (68/44 mmHg). Oxygenated superior vena cava (SVC) (97%), RA (94%), RV (96.8%), and PA (95.6%). Magnetic resonance angiography of chest was ordered for shunting identification, which pictured multiple engorged vessels in right posterior mediastinum and paravertebral region, with drainage vein of dilated azygos arch. Intracranial arteriovenous malformation (AVM) was also discovered. Genetic testing of serum with next generation sequencing (InheriNext) was done and revealed phosphatase and tensin homolog (PTEN) gene mutation. A diagnosis of PTEN gene mutation with mediastinal arteriovenous malformation causing left to right shunting and severe pulmonary hypertension was made. The patient was admitted to our intensive care unit, and after enhancing fluid balancing and diuretics adjustment, the patient was discharged on day 10 of admission uneventfully.

Discussion

Germline pathogenic PTEN gene variant is responsible for a variety of syndromes with different clinical presentations and malignant potential, collectively called PTEN hamartoma tumor syndromes (PHTS), and is inherited in an autosomal dominant fashion. Cowden syndrome (CS) and Bannayan-Riley-Ruvalcaba syndrome (BRRS) were the most recognized phenotypes. CS manifests multiple hamartomas involving mucocutaneous, breast, thyroid, genitourinary, gastrointestinal, and neurologic systems. On the other hand, BRRS embrace characteristics such as macrocephaly, penile lentigines, proximal muscle myopathy, joint hypermobility, lipomatosis, intestinal hamartomatous polyposis, developmental delay, and vascular anomalies such as arteriovenous malformation. In contrast to CS, BRRS tends to be diagnosed early in life. Other than hamartomas, AVM is also one of the distinct manifestation of PHTS. In one retrospective study, vascular abnormality was presented in 54% of patients with PTEN mutation.

We presented a case of multiple AVM involving intracranial, spinal dura, and mediastinum, causing extracardiac left to right shunting, resulting in severe pulmonary hypertension. Differential diagnosis such as hereditary hemorrhagic telangiectasia (HHT) was also considered, thus genetics testing was ordered. AVMs seemed to be the major clinical manifestation of PTEN mutation in this patient.

Conclusion

PTEN gene mutation caused a variety of syndromes, characterizing hamartomas involving multiple organs with malignant potential, and also arteriovenous malformations. Active surveillance of cancer in these patients is warranted, and genetic counseling of family members should be considered.

病例報告 113_C139

免疫檢查點抑制劑對原發不明癌症之療效

Efficacy of immune checkpoint inhibitor for cancer of unknown primary: a case report

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Introduction

The incidence of carcinoma of unknown primary (CUP), with poor prognosis, is around 5-10% among all malignancies. The one-year survival rate is 20%, and the median survival is only 3-6 months. It is a critical disease in clinical practice.

Case Report

This 60-year-old man had suffered from a progressively enlarging, firm, and tender mass over the left axillary region for two years. The excisional biopsy of the mass reported metastatic adenocarcinoma, with the immunohistochemistry (IHC) stains revealing CK7(+), p40(-), TTF-1(-), and PSA (-). Upper and lower gastrointestinal scopes, cystoscope, chest and abdominal computed tomography (CT), and whole-body Positron Emission Tomography-computed tomography (PET-CT) scan were used for primary site survey. Repeated sonography-guided needle biopsy of the left thigh mass also showed metastatic adenocarcinoma with CK7(+) and CK20(-). Other IHC stains presenting GATA3(+), CDX-2(-), TTF-1(-), and PAX-8(-), indicated that genitourinary tract origin was the most likely origin. Programmed death ligand 1 expression was reported in 60% of tumor cells. Because no significant primary origin was detected, CUP was diagnosed and favored urothelial cell carcinoma (UCC).

Palliative chemotherapy with cisplatin plus gemcitabine was administered initially. Unfortunately, his disease progressed after 2.5 months of treatment. The subsequent treatment with one course of CMV (cisplatin, methotrexate, and vinblastine) still failed. Immune checkpoint inhibitors (ICIs)-based therapy was prescribed for him with nivolumab alone in 200mg fixed dose every two weeks for 4.2 months with complete remission. After progression, combination therapy with nivolumab, plus lenvatinib and paclitaxel, for 17.2 and 2.0 months, respectively. The follow-up PET-CT scans remained in complete remission. Until now, the patient is still alive, with progression-free survival of 49.9 months calculated from the date of initial ICI up to the time of writing, and ongoing.

Discussion

ICIs have widely been used in metastatic UCC. IMvigor130 and Keynote-361 trials reported benefits of survival and response in the first-line setting. The US Food and Drug Administration has approved atezolizumab and pembrolizumab for platinum-ineligible patients with metastatic UCC. ICI monotherapy also showed significant survival benefits in the second-line setting. Although the patient was diagnosed with CUP, ICI-based therapy was still an ideal option because the IHC stain showed a metastatic UCC picture.

Conclusion

Cooperating with radiologists and pathologists is crucial in identifying the primary site of CUP. In our patients, an adequate discussion between the medical oncologist, radiologist, and pathologist supported the patient's diagnosis of CUP, suspected metastatic UCC. ICIs are still an effective option for this CUP patient.

病例報告 113_C140

胰管內視鏡介入治療後的罕見膽管炎原因-早期胰臟癌診斷的個案報告

A Rare Cause of Cholangitis After Pancreatic Duct Disruption Management

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Introduction

Pancreatobiliary cancer can also be the etiology for 5% of patients with unexplained acute pancreatitis and for up to 12% of those with recurrent pancreatitis. This case demonstrated a rare cholangitis after pancreatic duct disruption management.

Case Report

This 44-year-old man had history of repeated acute pancreatitis, alcoholism, dyslipidemia and hypertension. He was admitted for epigastric pain after abstaining from alcohol for 1 month. Physical examination revealed epigastric tenderness with rebound pain and muscle guarding. Abdominal computed tomography (CT) showed swelling of pancreas parenchyma, complicated with extensive peripancreatic fat stranding, multiple large cystic lesions at pancreatic tail (largest 11 cm), and some abscess accumulation at left subphrenic area. Pancreatic duct dilatation was noticed at pancreatic head without visible obstructive lesions on CT. Endoscopic ultrasonography (EUS) was arranged which showed no obvious obstructive lesions at pancreatic head. For acute pancreatitis with P-duct disruption and pancreatic tail pseudocyst formation, endoscopic retrograde cholangiopancreatography (ERCP) for P-duct stenting succeed. Percutaneous drainage of the largest pseudocyst was conducted. Repeated abdominal CT showed improvement of peri-pancreatic abscess and the diminishment of pseudocyst. After the removal of external drainage tube, the patient was discharged. However, epigastric pain with chillness, icteric sclera, yellowish discoloration of skin, and tea-colored urine were noticed 14 days later. Obvious elevation of biliary enzymes was noted. Abdominal CT demonstrated common bile duct (CBD) and intrahepatic duct (IHD) more dilatation than previous CT done 20 days before. During the second hospitalization for acute cholangitis, ERCP revealed a distal CBD stricture, which was not obvious in previous EUS or ERCP. Endoscopic trans-papillary under flow bile duct biopsies reported high grade dysplasia. The abdominal magnetic resonance imaging found a 2.2-centimeters tumor with restricted water diffusion at pancreatic head. After the Whipple operation, the histology revealed a pancreatic ductal adenocarcinoma, pT2N0M0 stage IB with CBD involvement.

Discussion

Acute pancreatitis with distal common bile duct stricture should be considered pancreatic head mass. Moreover, pancreatitis is also a risk factor of pancreatic cancer. A pooled analysis revealed recent pancreatitis was associated with an odds ratio of 21.35 (95% CI 12.03–37.86) for pancreatic cancer, while diagnosis of pancreatitis more than 2 years previously had a ratio for pancreatic cancer of 2.71 (95% CI 1.96–3.74) [1, 2]. In diagnostic accuracy of pancreatic or biliary cancer, contrast-enhanced harmonic EUS with Sonazoid showed significantly better than conventional EUS [3].

Conclusion

The patient had minute pancreatic head cancer related repeated pancreatitis episodes initially, even after conventional EUS. We believe that routine Sonazoid-enhanced EUS should be performed in this situation.

病例報告 113_C141

壞死桿菌引起的肝膿瘍

A rare case of liver abscesses caused by *Fusobacterium necrophorum*

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Introduction

Fusobacterium necrophorum is a gram-negative anaerobic pathogen of the normal oral flora. Liver abscesses caused by *F. necrophorum* in clinical settings are seldom reported, especially in individuals with normal immune function.

Case Report

A 23-year-old man with no underlying health conditions was admitted to our outpatient department due to intermittent high fevers accompanied by chills, fatigue, and severe throat pain for the past two weeks. Four weeks prior to this admission, he had unprotected sex with multiple partners. Aside from this sexual activity, he reported no other significant travel, occupational, or contact history. In the outpatient department, he exhibited a fever of up to 39°C and tachycardia. Physical examination showed bilaterally enlarged tonsils without pus discharge. Laboratory results indicated leukocytosis with a predominance of neutrophils, elevated high-sensitivity C-reactive protein (hsCRP), and mild hyperbilirubinemia. Upon admission, he was started on a treatment regimen of ceftriaxone and doxycycline due to fever and impaired liver function. Subsequent abdominal ultrasound revealed a 3.8-centimeter anechoic area with a hypoechoic irregular ring in segment 6 of the liver, and liver abscess was confirmed via triple-phase abdominal CT scan. On the third day of admission, blood cultures showed growth in the anaerobic bottle, leading to the addition of metronidazole to his treatment regimen alongside ceftriaxone. The pathogen causing the bacteremia was later identified as *F. necrophorum* on the sixth day. The patient also had ultrasound-guided fine needle aspiration to determine the cause of the liver abscess, and cultures from the aspirated fluid confirmed the presence of the sole pathogen, *F. necrophorum*. Consequently, ceftriaxone was discontinued, and he continued with metronidazole alone. During his hospitalization, there were no neck symptoms indicative of jugular vein thrombosis. HIV status and blood glucose levels were also assessed, showing no signs of diseases associated with immunocompromise. After 14 days of metronidazole treatment, all symptoms resolved, and he was discharged. He received an additional two weeks of oral metronidazole in the outpatient setting. Follow-up ultrasound showed a reduction in the size of the liver abscess, and he recovered well with no lasting sequelae.

Discussion

In Taiwan, *Klebsiella pneumoniae* is the most common pathogen responsible for liver abscesses, followed by other Enterobacterales. In young men with unprotected sexual exposure, amoebic liver abscess should also be considered. It is uncommon for liver abscesses to be caused by *F. necrophorum* in clinical settings, particularly in individuals with normal immune function. *F. necrophorum* is an obligate anaerobic, gram-negative bacillus that is part of the normal flora in

the oral cavity, gastrointestinal tract, and female genital tract. It can potentially cause opportunistic infections via the oropharynx, leading to conditions such as internal jugular vein thrombophlebitis, Lemierre's syndrome, or migratory infections, including infective endocarditis, brain abscesses, pleurisy, and liver abscesses.

Conclusion

F. necrophorum liver abscesses are uncommon because of the rarity of this bacterium and the difficulties in culture, which can result in misdiagnosis. In cases of liver abscesses with unknown causes, particularly when accompanied by oropharyngeal symptoms, clinicians should consider the potential for *F. necrophorum* infection.

病例報告 113_C142

合併上腸繫膜動脈剝離與中弓韌帶綜合症之罕見病例報告

Case Report of Concurrent Superior Mesenteric Artery Dissection and Median Arcuate Ligament Syndrome

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Case Report

This 49-year-old male patient with a history of smoking (half a pack per day) and no known history of hypertension, diabetes, or cardiovascular disease presented with recurrent abdominal pain. Approximately 10 months prior, he experienced sudden umbilical pain after eating, which resolved after medical treatment for suspected obstruction. One week before his current admission, he developed similar postprandial pain, which persisted despite treatment at a nearby hospital. He also reported mild diarrhea without fever, blood in stools, nausea, or vomiting. A CT scan revealed focal stenosis of the proximal celiac trunk with post-stenotic dilatation, suggesting Median Arcuate Ligament Syndrome (MALS), and the superior mesenteric artery dissection. Upon admission, the patient was treated with intravenous fluids and antibiotics, and his blood pressure was managed with Nicardipine and Norvasc. His condition stabilized, and he was discharged with plans for outpatient follow-up.

Discussion

This case presents a male patient with recurrent postprandial abdominal pain, who was diagnosed with both superior mesenteric artery (SMA) dissection and Median Arcuate Ligament Syndrome (MALS). These two conditions are rare vascular abnormalities that can lead to ischemic symptoms in the gastrointestinal tract. The patient's history of episodic abdominal pain, initially managed as an intestinal obstruction, and subsequent CT findings point to the complexity of these overlapping vascular syndromes.

The CT imaging in this case revealed focal stenosis of the celiac trunk, with post-stenotic dilatation consistent with MALS, and thickening of the SMA walls, suggestive of arterial dissection. MALS is characterized by the compression of the celiac artery by the median arcuate ligament, leading to postprandial pain due to compromised blood flow. SMA dissection, though less common, can occur due to various factors, including hypertension and atherosclerosis, as seen in this patient.

The decision to manage this patient conservatively aligns with the treatment strategy often recommended for stable cases of SMA dissection and MALS without signs of bowel ischemia or aneurysm rupture. In this case, controlling the patient's blood pressure using Nicardipine and transitioning to oral antihypertensives like Norvasc successfully stabilized the patient. This conservative approach, supported by intravenous fluids and antibiotics, allowed for symptom relief without the need for surgical intervention.

The co-occurrence of SMA dissection and MALS presents a unique clinical challenge. The stenosis of the celiac artery in MALS can increase blood flow through collateral pathways, potentially contributing to the development of SMA dissection. This case highlights the importance of recognizing these vascular conditions early, especially in patients presenting with recurrent,

unexplained postprandial pain. Early diagnosis using CT angiography is crucial to guide appropriate management and prevent complications such as bowel ischemia.

Conclusion

In conclusion, this case demonstrates the successful conservative management of a patient with concurrent SMA dissection and MALS. Careful monitoring and management of blood pressure, along with supportive care, resulted in a favorable outcome. Further research is needed to explore the long-term outcomes of patients with these coexisting conditions.

病例報告 113_C143

金黃色葡萄球菌毒素休克症候群合併敗血性肺栓塞 – 個案報告

Staphylococcal toxic shock syndrome (STSS) complicated with septic pulmonary emboli – Case Report

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Introduction

Staphylococcal toxic shock syndrome (STSS) was firstly reports in 1980s among young female cases with menstruation causes. Clinical manifestations include high fever, hypotension, diffuse erythematous rash, and multiple organ dysfunction, which may rapidly progress to severe and intractable shock. Diagnosis is made clinically and by isolating the organism.

Case Report

A 78-year-old female was transferred from one community hospital to the medical center on Sep 7, 2024 because of persistent shock and metabolic acidosis. The medical history included hypothyroidism, heart failure, and chronic kidney disease. She got intramuscular injection over left gluteal major one week before, and progressive left hip pain, fever, and confusion occurred later. The CT scan without contrast because of impaired renal function showed increased heterogenous density over left gluteal major and the nearby soft tissue. At ED, hypotension (91/57 mmHg), respiratory distress (23 breaths/min), diffuse erytheamtous macules over waist and left hip, and azotemia (BUN/Cr: 80/2.87 mg/dl) were noted. Toxic shock syndrome was impressed initially and ressuscitation with vigorous fluid infusion, empiric antibiotic therapy with oxacillin, and oxygen supplement were instituted immediatly. The renal fuction gradually recovered, and the CT scan with contrast demonstrated a long rhombic abscess in left gluteal major and wedge shaped nodules adhered to pleura in both lungs. Two sets of blood cultures yielded methicillin-susceptible *Staphylcococcus aureus* (MSSA). Staphylococcal toxic shock syndrome complicated with septic pulmonary emboli CT-guided drainage was done with much brownish pus drained. The symptoms of respiratory distress and confusion resolved remarkablely two days after admission. The followed CXR showed that infiltrations in both lungs resolved. At present time, she still received appropriate antibiotic therapy and drainage.

Discussion

Toxic shock syndrome often exhibits with nonspecific manifestations which may result in diagnosis delay and considerable mortality. Early diagnosis, aggressive ressuscitation, and appropriate antibiotics are essentials for lives rescusing of STSS patients.

病例報告 113_C144

化膿性子宮肌瘤合併大腸桿菌菌血症：以持續半年的多次敗血症表現

A rare case of pyomyoma with *Escherichia coli* bacteremia manifested as recurrent episodes of sepsis over a six-month period

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Case Report

A 41-year-old female presented to outpatient department of infectious disease with intermittent fever up to 39.5°C with chills for one week. Initially diagnosed with a partially treated urinary tract infection, her fever did not respond to oral antibiotic treatment. She was subsequently admitted for a comprehensive evaluation. She reported occasional lower abdominal pain, but no palpable mass or peritoneal signs were observed. Otherwise, the rest of the physical examination was normal. Blood test revealed leukocytosis, elevated hsCRP, and mild elevation of liver and biliary enzyme. Hematuria and bacteriuria were found on urinalysis. Because of fever with unknown origin (FUO), an abdominal CT was performed, showing unremarkable results apart from a suspicious uterine myoma measuring 6 cm x 5 cm, characterized by calcification and poor enhancement. The patient indicated that she had known about the uterine myoma for at least 15 years and had been followed up in the GYN outpatient department, but no additional intervention had been suggested. Her fever gradually subsided under ceftriaxone treatment. Subsequently, blood cultures revealed the presence of *Escherichia coli*. To identify the source of the *E. coli* bacteremia, an inflammation scan was performed, which revealed focal uptake in the leiomyoma, raising concerns about the potential for malignancy or a concurrent infection. A gynecologist was consulted; however, since all tumor markers were normal and her fever had improved, surgery was deferred, and follow-up in the GYN outpatient department was recommended.

After the episode, she experienced recurrent episodes of fever and low back pain over the next six months. Follow-up abdominal CT scans showed progression of the uterine myomas. After discussions with her gynecologist, surgical intervention was strongly recommended. Ultimately, the patient underwent a total hysterectomy with bilateral salpingectomy. During the procedure, pus was observed within the uterine myoma. The final pathology report indicated benign leiomyoma with abscess formation. Pus cultures later identified *E. coli* with the same resistance profiles as the previous bacteremia. Following the surgery, the patient was discharged without complications and remained free of fever or other signs of infection during her follow-up at the clinic for over eight months.

Discussion

We present a rare case of pyomyoma with *E. coli* bacteremia in a patient who experienced intermittent fever for more than six months. Uterine leiomyomas are the most common neoplasms of the uterus in women, but pyomyoma—a suppurative form of leiomyoma—is a rare condition caused by infarction and infection of the leiomyoma.

Pyomyoma is associated with a high fatality rate of about 21%. The condition may be difficult to diagnose especially in those with a nonspecific clinical presentation. Reported treatments of

pyomyoma included parenteral antibiotics, hysterectomy, myomectomy, and CT-guided drainage. Delayed diagnosis may result in serious complications, whereas adequate surgery and broad-spectrum antibiotics may decrease serious morbidity and mortality.

Conclusion

This case highlights the rarity of pyomyoma in a patient presenting with FUO. While uterine leiomyomas are common, pyomyoma is a rare and potentially life-threatening condition. Prompt recognition is important to avoid possible misdiagnosis of severe infection or malignancy.

病例報告 113_C145

Quetiapine 造成的抗精神病藥物惡性症候群及橫紋肌溶解症

Quetiapine-Induced Fatal Neuroleptic Malignant Syndrome

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Introduction

Neuroleptic Malignant Syndrome (NMS) is a severe reaction to dopamine-blocking drugs or sudden withdrawal of dopamine agents, often linked to antipsychotics like haloperidol and fluphenazine. Symptoms include mental changes, fever, muscle rigidity, and autonomic instability. This is suspect a rare case of Quetiapine-induced NMS and rhabdomyolysis.

Case Report

A 47-year-old woman with a 25-year history of bipolar I disorder presented to the emergency department with generalized weakness, tremor, and dysphagia persisting for two weeks. Her psychiatric symptoms had been managed with propranolol, alprazolam , duloxetine, lithium , flunitrazepam, and triazolam. Upon examination, she exhibited severe dysphagia, an apathetic expression, limb tremors, and generalized weakness. She had restricted herself to a liquid diet, refusing solid food, and displayed hand clenching with noticeable rigidity.

Initial investigations, including brain CT and laboratory tests, revealed no significant findings. An NG tube was inserted due to her dysphagia; however, she experienced a choking episode in the psychiatric ward, resulting in dyspnea and oxygen desaturation to 95% under a non-rebreather mask. Tachypnea, with a respiratory rate of 30-40/min, and blood gas analysis revealed respiratory acidosis and elevated lactate levels. She was transferred to the MICU for suspicion of aspiration pneumonia and impending respiratory failure.

In the MICU, intubation was performed without complications. Empiric treatment with Tazocin and low-dose Levophed was initiated for suspected septic shock. After three days, her condition improved, and she was extubated. Despite resolution of her respiratory issues, intermittent tremors persisted. The psychiatric team advised focusing on infection management before addressing her psychiatric symptoms.

During her stay, the patient exhibited acute delirium with involuntary movements. Her mood stabilizers, lithium, and lurasidone were continued, while quetiapine was increased to 100 mg HS as per psychiatric recommendation. A lumbar puncture ruled out CNS infection, but her psychotic symptoms worsened. Quetiapine was further titrated to 200 mg HS. Following medication adjustments, the patient developed high spiking fevers, body tremors, and muscle rigidity. Lab results revealed an extremely elevated CPK (51,190 U/L), creatinine (3.3 mg/dL), and tea-colored urine, indicating rhabdomyolysis. She was readmitted to the MICU due to unstable vital signs, prompting the cessation of all antipsychotic medications.

For suspected malignant hyperthermia and rhabdomyolysis, sedation with Dormicum was initiated. Lasix was administered as a diuretic to manage acute kidney injury secondary to rhabdomyolysis. The patient underwent a single session of continuous renal replacement therapy (CRRT) for metabolic acidosis and fluid overload. After aggressive fluid resuscitation, her urine

output improved, and lithium levels were confirmed within the upper normal range. A significant reduction in body temperature and CPK levels was observed following the discontinuation of quetiapine and adequate hydration.

Conclusion

This is suspect a rare case of Quetiapine induced neuroleptic malignant syndrome with acute kidney injury and rhabdomyolysis, recovered after holding Quetiapine and adequate hydration. It may be a clinical pitfall for patients who got unexpected fever and agitation during hospital course. Additionally, patients on quetiapine should be closely monitored for signs of dopamine deficiency, such as rigidity, akinesia, tremors, and tardive dyskinesia. Monitoring creatine phosphokinase (CPK) levels is also essential to detect rhabdomyolysis. Early recognition and prompt discontinuation of quetiapine can help prevent these potentially life-threatening complications.

病例報告 113_C146

嗜酸性肉芽腫性多血管炎：以影響小管徑血管之血管炎的兩位病例報告

Eosinophilic granulomatosis with polyangiitis(EGPA): 2 case reports involving small vessels vasculitis (SVV)

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Introduction

Asthma (one of the obstructive airway disease) with eosinophilia may coexisted with nasal polyps, especially in patients with eosinophilic granulomatosis with polyangiitis(EGPA). Herein, we reported two case reports with EGPA treating with mepolizumab.

Case Report

A 46-year-old man with chronic rhinosinusitis s/p functional endoscopic sinus surgery (FESS) in 2019/8 and 2024/6 proved to have polypoid change in pathology. He was also a patient with bronchial asthma with irregular follow-up at LMD and our chest OPD. The eosinophil count in 2022/2 was 1082/ul, and was up to 15650/ul in 2024/7. He also had bilateral bronchiectasis, elevated IgE, and negative for antineutrophilic cytoplasmic antibodies (ANCA) and Aspergillus fumigatus IgE. As the criteria of EGPA was met by 2022 ACR/EULAR classification criteria, mepolizumab was requested since 2024/9 instead of dupilumab by his self-pay.

Another 39-year-old man with bronchial asthma proved by methacholine challenge test positive in 2021/12 received symbicort rapihaler. He was also a patient with mononeuritis multiplex proved by nerve conduction velocity in 2022/9. The IgE was high (3478 in 2022/8), eosinophilia (6950 in 2022/9), and P-ANCA positive (1:40 X) & ANCA(PR3)(ELISA): 2.19 (negative; Max 30).

EGPA was confirmed, and then meprolizumab 300 mg subcutaneous injection. 2 months after meprolizumab injection, his oral glucocorticoids was totally tapered off.

Discussion

EGPA was a rare disease characteristics by small vessel vasculitis. As one can pay much more attention to asthma co-existed chronic rhinosinusitis with eosinophilia. As one can confirm EGPA, the biological agent “meprozilumab: can work very well.

Conclusion

EGPA is a rare disease, but early diagnosis made outcome less miserable. In addition, oral glucocorticoid tapering is another important issue for these patients.

病例報告 113_C147

肺炎克雷伯菌腦膿瘍：一病例報告及文獻回顧

Brain abscess caused by Klebsiella pneumoniae: a case report and literature review

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Introduction

Kelebsiella pneumoniae (K. pneumoniae) is a Gram-negative bacillus, and exists in human oral cavity and intestines. K. pneumoniae can cause various type of infection but K. pneumoniae related brain abscess is relatively uncommon. Herein, we report a patient infected with disseminated K. pneumoniae and brain abscess, which progressed on antibiotics, and successfully treated with surgical resection and prolonged antibiotic therapy. We also reviewed the literature on the prevelence, presentation, treatment and outcome of K. pneumoniae related brain abscess.

Case Report

A 59-year-old man visited emergency department with a 4-day history of right lower chest pain and productive cough. Physical examination disclosed right lower lung crackles. Lab data showed leukocytosis, neutrophilia, C-reactive protein elevation. Computed tomography showed multiple consolidative lesions over right lower lung and right side pleural effusion. The blood cultures yielded Klebsiella pneumonia. Intravenous antibiotic with Ceftriaxone was prescribed. The followed chest xray showed progressed right side pleural effusion. Empyema was diagnosed, and thoracoscopic decortication of pleura was performed. The abscess culture also yielded K. pneumoniae. The patient was discharged with oral Cefixime.

However, the patient visited emergency department again due to left-side limbs weakness. Magnetic resonance imaging of brain showed an lobulated lesion at right frontal lobe. He received craniotomy and a yellowish abscess with capsule 3.2 cm in diameter was found. The pathology report revealed suppurative inflammation. He received intravenous antibiotic with ceftriaxone for 30 days and was discharged with oral cefixime without neurologic deficit.

Discussion

K. pneumoniae brain abscess is an important issue in Taiwan. The mortality rate is 27%. Risk factors includes diabetes mellitus, alcoholism, etc. Infection routes includes direct spread, adjacent infection, or hematogenous spread. Common localization including supratentorial region and temporal lobe. Glasgow coma scale (GCS) greater than 12 are associated with better outcome. Neurosurgeon should be consulted at the time of initiation. For patients with brain abscess less than 2.5 cm in diameter, drainage may be not required. For brain abscess larger than 2.5cm in diameter, resection resulted in improved neurologic status.

No guideline on the usage of systemic steroids in treating bacterial brain abscesses. The duration of antimicrobial therapy is 6 to 8 weeks. Combine medical and surgical management, the cure rate could exceed 90%, which can reduce residual neurologic deficits.

Conclusion

Brain abscess caused by K. pneumoniae is relatively uncommon but should be considered in Taiwan. Surgical intervention should be considered in addition to antibiotic treatment, especially for those with large brain abscess. Adequate antimicrobial therapy for K. pneumoniae brain abscess up to 6-8 weeks is required.

病例報告 113_C148

大量冠狀動脈內血栓導致 NSTEMI 的循序治療：病例報告

A sequential approach to the management of a massive intracoronary thrombus in non-ST elevation myocardial infarction: a case report

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Introduction

Thrombus-laden coronary lesions present a particular challenge to the interventional cardiologist. Despite the development of multiple strategies to attack this problem, lesions with angiographically visible thrombus still carry a high risk of complications when coronary intervention is attempted. We would like to present a case of subacute inferior myocardial infarction presented as NSTEMI secondary to massive thrombus in an ectatic RCA. Sequential treatment with intracoronary urokinase injection, followed by systemic thrombolytic therapy with urokinase, and final coronary angioplasty over culprit lesion to achieve a satisfactory result. This case illustrates the potential benefit of combining various strategy to treat intracoronary thrombus.

Case Report

This is a 40-year-old gentleman who suffered from chest tightness in recent 1 week. He is a heavy smoker but denied any other systemic medical disease such as diabetes or hypertension. Elevated cardiac enzymes with T wave inversion was found over inferior leads on complete EKG. The coronary angiography showed total occlusion over proximal RCA with hazy antegrade blood flow over middle RCA and thrombus was highly suspected. However, collateral flow from
LAD was noted by retrograde flow over posterolateral branch of RCA. Chronic total occlusion cannot be excluded and intracoronary injection with urokinase 120000unit was done. Some small antegrade flow was found and continuous thrombolytic therapy was decided. After systemic thrombolytic therapy with urokinase 1000000unit/day for 5 days, good coronary flow was found over whole RCA and distal culprit lesion was noted with 90% stenosis. After balloon dilatation and stenting, we got an excellent result. The patient was discharged on the next day without any sequela.

Discussion

There is a consensus that intracoronary thrombus is a challenging target for revascularization because of its unique characteristics. It has a crucial impact on the performance and outcome of the percutaneous coronary intervention. The coronary thrombus can occlude the coronary artery and its branches, impairing epicardial and myocardial blood flow. The thrombus size and consistency are also important prognostic markers of distal embolization. Multiple studies have shown that distal embolization is associated with an increase in mortality.

Conclusion

Coronary artery thrombus occurs due to rupture or preexisting coronary artery plaque erosion, resulting in the artery's complete occlusion. It manifests clinically as an acute coronary syndrome,

including STEMI, NSTEMI, and unstable angina. For large thrombus burden in ectatic coronary artery, thrombolytic therapy may prevent more distal embolization compared with direct thrombus suction. In our case, good antegrade coronary flow was found after thrombolytic therapy and total revasculization was performed with plain old balloon angioplasty with stent successfully.

病例報告 113_C149

一位愛滋病患者罕見同時合併直腸卡波西氏肉瘤及巨細胞病毒直腸炎

Rare concurrent rectum Kaposi sarcoma with CMV colitis in a patient with HIV

許朝欽

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Introduction

Kaposi sarcoma is the most common neoplasm in patients with acquired immune deficiency syndrome (AIDS) and the gastrointestinal tract is a frequent site of visceral involvement. However, combined with CMV infection was rare. Hence, here we reported a HIV case with combined infection.

Case Report

We reported a 36-year-old male with a history of human immunodeficiency virus admitted dud to chronic cough for three weeks. This patient had irregular control for his HIV with Triumeg. This time, he had chronic cough for 3 weeks, associated with yellow sputum, fever, chest pain, and shortness of breath. Previous this episode, he had been to Indonesia. Physical examination revealed fever, tachycardia, pale conjunctiva, and bilaterally crackles breathing sound. Laboratory data revealed anemia (9.9g/dL), elevated CRP (166 mg/L), and low CD4 amount(22 cells/mm^3). Further chest CT image study showed bilateral pneumonia. Symptoms of this patient improved after antibiotics used. However, due to persistent anemia, esophageal gastric duodenal endoscopy and colonoscopy were preformed, and revealed esophageal candidiasis and rectum colitis with submucosal nodules, which further pathology were Kaposi sarcoma and cytomegalovirus infection. Typical picture with spindled endothelial cells and present of human herpesvirus 8 (HHV8). CT image reported with. Kaposi sarcoma with extraserosal extension. Then this patient received chemotherapy with liposomal doxorubicin and discharge.

Discussion

AIDS is the final stage of HIV infection and confers increased risk of opportunistic diseases due to immunosuppression, especially in the setting of low CD4 count. CMV colitis risk is higher when CD4 count is less than 50 and Kaposi sarcoma risk is higher when CD4 count is less than 100. Kaposi sarcoma is the most common cancer with HIV patients, and mostly were in gastrointestinal tract. CMV is also the common cause of gastrointestinal tract. There were only three reports for combined Kaposi sarcoma in gastrointestinal tract and another one report combined with TB infection of rectum. The endoscopy finding of Kaposi sarcoma included submucosal nodules, polypoids, and mass lesions with dark red mucosa. However, the endoscopy characteristic of CMV colitis varied. Hence, delayed diagnosis may lead to poor prognosis.

Conclusion

Opportunistic infection was critical for HIV patiens. Although Kaposi sarcoma combined with CMV infection of gastrointestinal tract was rare in HIV patients, early dection is important for those with immunosuppression patients.

病例報告 113_C150

異位性庫欣氏症候群合併惡性高血鈣

A case report of ectopic cushing syndrome and malignancy hypercalcemia

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Introduction

Paraneoplastic syndrome has variable presentation due to the function of hormone secretion, such as hypercalcemia from PTH rp (Parathyroid hormone-related protein) or ectopic cushing syndrome from excessive ACTH or ACTH-like substance. It also leads to poor prognosis. Although we could do symptomatic treatment, the crucial way to cure paraneoplastic syndrome is to treat the underlying tumor. Not until tumor burden decreased that the condition would be improved sequentially.

Case Report

A 82-year-old woman was admitted to hospital on 2024/03/26 due to sudden onset of consciousness disturbance within hours. The record past history were hypertension, diabetes mellitus, dyslipidemia, chronic heart failure with preserved ejection fraction and paroxysmal atrial fibrillation. High calcium and cortisol level was noted with CT scan showed multiple relative poor enhancing soft tissue lesion around both liver lobes and focal small nodular foci around both upper lung. Pathological report for hepatic CT-guide biopsy was carcinoma with undetermined origin with possible of pancreatobiliary tract, upper gastrointestinal tract, breast, lung or gynecological tract. The immune stains were CK7(+), CK20(-), TTF-1(-), synaptophysin(-) and chromogranin A(-).

Discussion

There were three case-reports below we had reviewed. The first case was a 6 year-old girl with hepatoblastoma. She had Cushing's syndrome appearance including moon face, buffalo hump, central obesity, and hirsutism. Lab data indicated hypercalcemia (14.2 mg/dl) with normal PTH (parathyroid hormone) level and high ACTH(819 pg/ml) after dexamethasone test were also noted. Abdominal CT indicated a giant liver tumor of the right lobe with a calculated volume above 1000 ml. Eventually, the tumor was surgically removed smoothly.

The second case was 74-year-old non-smoker woman with pulmonary mucoepidermoid carcinoma. CT reported a 9.1* 6.4 (cm) left lower lobe necrotic lung mass. Lab data revealed hypercalcemia (14.84 mmol/L) with normal PTH level. Hormone test revealed high cortisol(45.65 ug/dL), ACTH(76.7 pg/mL) and 24-hour urine cortisol level(527.04 ug/day). High cortisol level(23.72 ug/dL) was also noted after overnight dexamethasone test. There was no classic cushing appearance in this patient.

The last case was a 62-year-old woman who was diagnosed with malignant mesothelioma. Lab data revealed hypercalcemia to 13.3 mg/dL with low PTH(10 pg/mL) and PTHrP(<0.3 pmol/L). In addition, normal 25-Hydroxyvitamin D(calcidiol) and high 1,25-Dihydroxyvitamin D(calcitriol) levels were noted(each 55ng/mL & 118 pg/mL). We also discovered high cortisol (30 ug/dL) and

ACTH (50 pg/mL). 24-urine cortisol was examined with 360 (ug/day).

Conclusion

We could learn the strategy of diagnosing ectopic cushing syndrome according to special clinical manifestation. Although poor prognosis is common for these patients, we could do our best to release their discomforts.

病例報告 113_C151

直腸癌轉移造成的腦下垂體中風-一例罕見的病例報告

Pituitary apoplexy caused by colorectal adenocarcinoma metastasis- a case report

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Introduction

Pituitary apoplexy is a rare endocrine emergency that may be caused by infarction or hemorrhage of the pituitary gland. Clinical symptoms vary among patients. The major cause is pituitary adenoma and was rarely due to cancer metastasis with only a few care reports. Here we presented a case of colon adenocarcinoma with pituitary gland metastasis complicated with pituitary apoplexy.

Case Report

A 50-year-old female with colorectal adenocarcinoma stage IV was admitted to our hospital for urinary tract infection due to tumor compression-related bilateral hydronephrosis. Consciousness disturbance was noted one day with hypotension, hypoglycemia and fever. Brain MRI then revealed a long T2 change around the pituitary stalk and gland, which pituitary apoplexy was suspected. After treatment her consciousness improved dramatically then she was discharged. However, one week later she complained about bilateral eyes blurred vision, and the left eye progression. She was then transferred to an ophthalmologist which showed left abducens nerve palsy with pupil dilatation. She was then transferred to Tamshui MMH for further operation.Pre-operation MRI showed a 2.5cm lobulated enhancing soft tissue mass involving the sellar & suprasellar region. So the operation was arranged via an endoscopic endonasal approach and a 1.9cm yellow and white fragile tumor was located above the sella. The final report came out and was colon cancer metastasis. After surgery central diabetes insipidus was noted and she was transferred back to Taitung MMH for further palliative care.

Discussion

Pituitary apoplexy may be caused by ischemic infarction or hemorrhage of the pituitary gland. Clinical symptoms can be divided into two main components: neurologic and hormonal symptoms. Neurologic complications are mostly caused by compression of other adjacent structures, including headache, and loss of visual acuity of visual field defect. On the other hand, hormonal complications are mostly caused by hypopituitarism. The major cause of pituitary apoplexy is pituitary adenoma. Other precipitating factors include pregnancy, previous radiation therapy, coagulopathy, and rarely malignancy. There were only a few case reports were cancer metastasis-related. Brain MRI was used for diagnosis and we may see distinct image presentations during different phases. There were two Pituitary Apoplexy Score (PAS) proposed to grade the severity of patients. Though there was no consensus for management, patients who presented without visual symptoms or neurologic symptoms were preferred to receive conservative treatment with close follow-up; and those who had neuro-ophthalmologic symptoms were indicated for surgery intervention. Hormonal deficiency especially anterior pituitary hormones

was noted after surgery and might require a lifelong supplement.

Conclusion

Pituitary apoplexy may have various severities. Some may have no symptoms but some might present adrenal insufficiency or ophthalmologic symptoms which indicate rapid medication and surgery. However, for patients with mild or no symptoms, the timing for surgery intervention may be challenging. We may consider using pituitary apoplexy score and modify further treatment options case by case.

病例報告 113_C152

腹膜透析病人產生的腫瘤樣鈣質沉著症

Uremic tumoral calcinosis on peritoneal dialysis patients

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Introduction

Uremic tumoral calcinosis (UTC), a metastatic soft tissue with calcification, which usually deposited at the weight-bearing joint, such as shoulder or hip. It is a relatively rare complication in end-stage renal disease (ESRD) patients, developed due to uncontrolled phosphorus, calcium and intact parathyroid hormone (iPTH).

Case Report

We retrospectively reviewed 250 ESRD patients on PD at Taipei and Tamsui Mackay memorial hospital in recent ten years. This review enrolled 4 patients on PD. All patients were diagnosed as UTC by radiologic. They are undergoing chronic peritoneal dialysis around 4-7 years, all the cause of ESRD is chronic glomerulonephritis. There are two male and two female, and the age is around 44- to 54-year-old. The deposition of tumoral calcinosis is demonstrated at large joint, especially at shoulder and hip. Phosphorus in four patients is poorly controlled, around 6-10 mg/dL. Calcium is within a range from 8-11 mg/dL. One of patient who had parathyroidectomy (PTX) in the past, so her iPTH is low, about 50 pg/mL. Two patients have severe hyperparathyroidism (iPTH>1800 pg/mL), followed by parathyroidectomy, and one has hyperfunctioning parathyroid gland with mild elevated iPTH (400-600 pg/mL). With treatment includes parathyroidectomy and phosphate-lowering agents, partial remission of calcinosis were achieved.

Discussion

UTC is an unusual complication, develops in ESRD patients with uncontrolled hyperphosphatemia and hyperparathyroidism. It always presents on weight-bearing joints. There are many kinds of treatments of UTC, but the outcome is not satisfied. In our patient, UTC is just partial remission after parathyroidectomy. For the management of UTC, the most important thing is intensively controlling the serum phosphorus and hyperparathyroidism.

Conclusion

In patients with ESRD, UTC is a one of complication associated with poor management of hyperphosphatemia and hyperparathyroidism. Therefore, long-term control measures are recommended to prevent the development of UTC. Once UTC occurs, it is recommended to intensively manage serum phosphorus levels and hyperparathyroidism to mitigate further symptoms.

病例報告 113_C153

36 歲男性以體重減輕為表現典型的泛腦垂體機能低下症

36 years old man with initials body weight loss and typical presentation of pan hypopituitarism

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Introduction

Pan-hypopituitarism is a condition of inadequate or absent production of the anterior pituitary hormones. It is frequently the result of other problems that affect the pituitary gland and either reduce or destroy its function or interfere with hypothalamic secretion of the varying pituitary-releasing hormones. Pan-hypopituitarism can be the end result of various clinical scenarios. The signs and symptoms are diverse.

Case Report

A 36 years old man was admitted to general medical ward due to fever. According to old chat record, he had history of brain tumor 3.7x4.6x4.9 cm hypodense lesion over supra sella area and he took right modified fronto-orbit-zygomatic craniotomy on 2023/11/06 and bed ridden status since then. Review back to history, his mother said that sudden body weight loss 20kg within five to six month with general weakness and mother brought to emergency department. Patient cognitive function become declined and blurred vision was mentioned.

Nursing home nurse noticed fever with much sputum also low BP 86/43mmHg and he was readmitted to our hospital under impression of hyponatremia, pneumonia.

At presentation, his consciousness was stupor, only eye response when calling. His arterial blood pressure revealed 83/43 mmHg at nursing home and become 106/54 mmHg at emergency department and a heart rate of 94/min. He had fever with mild respiratory distress.

Physical examination reveal that thin body built and chronic ill looking status, comprehensive neurological examination were not detected. His eye movements were free and full. Eye-brows were slightly thinner. His neck was supple. Hearts sounds were regular without murmurs. The lungs show rhonchi on both sides. The pigmentation of areola was diminished. The abdomen was soft and non-tender. There was sparse axillary and pubic hairs. The extremities showed spastic contracture with atrophies. The peripheral pulsations were normal. Muscle powers and deep tendon reflexes of all four limbs were reduced.

Complete blood count show mild leukocytosis and serum chemistry tests were within normal limit except hyponatremia 120 mmol/L. The data were summarized in table 1. Chest X ray film show mild infiltration.

The hormonal profile was summarized in Table 2. The reduced morning cortisol, ACTH, free thyroxine (free T4), thyroid stimulating hormone (TSH), reduced testosterone and follicular stimulating Hormone, luteinizing hormone were compactible with pan- hypopituitarism. The patient was given with thyroxine 100mcg once daily for secondary hypothyroidism and intravenous hydrocortisone 100mg every 8 hourly then shift to oral cortisone 25mg 1tab at morning and 1/2tab at evening for secondary hypo-cortisolism. His condition become improved

gradually.

Discussion

Hypopituitarism is defined as deficiency of one or more pituitary hormones produced in the anterior lobe and posterior lobe, including GH, the gonadotropins follicle stimulating hormone (FSH) and luteinizing hormone (LH), ACTH, thyroid stimulating hormone (TSH), prolactin, oxytocin, and anti-diuretic hormone (ADH).

Metabolic encephalopathy can also present with disorders of consciousness and it can be differentiated from NCSE by etiology, laboratory examination, and EEG. While increasing slowing of the waking background frequency was observed in hyperglycemia and hypoglycemia, hyponatremia may initially produce posterior slowing followed by more diffuse delta activity, hypopituitarism and hypoadrenalism may cause diffuse theta and delta activity.[1]

Hypopituitarism can have a range of presentations with the most common electrolyte abnormality on lab work being hyponatremia which is usually secondary to adrenal insufficiency or central hypothyroidism. Both adrenal insufficiency and central hypothyroidism can cause decreased free water clearance leading to hyponatremia. Adrenal insufficiency also causes an increase in anti-diuretic hormone contributing to hyponatremia.[2]

病例報告 113_C154

原發性體液淋巴瘤伴後腹腔淋巴結轉移在非人類免疫缺乏病毒感染的肝硬化患者，一例病例報告

Primary Effusion Lymphoma with Retroperitoneal Lymph Node Involvement in a Human Immunodeficiency Virus-Negative Patient with Cirrhosis

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Introduction

Primary effusion lymphoma (PEL) is a rare and aggressive subtype of large B-cell lymphoma characterized as a lymphomatous effusion in the body cavities, which is universally associated with the human herpesvirus 8 (HHV8), human immunodeficiency virus (HIV) infection and severe immunodeficiency conditions. In this case report, we describe an instance of HHV8-positive PEL in a patient with alcoholic liver cirrhosis but without HIV infection.

Case Report

A 57-year-old male had a long-standing history of alcohol abuse with medical history included type 2 diabetes mellitus, chronic kidney disease and alcoholic liver cirrhosis. Between 2005 and 2020, he experienced multiple hospital admissions due to alcohol- and cirrhosis-related complications. The patient presented to hospital with abdominal fullness and poor appetite in November 2021. Physical examination revealed a distended abdomen with shifting dullness and abdominal sonography confirmed massive ascites. The fluid analysis revealed liver cirrhosis with portal hypertension. The patient was diagnosed with spontaneous bacterial peritonitis and treated with antibiotics. Cytologic and cell block analysis revealed abundant polymorphonuclear leukocytes but no abnormal lymphoid cells. He waitlisted for liver transplant in December 2021 due to progressive deterioration of liver function and refractory ascites. In July 2022, computed tomography revealed enlarged retroperitoneal lymph node, which were subsequently confirmed by positron emission tomography. The ascites analysis yielded a predominance of 79% monocytes. Ascites cytology revealed atypical lymphoid cells. Multinucleated cells resembling Reed–Sternberg cells were also observed. Immunohistochemical staining demonstrated expression of CD138, CD30, and HHV-8 latency-associated nuclear antigen (LANA) of the atypical lymphoid cells. The flow cytometry study indicated an abnormal large-sized cell clone (88.3% of total nucleated cells) in the ascites which was positive for CD45 and negative for CD19, CD20, CD79b, CD10, CD38, and CD23. Primary effusion lymphoma was diagnosed. He received two courses of a reduced-dose CHOP regimen (cyclophosphamide, doxorubicin, vincristine, prednisone) followed by one course of a reduced-dose EPOCH regimen (etoposide, prednisone, vincristine, cyclophosphamide, doxorubicin). Unfortunately, he succumbed to disease progression.

Discussion

PEL primarily affects HIV-positive individuals and immunocompromised HIV-negative patients. A prior large case series study reported that PEL patients in Taiwan were more frequently unrelated to HIV. PEL cells typically exhibit large, multinucleated or multilobed lymphoid morphology with

prominent nucleoli, basophilic cytoplasm containing small vacuoles, and high mitotic activity. Immunophenotypically, these malignant cells are CD45-positive but negative for B-cell (CD19, CD20, CD79a) and T-cell (CD3, CD4, CD8) markers. PEL can present in lymph node or as solid tumors in extra-nodal sites, including the central nervous system, gastrointestinal tract, lung, and skin. Our case involved both liquid and solid components. Patients with PEL respond poorly to chemotherapy and have a very poor prognosis. Treatment options include conventional chemotherapy regimens used for non-Hodgkin lymphoma, such as CHOP or EPOCH. Our patient received two courses of reduced-dose CHOP regimen followed by one course of reduced-dose EPOCH regimen. Unfortunately, he died of progressive disease.

Conclusion

This case underscores the importance of carefully examining ascitic fluid of patients with refractory ascites for early diagnosis and effective treatment for this rare lymphoma subtype.

病例報告 113_C155

中度阻塞型睡眠呼吸中止症及週期性腿部抽動症

Obstructive Sleep Apnea (Moderate) and Periodic Limb Movement Disorder

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Case Summary

The patient came to sleep medicine OPD for chief complaint of daytime sleepiness, and periodically involuntary and repetitively stereotypic leg movement during sleep for 2-3 years. He had also frequent snoring and impaired restorative sleep for few years. He had HTN, nightly insomnia. He denied of DM, CKD, neurodegenerative disorder and intake of anti-depressants. EPSS was 6. BW was 90 kg and Ht. of 170 cm, BMI of 31, neck circumference of 37 cm was noted. On PE, no pallor, Mallampati score of 2 and tonsillar hypertrophy of Gr.2, marked lateral peritonsillar narrowing were noted. Firstly, we prescribed clonazepam for initial 3 weeks. On 2nd time visit, nocturnal leg symptoms still unimproved, therefore we arranged nighttime PSG as we suspected both sleep related movement and breathing disorders. Total sleep time was 5.8 h, N2 stage was 70%. AHI of 19.6/h, OHI of 17/h, and Snore index was 284/h. His ODI was 12.8/h and mean SpO2 was 92%. Periodic limb movement index (PLMI) was 15.5/h, and non-PLMI was 77/h, and respiratory arousal index is 6/h. Periodic leg movement of sleep (PLMS) mostly occurred during deep sleep (N2 and N3 stages). So, final diagnosis was OSA (moderate) and PLMD. Then, we shifted to pregabalin in daily dose of 100mg as he also had nightly insomnia and HTN and anxiety disorder which are common side effects of dopamine agonists. Serum Fe profile and Mg level were WNL. Then, we titrated the dose up to 300mg upon leg symptoms and omit sedatives. On 3rd month of daily pregabalin dose in 300 mg, leg symptoms improved significantly and EPSS down to 3 but snoring persisted and didn't complain of any adverse effect of pregabalin. For OSA, he chose adoption of lifestyle modification and regular physical activities for body weight reduction as initial choice. We will do follow up post-pregabalin PSG and will suggest CPAP therapy for pneumatic splint of upper airway if worsened OSA symptoms and/or worsening AHI.

Discussion

Nightly insomnia and daytime symptoms are probably related to frequent nocturnal sleep disruption due to frequent, short lasting, repetitive PLMS and OSA. Physiologically, sleep influences cardiovascular homeostasis (that is cardiac physiology and vascular tone) through autonomic nervous system and circadian rhythm. Sympathetic hyperactivity related to both PLMS and OSA-induced respiratory related arousal, lead patients more vulnerable to greater heart rate variability and substantial basal sympathetic predominance during the whole sleep time and sleep deprivation, causing increased coagulation dysfunction and further detrimental effects like increased risk of CV morbidity and mortality if his OSA and PLMD were not diagnosed and not under adequate control.

Conclusion

Physical inactivity, male gender and AHI are positively correlated with PLMS in patients with OSA

and PLMD. HTN also correlates with severity and duration of PLMS. Selection of therapy for PLMD depends on disease severity, patient age, comorbidities (e.g. IDA), drug side effects and patient preferences after exclusion of RLS, RBD and narcolepsy. Goals of therapy are to reduce symptoms, and improve daytime function, sleep and quality of life.

病例報告 113_C156

麴菌肺積膿發生於支氣管擴張個案:少見的病例

A Case Study on a 74-year-old Male Patient with Bronchiectasis developed lung Empyema

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Introduction

This study aims to investigate the case of a 74-year-old male patient with a history of bronchiectasis, who presented with a persistent cough and chest pain for 2 months. The patient underwent evaluation for pulmonary empyema using chest X-ray, chest ultrasound, and blood tests, which suggested the presence of lung abscess. The recommendation for diagnostic aspiration and treatment was given. Due to unbearable chest pain, the patient consented to undergo surgical intervention on the following day, which included chest tube insertion and pleural biopsy. The pleural biopsy revealed a significant amount of Aspergillus. However, the fungal culture of the pleural fluid was negative. The patient was discharged after receiving appropriate drainage and antifungal therapy, but continued to experience chest pain and wound discharge. Following further chest tube drainage, the symptoms improved, and the patient was kept on follow-up treatment.

Case Report

A 74-year-old male patient with medical history of bronchiectasis, symptoms: Persistent cough and chest pain for 2 months. CXR, chest ultrasound, blood tests, pleural biopsy, fungal culture of pleural fluid were examined. Treatment with diagnostic aspiration, surgical intervention, drainage, antifungal therapy- Chest tube drainage, symptom observation, follow-up treatment. CXR and chest ultrasound revealed pulmonary empyema, pleural biopsy showed a significant amount of Aspergillus Fungal culture of pleural fluid was negative.

Discussion

In this case, the patient had a history of bronchiectasis and presented with persistent cough and chest pain. Based on the findings from chest X-ray and chest ultrasound, pulmonary empyema was impressed. However, determining the specific causative pathogen of the pulmonary empyema proved to be challenging. The pleural biopsy showed a notable presence of Aspergillus, but the fungal culture of the pleural fluid turned out to be negative. This could be attributed to a low quantity of fungi in the sampled pleural fluid or limitations in the culturing technique. Due to the severity of the condition and intolerable symptoms, the patient agreed to undergo chest tube insertion and surgical intervention. After the treatment, the symptoms improved.

Conclusion

Aspergillus empyema is uncommon, pleural drainage Anti-fungal agents and aggressive surgical intervention can provide effective disease control for improving outcome. The importance of considering fungal infections in patients with empyema, especially those with underlying bronchiectasis anti-fungal therapy is crucial. Continued follow-up is necessary to ensure resolution of symptoms and prevent recurrence.

病例報告 113_C157

芽枝狀枝孢菌肺炎與慢性阻塞性肺病急性惡化:罕見的病例報告

Claudosporium pneumonia and COPD with acute exacerbation (AE).

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Introduction

This study aims to report a case of a taxi driver with concomitant Claudosporium pneumonia and COPD with acute exacerbation (AE). The patient frequently sought medical attention in the past due to respiratory distress, but the symptoms ceased after treatment with the antifungal medication Itraconazole. Chest X-rays and computed tomography showed improvement in the right upper lung lesion. Currently, the patient remains stable and is undergoing treatment with a triple inhaler and antifungal medication.

Introduction. Claudosporium is a rare fungus that can pose a threat to human health. COPD is a chronic lung disease that has been associated with respiratory infections and acute exacerbations (AE). This study reports a rare case of combined Claudosporium pneumonia leading COPD with AE. The objective of this study is to report the patient who developed changes in the right upper lung lesion as seen on chest X-rays and computed tomography, along with concomitant Claudosporium pneumonia and COPD AE. Additionally, we aim to explore the patient's condition and treatment outcomes after using itraconazole

Case Report

The case involved a taxi driver diagnosed with Claudosporium pneumonia from sputum culture and environmental exposure history. The patient had frequent emergency department visits due to respiratory distress. During treatment, the patient received Itraconazole, antifungal medication. Following treatment with Itraconazole, the patient's condition improved significantly. Chest X-rays and computed tomography showed changes in the right upper lung lesion. The patient's respiratory distress improved, and there were no further COPD AE episodes.

Discussion

his case report highlights the importance of Claudosporium pneumonia in COPD patients and suggests the potential efficacy of antifungal medication in treating this infection. However, further research is needed to determine the optimal duration of Itraconazole treatment for COPD with concomitant Claudosporium pneumonia.

Conclusion

Claudosporium pneumonia is a rare infectious lung disease that holds clinical significance, especially in COPD patients with mold exposure. This study reports a rare case of successfully controlling COPD with AE and concomitant Claudosporium pneumonia using antifungal medication and a triple inhaler.

病例報告 113_C158

肺腺癌接受表皮生長因子受體一酪氨酸激酶抑制劑治療期間出現肺結核

Tuberculosis Pleurisy Development During Epidermal Growth Factor Receptor Tyrosine Kinase Inhibitor Therapy for Advanced Lung Adenocarcinoma with Malignant Pleural Effusion

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Introduction

Lung cancer is the most common cancer and the leading cause of cancer-related deaths in Taiwan. Malignant pleural effusion (MPE) is frequently observed in patients with lung adenocarcinoma. Epidermal growth factor receptor (EGFR) mutations are prevalent in the majority of female, non-smoking Asian patients with lung adenocarcinoma. Treatment with EGFR tyrosine kinase inhibitors (TKIs) significantly improves the prognosis of patients with advanced lung adenocarcinoma harboring EGFR gene mutations. However, the development of tuberculosis pleurisy in patients with lung adenocarcinoma and MPE undergoing EGFR-TKI therapy is rare. Here, we report the case of a 90-year-old female with lung adenocarcinoma (exon 19 deletion) and MPE who developed tuberculosis pleurisy after 1 year of EGFR-TKI therapy.

Case Report

A 90-year-old female was admitted with left-sided pleural effusion. Her medical history included abdominal aortic aneurysm and an old cerebrovascular accident, with prolonged bedridden status for two years. Echo-guided thoracocentesis and a CT-guided lung biopsy on August 29, 2023, revealed adenocarcinoma (acinar component, well-differentiated, exon 19 deletion) with left malignant pleural effusion. The pleural fluid adenosine deaminase (ADA) level was 15 U/L, and acid-fast bacilli (AFB) tests and mycobacterial cultures were negative. Imaging studies, including chest CT, brain MRI, bone scans, and abdominal ultrasound, indicated cT1cN2M1c, stage IVA disease, with an ECOG performance status of 3-4. Afatinib 30 mg per day was prescribed starting on September 22, 2023. The patient experienced grade 2 side effects, including a skin rash and diarrhea. Despite a reduction in pleural effusion volume during afatinib treatment, the effusion did not fully resolve. Several thoracocenteses were performed, consistently yielding adenocarcinoma cells, while AFB tests and mycobacterial cultures remained negative. However, on September 12, 2024, the pleural fluid mycobacterial culture tested positive and identified Mycobacterium tuberculosis (MTB).

Discussion

Exon 19 deletion is a sensitive mutation of the EGFR gene in non-small cell lung cancer (NSCLC). Previous studies have demonstrated a significant response to first- and second-generation EGFR-TKIs in NSCLC patients with EGFR mutations. In this case, we prescribed a second-generation EGFR-TKI (afatinib 30 mg QD) for this elderly female patient with adenocarcinoma and an exon 19 deletion. Radiographic assessment using the Response Evaluation Criteria in Solid Tumors (RECIST) indicated a partial response. Therefore, we continued periodic thoracocenteses to monitor treatment response until MTB was identified from the pleural effusion. To our knowledge,

no previous studies have reported the development of MTB after EGFR-TKI therapy in NSCLC patients.

病例報告 113_C159

Rasmussen's Aneurysm：肺動脈假性動脈瘤，一種罕見但嚴重的肺結核併發症

Rasmussen's Aneurysm: Pulmonary artery pseudoaneurysm, a rare yet severe complication of pulmonary tuberculosis

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Introduction

Rasmussen's aneurysm: Pulmonary artery pseudoaneurysm characterized by localized dilation of the pulmonary artery wall secondary to invasion of adjacent intraparenchymal tuberculous cavities in the lung, which may result in life-threatening hemoptysis.

Case Report

We reported a 44-year-old man with a history of incomplete treatment for latent tuberculosis presented with a 3-day history of blood-tinged sputum. Cavitary lesion at right upper lung was noted by chest radiograph. On the third day of admission, he experienced life-threatening hemoptysis and desaturation, leading to endotracheal intubation and intensive care unit monitoring. Computed tomography angiography revealed a pseudoaneurysm in the right upper lung cavity. Successful embolization of the right bronchial artery branch was performed. However, hemoptysis recurred on the twenty second day of admission and was resolved with subsequent embolization of the right pulmonary artery branch. Tuberculosis treatment was initiated after a positive acid-fast stain and continued at outpatient setting after discharge without recurrent hemoptysis

Discussion

To address life-threatening hemoptysis, three simultaneous goals include protecting non-bleeding lungs, identifying bleeders, and controlling bleeding. About one-third of patients with pulmonary tuberculosis may develop hemoptysis, and bronchial arteries and other systemic arteries (~90%), including the intercostal arteries, internal mammary arteries, thyrocervical arteries, and inferior phrenic arteries, are the main sources of life-threatening hemoptysis. The pulmonary artery (<10%) is not a major source of bleeding and can easily be missed during evaluation. Regardless of the origin, if left untreated, life-threatening hemoptysis has a mortality rate of over 50%. Contrast-enhanced multidetector CT angiography is considered the imaging of choice for both initial diagnosis and treatment planning. The first-line treatment is transarterial catheter embolization (TAE), with an immediate success rate of about 85% and a long-term (6-12 months) recurrence rate of 10-20%. The second-line treatment after failure is surgical intervention.

Conclusion

We aim to elucidate a rare yet severe complication of pulmonary tuberculosis, Rasmussen's Aneurysm, through this clinical case, as keeping this diagnosis in mind is pivotal for guiding effective patient management with life-threatening hemoptysis. Early collaboration with an interventional pulmonologist, interventional radiologist, and chest surgeon is imperative.

Furthermore, we advocate for the inclusion of dual pulmonary blood supply assessments when evaluating life-threatening hemoptysis, as relying solely on systemic or pulmonary circulation examinations may fail to achieve hemostasis effectively.

病例報告 113_C160

病例報告- 神經精神紅斑性狼瘡合併廣泛性全身良性淋巴結腫大

A case report of neuropsychriatric systemic lupus erythematosus with generalized lymphadenopathies

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Introduction

The diagnosis of systemic lupus erythematosus(SLE) is based on characteristic clinical features and autoantibodies[1]. Even autoantibodies were much, but specificity in diagnosis of SLE was limited.

Case Report

This 41-year-old man with hypertension and gout was in his health usual until one month before admission when subacute onset of distal numbness over four limbs developed. He was suggested to visit the emergent department by the neurologist of the local clinic. Due to diplopia was noted for 2 days, he went to our emergent department for help.

Physical examination showed no palpable mass. Laboratory test revealed leukocytosis (11730k/uL), elevated CRP level(11.2mg/dL) and procalcitonin level(1.25ng/ml), normal LDH level(183U/L), pre-diabetes mellitus (HbA1c: 6.0%), acute on chronic kidney disease (Cre: 2.69mg/dL). Whole body CT scan without contrast revealed no acute intracranial hemorrhage, but diffuse lymphadenopathies. He also reported fever, night sweat and body weight loss of 6 kilograms within one month. Mild hoarseness of his voice was mentioned in recent months too. For suspect lymphoma with acute polyneuropathies, the patient was admitted for evaluation.

After admission, empirical antibiotic of Ceftriaxone was prescribed even though there was no fever, his progressive muscle weakness persisted and he can not walk due to lower limb weakness. NCV reported bilateral sensori-motor polyneuropathy with axonal demyelinating features. CSF studies showed aseptic meningitis and IgG index more than 0.7. Core needle biopsy of right axillary lymph nodes revealed necrotizing lymphadenopathy. Excisional biopsy of left inguinal lymph nodes reported reactive lymphadenopathy. Started double filtration plasmapheresis but his neurologic problem did not improve.

There was elevated IgG and beta-2 microglobulin. Also, ANA positive. We consulted rheumatologist and systemic lupus erythematosus was impressed. Kidney biopsy found full house nephropathy. His neurologic problem were resolved after treatment with cyclophosphamide plus pulse therapy and out-of-pocket Belimumab. Finally, he can walk when he is discharged after rehabilitation.

Discussion

Lymphadenopathy occurs in approximately 40% of SLE patients, usually at the onset of disease or during disease flares. Though it is not considered a clinical criterion in the 2019 EULAR/ACR classification criteria, some reports have suggested that it can be the first manifestation, with a cumulative incidence ranging from one-third to one-half of the cases. Lymph nodes are typically soft, non-tender, discrete, and usually detected in the cervical, axillary, and inguinal area. Biopsy

commonly shows reactive follicular hyperplasia, but can have distinctive pathologic features with necrotizing lymphadenitis.

Conclusion

Classification criteria of SLE is not the diagnostic criteria. Therefore, diagnosis of systemic lupus erythematosus was still challenging although we know more about the disease in this generation.

病例報告 113_C161

一例唐菖蒲伯克氏菌引起的呼吸器相關肺炎與急性腎功能損傷

Ventilator-Associated Pneumonia and Rapid Renal Failure Due to *Burkholderia gladioli*: A Case Report

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Introduction

Burkholderia gladioli is a gram-negative, rod-shaped bacterium commonly found in the environment and known to produce bongkrekic acid (BKA) in contaminated foods. It primarily affects immunocompromised individuals, including those with cystic fibrosis. In March 2024, a food poisoning outbreak in Taiwan linked to BKA from *B. gladioli* pathovar *cocovenenans* resulted in 35 cases and six fatalities due to multi-organ failure. This report highlights a case of ventilator-associated pneumonia (VAP) with rapid renal failure caused by *B. gladioli*.

Case Report

A 35-year-old male with a medical history of type II diabetes mellitus, alcohol-related dilated cardiomyopathy, a history of ventricular tachycardia, and an implanted cardioverter-defibrillator presented to the emergency department with progressive shortness of breath and bilateral lower leg edema for one month. CXR revealed cardiomegaly and pulmonary edema. Laboratory results showed a normal complete blood cell counts, elevated BUN (66 mg/dL), elevated serum creatinine (2.5 mg/dL), and NT-proBNP of 4021 pg/mL. The patient was diagnosed with decompensated heart failure and was treated with continuous infusions of furosemide, dopamine, and dobutamine. Arterial blood gas analysis indicated mixed metabolic and respiratory acidosis. The patient subsequently underwent endotracheal intubation. Over the following days, his hemodynamic status stabilized, and renal function improved. However, on day 8 post-intubation, the patient developed a new fever and profound septic shock. A repeat CXR revealed worsening bilateral lung consolidation. Concurrently, he experienced rapid renal deterioration with anuria and worsening metabolic acidosis. Continuous renal replacement therapy (CRRT) was initiated, along with high-dose vasopressors, and antibiotic therapy was escalated to meropenem. Sputum cultures from tracheal aspirates grew *Burkholderia gladioli*, identified by matrix-assisted laser desorption ionization-time of flight mass spectrometry. Susceptibility testing revealed susceptibility to trimethoprim/sulfamethoxazole, meropenem, and levofloxacin. Despite aggressive treatment, the patient developed multi-organ failure and shock refractory to inotropic support. He suffered cardiac arrest and died.

Discussion

Burkholderia gladioli, formerly known as *Pseudomonas gladioli*, was first identified as a plant pathogen responsible for vegetation rot. Human infections caused by *B. gladioli* are rare but have been reported in immunocompromised individuals, such as lung transplant recipients, patients with cystic fibrosis, and co-infection following COVID-19. Most *B. gladioli* infections have a poor prognosis, with high mortality rates despite appropriate antibiotic therapy. These findings

highlight the need for a deeper understanding of *B. gladioli* and its pathogenesis to improve clinical outcomes.

In the present case, the patient had multiple comorbidities and was admitted with decompensated heart failure. Although he improved early in the hospital course, VAP caused by *B. gladioli* led to refractory septic shock, hypoxemia, acute kidney injury, and rapidly progressing metabolic acidosis. Although the empiric antibiotics with meropenem, was effective against the isolated *B. gladioli*, the patient ultimately succumbed multi-organ failure.

Conclusion

We report a case of VAP caused by the rare organism *Burkholderia gladioli*, leading to rapid renal deterioration and multi-organ failure. Despite prompt recognition and aggressive treatment, the infection proved fatal. This case highlights the challenges posed by *B. gladioli* infections and the need for greater understanding of its pathogenesis to improve patient outcomes.

病例報告 113_C162

未溶解的清腸劑所造成的急性腐蝕性傷害

Acute Corrosive Injury Caused by Ingestion of Undissolved SPMC Powder for bowel preparation

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Introduction

Bowklean® Powder (Genovate, Taiwan) is a sodium picosulfate/magnesium citrate (SPMC) preparation widely used for bowel cleansing due to its dual laxative effects. To prevent chemical burns in the esophagus and stomach, it is vital to fully dissolve the powder in water before consumption.

This report presents a rare case of acute esophageal and gastric corrosive injury resulting from the direct ingestion of Bowklean® dry powder.

Case Report

A 72-year-old man arrived at the emergency department (ED) with chest pain and epigastric discomfort after ingesting Bowklean® powder undissolved prior to a colonoscopy. He experienced an immediate burning sensation. On examination, he was stable but exhibited epigastric tenderness. Cardiac and pulmonary conditions were ruled out, with initial hemoglobin at 14.0 g/dL and normal electrolyte and coagulation tests. Imaging studies showed no significant findings.

Twelve hours post-ingestion, an esophagogastroduodenoscopy (EGD) revealed significant mucosal damage: edematous and friable mucosa with hemorrhagic erosions coated in white membranes in the middle to lower esophagus, alongside shallow brown-black ulcers in the gastric body. Additionally, tiny hemorrhagic spots were noted in the second portion of the duodenum. Histopathology indicated infiltration of lymphocytes and neutrophils with fresh hemorrhage in the lamina propria, while H. pylori was absent. The patient was diagnosed with acute corrosive esophagitis and gastric ulcers. Treatment included fasting, intravenous proton pump inhibitors, and oral sucralfate. Following symptom improvement, he was discharged on a liquid diet without further complications.

Discussion

Sodium picosulfate/magnesium citrate (SPMC), like Bowklean, is a common oral bowel preparation. Mixing with water, picosulfate sodium creates highly acidic liquid through hydrogen ion release. When water interacts with magnesium oxide and citric acid, it forms corrosive magnesium citrate, potentially harming the esophageal and gastric mucosa.

When dealing with caustic esophageal injuries, the initial EGD is a crucial diagnostic tool. According to Zargar's classification, lower-grade injuries (grades 1-2a) are less likely to lead to strictures, while severe burns (grade 3b) can result in strictures in up to 80% of patients. Strictures typically peak around 8 weeks post-ingestion but can occur as early as 3 weeks. In a recent 2021 case, a patient experienced Grade 2b ulceration in the middle esophagus after accidentally ingesting SPMC powder, leading to subsequent stricture complications.

During the EGD, we observed Grade 2a esophageal erosions with white membranes and Grade 2a

gastric ulcers with brown-black discoloration. The patient received 48 hours of conservative treatment, including restricted enteral feeding and sucralfate with intravenous esomeprazole. After successfully transitioning to oral feeding, the patient completed the treatment course without any complications.

Conclusion

This case illustrates the significant risks associated with the direct ingestion of SPMC without proper dissolution. Enhanced awareness among healthcare providers and patients is crucial for avoiding similar incidents in the future.

病例報告 113_C163

糞便移植對於同時患有猛爆性困難梭狀桿菌感染及發炎性腸疾病人之治療效果：一例病例報告

A case report of fecal microbiota transplantation in a patient with concomitant fulminant *Clostridioides difficile* infection and inflammatory bowel disease

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Introduction

Fecal microbiota transplantation (FMT) is a recommended therapy for recurrent *Clostridioides difficile* infection (CDI) but lacks confirmed evidence for patients with concomitant fulminant CDI and acute severe ulcerative colitis (ASUC).

Case Report

A 27-year-old female without systemic diseases presented to the Emergency Department with progressive watery diarrhea for 2 weeks, accompanied by bloody stool, lower abdominal pain, and intermittent fever. Abdominal CT showed diffuse wall thickening from the transverse colon to the rectum, consistent with colitis. A stool PCR test confirmed toxigenic *Clostridioides difficile*, and oral vancomycin was initiated. Colonoscopy revealed multiple diffuse A1 ulcers with mild oozing from the ascending colon to the rectum, suggesting ulcerative colitis (UC).

Due to persistent bloody diarrhea and fever, treatment was switched to fidaxomicin, and FMT was performed for fulminant CDI. Follow-up colonoscopy showed improvement, and a stool PCR test for C. difficile was negative. Steroid therapy (methylprednisolone 40 mg/day), azathioprine, and mesalazine were started for UC treatment.

After beginning enteral feeding, the patient developed diffuse abdominal pain, and CT revealed bowel perforation with peritonitis. Laparoscopic exploration found a 1 mm perforation in the descending colon. Due to poor response to steroid treatment, cyclosporine infusion was given, and then her symptoms improved gradually since then. Given her steroid-refractory acute severe UC, vedolizumab was administered. She reported no recurrence of bloody stool or diarrhea, and her condition remains stable.

Discussion

Clostridioides difficile is a gram positive, anaerobic, spore forming bacteria, which affects the large colon primarily leading to colitis. Patients with inflammatory bowel disease (IBD) have higher risk of CDI. Vancomycin or fidaxomicin are currently the recommended first-line treatments for an initial episode of CDI. FMT is a recommended therapy for recurrent CDI according to treatment guidelines. In some studies, FMT may also be beneficial in patients with severe or fulminant CDI. Furthermore, in current studies, FMT is well-tolerated without severe adverse events in IBD with CDI.

Conclusion

Our case demonstrated that FMT may be beneficial for patients with ASUC combined with fulminant CDI. However, there is a need for more prospective studies to determine the best treatment strategy in managing ASUC and fulminant CDI concurrently.

病例報告 113_C164

牙源性細菌直接感染所導致的腦膿瘍

Brain Abscess Caused by Direct Infection from Odontogenic Bacteria

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Introduction

In Taiwan, the prevalence of brain abscesses is approximately 1.88 per 100,000, with peak incidence around 30 to 40 years. Most cases have predisposing factors such as HIV, immunosuppressive drug use, or prior neurosurgery. Brain abscesses arise from either hematogenous or contiguous spread, with the latter accounting for over half of cases. Contiguous spread is often linked to skin flora or adjacent infections (e.g., otitis media) caused by *Streptococcus* species, anaerobes, or gram-negative bacilli. Odontogenic brain abscesses, though rare, may arise from dental infections, with *Olsenella spp.* identified as a potential pathogen.

Case Report

A 40-year-old male with a history of betel nut, alcohol, and tobacco use for over 15 years presented with persistent tooth pain after treatment for tooth #27. Despite debridement and amoxicillin, his condition worsened, and he developed high fever, toothache, confusion, and neurological symptoms including dysarthria and strabismus. Brain imaging revealed a pyogenic abscess at the left skull base extending to the masseter and temporalis muscles. CSF analysis showed high protein, low glucose, and neutrophil-predominant pleocytosis. The patient was intubated and transferred to the ICU. MRI confirmed a brain abscess with bilateral ventricular empyema. Blood cultures grew *Olsenella phocaeensis*, and the patient was treated with Ceftriaxone, Vancomycin, and Metronidazole. After abscess drainage and continuous debridement, his fever subsided, and his neurological status improved. He was later stabilized with a V-P shunt and prepared for discharge.

Discussion

Odontogenic brain abscesses account for 2-5% of cases and are often linked to poor oral hygiene or dental infections. With advances in molecular techniques, more oral pathogens like *Olsenella* are being identified. The most common symptoms include neurological deficits, headache, and fever. Recent improvements in imaging, surgery, and antibiotics have reduced brain abscess mortality from 40% to 10%, and recovery rates have increased from 33% to 70%. This patient had a favorable recovery, though some language impairment persisted.

Conclusion

This case highlights the importance of considering odontogenic sources in brain abscesses and emphasizes the critical role of surgical intervention and antimicrobial therapy in management.

病例報告 113_C165

庫倫氏徵象以及胰臟囊腫破裂伴隨出血

Cullen sign and Hemorrhagic Pancreatic cyst rupture with hemoperitoneum

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Introduction

Cullen sign is the sign of abdominal wall hemorrhage and is generally associated with acute pancreatitis. The bruise on the umbilicus is Cullen's sign. The bruising shows as a blue discoloration and is an indication of retroperitoneal hemorrhage, or bleeding in the peritoneum. We reported a case of hemorrhagic pancreatic cyst rupture, with hemorrhage as the cause of the Cullen sign.

Case Report

A 48-year-old man who had acute left flank pain for one day was admitted to our emergency room. Patient did not have any underlying disease and his physical study and laboratory study showed normal finding. The patient had a history of consuming six cans of beer per day for more than 20 years. About 15 days ago, the patient experienced a sharp episode of left flank pain while carrying a heavy generator during farming, and has since been experiencing intermittent pain in the same area. His previous CT showed a pancreatic cyst with hemorrhage at tail portion. On the second day, we observed ecchymosis over the umbilicus. Anemia was also noted. PRBC transfusion was made. A repeat CT revealed a pancreatic cyst rupture with hemoperitoneum. Ascites tapping was made that the ascites was bloody with SAAG<1.1. It was concluded that the Cullen sign in this patient was caused by hemorrhagic pancreatic cyst rupture with hemoperitoneum.

Discussion

Cullen sign refers to periumbilical ecchymosis and is often associated with intra-abdominal hemorrhage, commonly seen in acute pancreatitis, ruptured ectopic pregnancy, or other intra-abdominal bleeding events. In this case, the Cullen sign developed in a 48-year-old man with a history of a pancreatic cyst located in the tail, which ruptured and led to hemoperitoneum. The patient experienced intermittent left flank pain and later developed anemia, requiring PRBC transfusion. Ecchymosis around the umbilicus was noted on the second day, indicative of the Cullen sign, which occurs due to tracking of blood along the fascial planes to the umbilicus. The bloody exudative ascites with SAAG <1.1 further confirmed the diagnosis of hemorrhagic pancreatic cyst rupture. This case shed a light for the role of Cullen sign as an important physical manifestation during daily practice for detecting serious intra-abdominal bleeding such as pancreatic cyst hemorrhagic rupture.

Conclusion

We reported a case of Cullen sign with hemorrhagic pancreatic cyst rupture with hemoperitoneum.

病例報告 113_C166

憂遁草(沙巴蛇草)治療肝細胞癌的意外效果

Clinacanthus nutans' unexpected effect in treating hepatocellular carcinoma

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Introduction

Clinacanthus nutans (C. nutans), a perennial herb from the Acanthaceae family, is traditionally used in Southern Asian countries for treating various ailments, including snake bites, skin infections, burns, dysentery, diabetes, viral infections, and cancers. We reported a case of old age patient who refused traditional treatment of hepatocellular carcinoma and opted for the Clinacathus nutans' medication with unexpected disappearance of the tumor.

Case Report

A 95-year-old woman was admitted with diarrhea, which prompted further investigation. Computed tomography (CT) revealed a 5 cm liver nodule in segment 6, alongside liver cirrhosis (Figure 1). The patient tested negative for hepatitis B and C, but her alpha-fetoprotein (AFP) levels were elevated at 389 μg/mL, raising suspicion of HCC. After being treated for infectious diarrhea and discharged, she was advised to undergo a three-phase CT scan for further evaluation of the liver nodule. However, due to her age, she chose not to pursue invasive treatments. Instead, she opted for an alternative remedy using CN. For two months, she consumed a blended mixture of 20 CN leaves with an apple in juice form daily (250 cc per day). Remarkably, upon follow-up, ultrasound imaging indicated resolution of the liver nodule, and AFP levels had normalized. A subsequent three-phase CT scan showed early-enhancement and early washed-out formation of the residual nodule that confirmed the characteristics of HCC.

Discussion

Research has demonstrated that C. nutans exhibits a range of biological activities, notably its anticancer effects. Extracts and phytochemicals from C. nutans have shown significant cytotoxicity against HepG2 human hepatoma cells, with methanol extracts displaying the strongest activity. In animal models, a 30% ethanol extract (CN30) significantly inhibited tumor growth and induced apoptosis by modulating the immune response. These findings highlight the potential of C. nutans as a promising therapeutic agent for liver cancer. However, no clinical studies in human have been conducted specifically on HCC.

Conclusion

To our knowledge, this is the first reported case of CN's potential effect on HCC. This case suggests that CN might offer a complementary approach to traditional treatments such as transarterial embolization (TAE) or radiofrequency ablation (RFA) in managing HCC. Further research and clinical trials are necessary to confirm the efficacy and safety of CN in cancer treatment. We hope

this report stimulates interest and prompts further investigation into CN's potential role in oncology.

病例報告 113_C167

罕見的成人迴腸腸套疊: 神經束膜瘤

A Rare Adult Ileal Intussusception: Perineurioma

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Introduction

Perineuriomas are benign neoplasms that are uncommon and primarily found in the subcutis, although they can also develop in soft tissues or intraneural locations. Perineuriomas are composed of neoplastic cells that are similar to those of the normal perineurium. Lazarus and Trombetta initially described these neoplasms in 1978. Only a few reports have documented perineuriomas in the gastrointestinal tract, with the majority of cases occurring in the stomach, colon, and jejunum. We present a soft-tissue ileal perineurioma case that was clinically evident as intestinal obstruction as a result of intussusception in the terminal ileum.

Case Report

A 64-year-old woman presented with intermittent abdominal pain that suddenly intensified, became acute and unbearable, and lasted for more than half an hour, leading her to the emergency room. This severe episode was accompanied by vomiting and significant abdominal distension. Computed tomography revealed ileal intussusception. Emergent surgery was performed, and the pathological report identified a rare ileal perineurioma as the lead point for the formation of intussusception. The patient underwent successful resection of the affected segment, and her postoperative recovery was uneventful.

Discussion

Perineuriomas are rare benign peripheral nerve sheath tumors predominantly found in the subcutis, with gastrointestinal involvement, especially the terminal ileum, being particularly uncommon. Perineurioma usually presents as an asymptomatic intramucosal lesion, which is incidentally found in imaging studies. Differential diagnoses include gastrointestinal stromal tumors, schwannoma and leiomyomas. Unlike in children, surgery is usually mandated in adult intussusception because neoplasm is usually the underlying cause.

Conclusion

We reported a rare adult case of ileal intussusception with perineurioma as the offending agent.

病例報告 113_C168

肌肉強直症，一種伴有多種內科疾病的肌肉罕見疾病：病例報告

Myotonia dystrophy , a rare muscular dystrophy associated with many medical diseases: case report

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Introduction

Myotonic dystrophy (DM) is a progressive, multisystem disease characterized by skeletal muscle weakness, myotonia, cardiac conduction abnormalities, cataracts, endocrine dysfunction, and other systemic involvements. The multisystemic nature of DM necessitates thorough and regular monitoring across multiple organ systems for early detection and management of complications.

Case Report

We present a 45-year-old female with a 10-year history of progressive bilateral lower limb weakness, which eventually spread to all four limbs. She also experienced myotonia, predominantly affecting the distal muscles of her hands. Electromyography revealed electrical myotonia and myopathic changes. Her past medical history included a left ovarian tumor, cervical intraepithelial neoplasia, bilateral cataracts, and multiple injuries due to traffic collisions. A strong family history of similar symptoms was noted, as her mother, uncles, aunts, and younger sister all exhibited similar clinical features. Genetic testing confirmed a complete mutation of the DMPK (dystrophia myotonica protein kinase) gene, leading to the diagnosis of myotonic dystrophy type 1 (DM1). Her hemoglobin A1c (HbA1c) was 7.9%, confirming a new diagnosis of diabetes mellitus. Initial electrocardiography (ECG) showed a normal sinus rhythm, but first-degree atrioventricular (AV) block was noted during follow-up one year later. Her cognitive function remained intact, as evidenced by a cognitive abilities screening instrument (CASI) score of 99 points. Thyroid hormone levels and other endocrine tests were within normal limits.

Discussion

Myotonic dystrophy (DM) is a hereditary muscle disorder affecting skeletal muscles and multiple other organ systems, including the cardiac, respiratory, endocrine, gastrointestinal, and central nervous systems. The disease presents in two major forms: myotonic dystrophy type 1 (DM1), caused by an expansion of cytosine-thymine-guanine (CTG) repeats in the DMPK gene, and myotonic dystrophy type 2 (DM2), which results from cytosine-cytosine-thymine-guanine (CCTG) repeat expansions in the CNBP gene. The severity of the disease correlates with the length of the repeat expansions, with longer expansions typically resulting in earlier onset and more pronounced systemic involvement.

Despite its well-defined genetic cause, DM can be challenging to diagnose in its early stage or when symptoms are mild. Systemic involvement, including cardiac arrhythmias, diabetes mellitus, and cataracts, may provide important diagnostic clues. Regular follow-up and systematic evaluations are crucial for monitoring disease progression and managing complications. Annual electrocardiography is recommended to detect arrhythmias, particularly AV block. Pulmonary

function, as measured by forced vital capacity (FVC), should be assessed annually. Annual screening for fasting blood glucose and HbA1c levels is essential due to the increased risk of diabetes mellitus. Thyroid function, particularly thyroid-stimulating hormone (TSH) levels, should be evaluated every 2-3 years. Additionally, ophthalmologic examinations are recommended to monitor for cataracts every 2 years.

Conclusion

Myotonic dystrophy is a multisystem disease affecting both skeletal muscles and other organ systems. Regular monitoring, including cardiac, endocrine, respiratory, and ophthalmologic evaluations, is essential for early detection of complications and for optimizing patient outcomes.

病例報告 113_C169

骨巨細胞腫瘤異時性肺部轉移：病例報告

Metachronous Lung Metastasis of Giant Cell Tumor of Bone: A Case Report

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Introduction

Giant cell tumor of bone (GCTB) is a locally aggressive primary bone neoplasm that accounts for 5% of primary skeletal tumors and has a rare tendency to metastasize. We report a case of metachronous lung metastasis of knee GCTB.

Case Report

A 42-year-old woman with history of left knee giant cell tumor underwent tumor excision at NTUH in December 2012 and December 2014. She also has asthma with regular follow-up. On December 28, 2022, CXR revealed a new RUL lung nodule, and subsequent chest CT showed several small nodules up to 1.5cm in the LUL, RUL, and RLL. Bilateral lung nodules persisted in chest CT three months later. Surgical intervention for tissue proof was suggested, and the patient underwent uniport VATS wedge resection of LUL, RUL, RLL, and radical left lymph node dissection on April 18, 2023. Operative findings revealed three grayish tumors with mild pleural retraction, measuring 1.7cm (LUL), 1.2cm (RUL), and 1.1cm (RLL) respectively. The postoperative condition was stable, and the pathological result showed that the lung nodules were metastatic malignant giant cell tumor of bone. No recurrence has been observed so far.

Discussion

Giant cell tumor of bone (GCTB) primarily affects skeletally mature individuals, peaking in the third and fourth decades of life with a slight female preponderance. GCTB is a locally aggressive primary bone neoplasm that accounts for 4-8% of all primary bone tumors with 18-50% local recurrence rate and 2-3% distant metastasis rate, mainly to the lungs.

Pulmonary metastases are histologically benign, and malignant transformation occurs in less than 1% of cases. For lung metastasis, appropriate surgical resection should be suffice. Chemotherapy and radiotherapy are commonly used, especially in inoperable cases. However, the efficacy of chemotherapy for GCTB has been limited, primarily due to variations in drug selection. Radiotherapy carries a risk of up to 5% of post-radiation sarcomas, which is a significant concern, particularly in young patients with indolent disease.

Denosumab is often used as an alternative to chemotherapy or radiotherapy for unresectable GCTB. A phase 2 trial of Denosumab in GCTB showed long-term safety profile and disease control. However, given the generally favorable prognosis of lung metastases in patients with GCTB, observation can be seen as a viable option. Denosumab is preferred initially, with surgery reserved for cases of progression.

Conclusion

In conclusion, lung metastases are not uncommon in GCTB. They occur synchronously with

primary lesions in fewer than 10% of cases, but more commonly develop metachronously. Therefore, careful periodic surveillance with chest CT after diagnosing GCTB is recommended, especially for patients younger than 30 years old. Typically, lung metastases are multiple and bilateral, with a favorable prognosis. Once the histological diagnosis is confirmed, a watch-and-wait approach should be favored, considering the high rates of spontaneous regression or stable disease during long-term follow-up. Metastasectomy should be considered for single or few actively growing lesions. In addition, by reducing the disease progression, Denosumab can avoid surgery and prevent lung failure.

病例報告 113_C170

抗 MDA-5 抗體陽性的皮肌炎的臨床表現：個案系列報告

Clinical Course of Anti-MDA-5 Positive Dermatomyositis：Case Series

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Introduction

Anti-melanoma differentiation-associated gene 5 (MDA5) dermatomyositis is frequently associated with rapidly progressive interstitial lung disease (RP-ILD) and high mortality rates, highlighting the importance of early diagnosis. We present four cases of anti-MDA5 dermatomyositis with poor outcomes due to rapid-progressive interstitial lung disease.

Case Report

These four patients had a mean age of 58 years old (range 38 to 74 years old). Only one patient was a woman；one male patient had underlying diseases of hypertension and chronic kidney disease, while the other two male patients were healthy prior to admission. All of them presented with progressive exertional dyspnea and dry cough of variable duration (1 to 4 weeks), while two of them had non-specific skin rash over face and extremities. CXR showed variable degrees of increased infiltration and mixed alveolar pattern. All chest CT scans during admission revealed some degrees of basal fibrotic changes, consolidation and variant degrees of ground-glass opacity lesions. Serology panels of these four patients all showed strong positive of anti-MDA5 antibody in the serum, with some differing serological results. All of them developed acute respiratory failure within 1 month after symptoms onset (1 week to 1 month), and all ultimately required ECMO support. None of these four patients survived despite aggressive treatments, including pulse steroid therapy, IVIG, immunosuppressants, and even plasma exchange.

Discussion

Idiopathic inflammatory myopathies (IIMs) are a rare group of autoimmune disorders characterized by diverse clinical features including muscle inflammation, skin rashes, arthritis, and systemic manifestations. These conditions often coincide with specific antibodies, such as myositis-specific and myositis-associated antibodies, which aid in diagnosis and prognosis. Among these disorders, anti-MDA5 dermatomyositis stands out due to its association with rapidly progressive interstitial lung disease (RP-ILD), inflammatory arthritis, and distinct cutaneous symptoms. Initially identified in a Japanese cohort, anti-MDA5 dermatomyositis predominantly affects middle-aged women, although mortality rates are higher in men.

ILD is a common and severe complication in anti-MDA5 dermatomyositis, particularly RP-ILD, which significantly impacts mortality. Radiographic patterns such as organizing pneumonia (OP) and non-specific interstitial pneumonia (NSIP) are frequently observed, which impact disease management and prognosis. Diagnosis involves recognizing characteristic clinical features and confirming anti-MDA-5 antibodies by using various laboratory methods. High-resolution computed tomography (HRCT) and pulmonary function testing are crucial for screening and monitoring lung involvement, especially in the early stages.

Management typically involves aggressive immunosuppressive therapy, often combining Glucocorticoids with calcineurin inhibitors like Tacrolimus or Cyclosporine, and occasionally Cyclophosphamide. Recent studies also explore therapies such as Rituximab, JAK inhibitors like Tofacitinib, plasma exchange, and antifibrotic agents such as Pirfenidone, based on disease severity and individual responses.
Prognostic biomarkers like elevated ferritin and anti-Ro52 antibody correlate with disease severity and survival outcomes, guiding therapeutic decisions. Early intervention is critical given the rapid progression and high mortality associated with anti-MDA5 dermatomyositis RP-ILD, highlighting the importance of ongoing research and individualized treatment approaches.

Conclusion

In summary, anti-MDA5 dermatomyositis represents a challenging subset of IIMs characterized by severe systemic involvement, particularly RP-ILD. Timely identification, comprehensive evaluation, and aggressive treatments are necessary to improve patient outcomes.

病例報告 113_C171

內視鏡超音波導引之胃囊吻合術：治療胰臟假性囊腫的安全且有效的微創方法

Endoscopic Ultrasound-Guided Cystogastrostomy : A Safe and Effective Minimally Invasive Approach for Pancreatic Pseudocysts Management

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Introduction

Endoscopic ultrasound (EUS)-guided cystogastrostomy is a minimally invasive technique for managing pancreatic pseudocysts (PPCs), providing an alternative to surgery with fewer complications. By creating a direct communication between the pseudocyst and gastric lumen, EUS enables internal drainage without incisions. This method is highly effective, making it a safer and more efficient approach compared to traditional surgery.

Case Report

A 60-year-old male, previously diagnosed with an 8 cm pancreatic pseudocyst 10 months ago, presented to our hospital with persistent and progressive epigastric pain. According to past medical records, he experienced recurrent epigastric pain and was admitted twice due to pancreatitis since the initial diagnosis, with regular follow-up via abdominal sonography at our GI outpatient department. Upon ER evaluation, abdominal computed tomography revealed that the pseudocyst progressed in size, with features of walled-off necrosis. Following admission, the cystogastrostomy was performed with a 10 mm × 10 mm lumen-apposing metal stent. Immediately after stent deployment, a copious amount of turbid yellowish fluid drained from the cyst. The stent was positioned well along the posterior wall of the stomach. The patient was discharged 2 days later, and the metal stent was successfully removed 21 days post-procedure, with no recurrence of symptoms.

Discussion

A pancreatic pseudocyst (PPC) is a localized fluid collection rich in amylase, typically forming near the pancreas due to inflammation or injury. While most PPCs are asymptomatic and resolve spontaneously, treatment is necessary when they persist and cause symptoms such as abdominal pain, infections, or compression of nearby structures. Traditionally, surgical intervention has been the primary treatment, but it carries significant risks, including complications and mortality.

Endoscopic drainage provides a minimally invasive alternative, utilizing either a trans-papillary or trans-mural approach. The trans-mural method involves placing a stent to drain cyst fluid into the gastric lumen (cystogastrostomy) or duodenal lumen (cystoduodenostomy). This can be done “semi-blind” or guided by endoscopic ultrasound (EUS), which reduces the risk of damaging nearby vessels. The choice of drainage technique depends on the fluid collection type; for simple pseudocysts, a single 10-15-French plastic stent may suffice, while infected collections often require multiple stents or lumen-apposing metal stents. Current management algorithms favor EUS-guided drainage, which has higher technical success rates and fewer severe complications compared to percutaneous or unguided endoscopic drainage and surgical options.

Conclusion

EUS-guided drainage is an effective treatment for pancreatic pseudocysts, achieving successful outcomes in most patients. The findings of this case indicate that EUS-guided cystogastrostomy with a lumen-apposing metal stent is both safe and effective for managing pancreatic pseudocysts.

病例報告 113_C172

腹膜透析液中的蘑菇狀懸浮物-多病原菌引起的腹膜透析相關性腹膜炎

A Mushroom-like Floating Mass in Cloudy Peritoneal Dialysate: Peritoneal Dialysis-Associated Peritonitis with Multiple Pathogens

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Case Report

We report a rare case of a 71-year-old female patient with a history of type 2 diabetes mellitus and diabetic nephropathy, who has been undergoing peritoneal dialysis for four years. The patient was admitted to the chest ward for pneumonia and received non-invasive ventilation therapy for hypercapnic respiratory failure. During her hospitalization, she continued with continuous ambulatory peritoneal dialysis.

One night, the patient experienced acute diffuse abdominal pain, drowsiness, hypotension, and shortness of breath. Notably, a mushroom-like floating mass was observed in her peritoneal dialysate, which had become cloudy. Laboratory results revealed a white blood cell count (WBC) of 11,420/μL, with 9.2% band forms, lactate levels of 3.2 mmol/L and alanine aminotransferase (ALT) of 13 U/L. Dialysate analysis indicated a WBC of 4,590/μL, with 95% neutrophils and 30/μL red blood cells.

A CT scan of the abdomen was performed to rule out acute abdominal conditions, revealing mild pneumoperitoneum. A general surgeon was consulted for suspected hollow organ perforation, but surgery was ultimately refused by the family due to the patient's multiple comorbidities and the high risk of mortality.

Two days later, dialysate culture identified *Enterococcus faecium* and *Candida glabrata*. Despite antibiotic treatment, subsequent cultures revealed *Acinetobacter baumannii* and *Escherichia coli*. Unfortunately, the patient succumbed to sepsis after two weeks of treatment.

Discussion

The presence of a specific mass in peritoneal dialysate is rare; fibrin material may occasionally be observed. In this case, the patient was in septic shock, making the choice of empirical antibiotics critical. However, the mushroom-like floating mass did not provide definitive information about the pathogens involved. A review of the literature through PubMed yielded no related findings regarding such objects in dialysate fluid. Thus, we present this unique case as a reference for clinical practice.

Conclusion

A mushroom-like floating mass or similar object in cloudy peritoneal dialysate may indicate severe and complicated peritoneal dialysis-associated peritonitis with multiple pathogens.

病例報告 113_C173

自體免疫疾病的治療經驗 - 病例報告

Clinical Experiences on Treating Autoimmune Diseases - Case Studies

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Introduction

To date, approximately 80 types of autoimmune diseases have been reported. Autoimmune diseases often have chronic clinical courses, with some being easier to diagnose while others are more difficult and thus lead to delayed detection.

Case Report

Over the past eight years (2016–2024), we have collected 27 cases of autoimmune diseases at our clinic. Diagnosis was made through careful history-taking, physical examination, biochemical data, X-rays, and abdominal ultrasound examinations, etc. In addition to routine laboratory tests such as complete blood count, lipid profiles, liver, renal, and thyroid functions, we also conducted immunological studies for diagnosing autoimmune diseases, including ANA, RA factor, Anti-TPO, TSHR-Ab, and AMA.

Discussion

Case Report: Successful treatment of a case of autoimmune hepatitis complicated with liver cirrhosis (Child-Pugh class B).

An 81-year-old woman complained of general weakness, poor appetite, abdominal fullness, and lower leg edema for four months. Her first visit was on July 7, 2022. Biochemical tests revealed: GOT 46, GPT 12; Total Bilirubin 1.8, Direct Bilirubin 1.24; Albumin 2.7; HBsAg (-); Anti-HCV (-); No history of alcohol consumption or long-term medication use; but ANA 640X (+) (Table 1).

An abdominal ultrasound showed massive ascites and liver cirrhosis (Fig. 1). Then, a clinical diagnosis of autoimmune hepatitis complicated by liver cirrhosis (Child-Pugh class B) was made. Treatment with albumin infusion, diuretics, and steroids was administered. Follow-up ultrasound on March 16, 2023 showed normal liver function, no cirrhotic changes in the liver, and no ascites (Fig. 2). She currently enjoys a normal life.

Among the 27 patients, there were 7 cases of autoimmune liver disease, 3 cases of both rheumatoid arthritis and SLE, 2 cases of Hashimoto's disease, 2 cases of autoimmune thrombocytopenia, and 2 cases of Sjögren's disease. There was 1 case each of Grave's disease, autoimmune bronchitis, autoimmune vasculitis, autoimmune myositis, and primary biliary cirrhosis (Table 2).

One case had a very high ANA titer of 5,200X (+) but showed no clinical symptoms.

Conclusion

The exact etiology of autoimmune diseases is unknown, but both hereditary and environmental factors likely play a role. Among our patients, autoimmune liver disease was the most common,

and it generally responded well to medication. Notably, one case of AILD with liver cirrhosis (Child-Pugh class B) showed significant clinical improvement.

Grave's disease is a B-cell-mediated autoimmune disorder, where autoantibodies bind to the TSH receptor and stimulate it, resulting in increased levels of T3 and T4, and decreased TSH. In Grave's disease, the anti-TSHR antibody titer is elevated in 95% of cases, and Anti-TPO antibody levels rise in about 70%.

Hashimoto's disease is an autoimmune disorder characterized by hypothyroid symptoms such as cold intolerance, weakness, pale skin, and lower leg edema (myxedema). Biochemical tests typically show decreased T3 or T4, elevated TSH, and increased Anti-TPO antibody.

Patients with primary biliary cirrhosis often experience weakness, poor appetite, anemia, jaundice, and itching. Laboratory tests commonly reveal elevated levels of GOT, GPT, Bilirubin, Alkaline Phosphatase, and GGT, along with low albumin and hemoglobin. Abdominal ultrasounds may show biliary tract sclerosis, liver cirrhosis, and splenomegaly. The treatment of choice is ursodeoxycholic acid (UDCA), with early treatment leading to a better prognosis.

病例報告 113_C174

支氣管動靜脈異常形成伴隨反覆大量咳血發生在一位支氣管擴張症病人

Bronchial arteriovenous malformation complicated with refractory massive hemoptysis in a bronchiectasis patient

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Introduction

Pulmonary arteriovenous malformation was defined as the communication between the pulmonary artery and pulmonary vein and over 70% of the disease was associated with hereditary hemorrhagic telangiectasia. Here, we reported a rare case of bronchial artery communicating with pulmonary vasculature complicated with refractory massive hemoptysis.

Case Report

A 58-year-old man has underlying diseases of polycystic kidney disease, end stage renal disease under regular hemodialysis, coronary artery disease, and hyperparathyroidism. He firstly came to our hospital in May of 2020 for intermittent hemoptysis for 1 week. The computed tomography (CT) showed mild bronchiectasis in the right upper lobe of lung. The hemoptysis resolved after taking tranexamic acid and discontinuing antiplatelet agent. Thereafter, the patient had several episodes of pulmonary infectious over right upper lung, including one episode of lung abscess. However, due to the poor compliance to medication, these episodes of infection were considered to be treated partially. The hemoptysis was on and off during this period.

Recurrent hemoptysis with massive amount over 300mL of blood was noted in the October of 2023. The patient received angiography with bronchial arterial embolization (BAE). The angiography showed vascular structure at distal right bronchial artery with early appearing venous structure and bronchial arteriovenous malformation (AVM) was diagnosed. The massive hemoptysis resolved after BAE, but recurred again in December of 2023. The patient received repeated angiography with BAE. Hemoptysis resolved after the secondary BAE.

In February of 2024, blood-tinged sputum recurred and gradually progressed in weeks. The patient presented to our emergent department again due to once massive hemoptysis with over 800mL of fresh blood. He was intubated due to acute hypoxemic respiratory failure. RUL lobectomy was performed. The pathology of resected lung discovered hemorrhagic area and some vessels with inflammation. RUL AVM fed by the right bronchial artery with communication to the pulmonary vessels was impressed. The patient was stable after operation and was discharged with no more hemoptysis recurred in the six-month follow-up after discharge.

Discussion

Hemoptysis was the expectoration of blood from the lung parenchyma or airways. Pulmonary arteriovenous malformation (PAVM) was defined as the communication between the pulmonary artery and pulmonary vein. Over 70% of the PAVM was associated with hereditary hemorrhagic telangiectasia. Bronchial arteriovenous malformation is a rare subset of PAVM, approximately 2-4% of all PAVM. The bronchial AVM is a communication between the bronchial artery and the

pulmonary vessels. Bronchial AVM rarely originated from congenital diseases. Instead, bronchiectasis was a considered etiology for the bronchial AVM. Bronchiectasis with recurrent airway inflammation promotes angiogenesis and bronchial artery dilatation.

Conclusion

We reported a rare case of bronchial artery communicating with pulmonary vasculature complicated with refractory massive hemoptysis in a bronchiectasis patient which was managed successfully with lung resection.

病例報告 113_C175

攜帶 LMNA 基因患者接受電燒後的症狀改善

Case report- a case with LMNA gene mutation and atrial fibrillation receiving catheter ablation with symptoms improvement

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Introduction

LMNA gene mutations were often related to dysrhythmias, including atrial fibrillation or atrioventricular block. We present a case of 39-year-old with a *LMNA* gene mutation who had conduction system disturbance and atrial fibrillation with associated clinical symptoms. After receiving catheter ablation, the clinical discomfortabilities disappeared.

Case Report

A 39-year-old previously healthy male complained of intermittent chest tightness with shortness of breath after running. The electrophysiology study revealed no arrhythmia but a high degree AV block in 2015. Pacemaker implantation was suggested, but the patient rejected it. However, the symptoms got worse 8 years later even during ambulation. Electrocardiogram showed atrial fibrillation. A 24-hour Holter displayed incessant atrial fibrillation. The echocardiography revealed no significant structural disorder with the left ventricular ejection fraction was 63.6%. Because the symptoms were correlated with atrial fibrillation, catheter ablation was arranged.

After the procedure, follow-up 24-hour ECG recorded sinus rhythm with multiple atrial and ventricular premature complexes. Although there were multiple long pause due to second-degree AV block with Mobitz type 1 AV block and 2:1 AV block with some escape junctional beats, the symptoms improved a lot.

As reviewing his pedigrees, many family members have pacemaker implantation. In suspicious of genetic disorder, molecular genetic analysis was done which showed a heterozygous mutation in LMNA gene: c.1560G>A(NM_170707.4) p.Trp520Ter (NP_733821.1), which contributes to atrial fibrillation and conductive disorder presented in this patient.

Discussion

The *LMNA* gene encodes structural proteins of the nuclear envelope[1]. Mutations can lead to disorders in multiple organs, including striated muscle and vascular tissue, known as laminopathies[2].

LMNA cardiomyopathy include arrhythmia or dilated cardiomyopathy. Atrial fibrillation is frequently observed in LMNA cardiomyopathy and may precede an eventual atrial standstill. Early changes in atrial contractile strain are distinctive markers of intrinsic atrial myopathy, often preceding left atrial dilatation, left ventricular dysfunction, or the onset of atrial fibrillation.

Early-onset atrial fibrillation and progressive conduction system disease have a full penetration with *LMNA* gene mutations[3] suggesting the potential linkage between them.

Our patient presented with atrial fibrillation and conduction system disturbance. The echocardiogram showed a fair left ventricular ejection fraction with a fair left ventricle size. Symptoms significantly improved after catheter ablation. A nonsense mutation was noted in molecular genetic analysis and favorable risk was attributed based on previous study.

Conclusion

LMNA gene may be related with atrial fibrillation and conduction system disturbance. We reported a case with *LMNA* gene represented with chest tightness majorly coming from atrial fibrillation and improved after catheter ablation.

病例報告 113_C176

成功對 Pembrolizumab 產生嚴重過敏性休克的肺癌病患進行減敏治療

Successful Desensitization of Pembrolizumab-Induced Anaphylactic shock in a Lung Cancer Patient

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Introduction

Pembrolizumab, an anti-PD-1 monoclonal antibody, is associated with risks of infusion reactions and anaphylaxis. We reported a novel 16-step protocol specifically for a lung cancer patient, emphasizing saline restriction to minimize fluid overload. This approach combines minimal solvent use and effective management of severe hypersensitivity, allowing for the safe continuation of therapy.

Case Report

A 50-year-old man was diagnosed with advanced stage IVb pleomorphic carcinoma of the lung (cT4N3M1c). Lymph node biopsy revealed a PD-L1 expression of 98%. The treatment plan included a combination of pembrolizumab, cisplatin, and paclitaxel. However, during the administration of his second dose of pembrolizumab, the patient experienced an anaphylactic reaction characterized by tachycardia, shortness of breath, flushing, and profound shock.

The allergist was consulted for pembrolizumab desensitization therapy. Given the patient's compromised pulmonary function and concerns about fluid overload, we employed a 16-step desensitization protocol, targeting a dose of 200 mg of pembrolizumab, with water restriction in place. Pembrolizumab was diluted into 5 ml, 10 ml, 20 ml, and 40 ml normal saline solutions respectively to prepare solutions A (0.005 mg/ml), B (0.05 mg/ml), C (0.5 mg/ml), and D (5 mg/ml). The protocol commenced with Solution A at a dosage of 0.00125 mg administered over 15 minutes, with a subsequent doubling of the dose at each step. At step 15, involving Solution D and a 50 mg administered dose (cumulative dose of 97.36 mg), the patient developed urticaria on the neck and right eyelid angioedema. In response, antihistamine was given and the protocol reverted to step 13 of Solution D. The infusion rate was then maintained, allowing the desensitization to be completed without further anaphylaxis.

Discussion

This case represents the first reported instance of using a 16-step pembrolizumab desensitization protocol, distinguished by minimal saline use. Our protocol involved the infusion of a total of 75 ml of saline, significantly lower than the 200 ml used in previous therapies. Pembrolizumab maintains stability at an infusion concentration of 1–10 mg/ml and can be stored at room temperature for up to 6 hours. We prepared the drug at a concentration of 5 mg/ml, optimizing solvent use while meeting the minimum pump rate requirements. Although a minor allergic reaction occurred at step 14, the total therapy duration was able to controlled in approximately 5 hours. The patient is scheduled to undergo subsequent pembrolizumab desensitization courses following this protocol.

Conclusion

We have developed a 16-step desensitization protocol with water restriction for managing pembrolizumab-induced anaphylaxis in lung cancer patients.

病例報告 113_C177

反覆沙門氏菌菌血症與抗生素抗藥性之個案報告

Recurrent Salmonella bacteremia and antibiotic resistance: a case report

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Introduction

Salmonella, a genus of Gram-negative bacteria belonging to the family Enterobacteriaceae, comprises two main species: *Salmonella enterica* and *Salmonella bongori*. Within *S. enterica*, there are six subspecies, with *S. enterica* subsp. *enterica* being the most predominant, accounting for more than half of the species. Notably, *S. enterica* subsp. *enterica* includes serovar Enteritidis (commonly referred to as *S.* Enteritidis) and serovar Typhimurium (commonly referred to as *S.* Typhimurium), which are frequently encountered.

Clinical presentation of *Salmonella* infection commonly involves enterocolitis, which tends to be self-limited. However, severe cases may progress to invasive disease, with bloodstream infection occurring in approximately 6% of patients. Notably, mycotic aneurysm is a significant concern due to their high mortality rates. Other manifestations of invasive disease include focal abscesses and meningitis.

We present a case of recurrent *Samonella* bacteremia, detailing its antibiotic resistance and whole genome sequencing.

Case Report

A 73-year-old woman with history of hypertension. She was diagnosed with AML followed by induction chemotherapy and consolidation chemotherapy. Bone marrow transplant had been done 3 months before first episode of *Salmonella* bacteremia. However, another *Salmonella* bacteremia episode appear 1 year after first episode. About culture evidence other than blood, stool culture with *Salmonella* was noted in first episode, and urine culture with *Salmonella* was noted in second episode. There are some differences of minimum inhibitory concentrations (MICs) for antibiotics in 2 samples.

Discussion

To analyze resistance patterns and genetic sequences, the samples underwent whole genome sequencing. These two samples were identified as *Salmonella* Enteritidis. Each sequence was analyzed using PubMLST.com, a website dedicated to public databases for molecular typing and microbial genome diversity. The allelic profiles of seven housekeeping genes (aroC, dnaN, hemD, hisD, purE, sucA, and thrA) were constructed. There were no discernible differences between the two samples, and both sequences were typed as ST11. It is possible that antibiotic resistance may be linked to allelic profiles beyond those mentioned above.

Conclusion

We present a case of recurrent *Salmonella* bacteremia. These two samples were identified as *Salmonella* Enteritidis, and some differences in antibiotic resistance was noted. Despite analyzing

seven housekeeping genes (aroC, dnaN, hemD, hisD, purE, sucA, and thrA), no significant differences were noted. Both sequences were typed as ST11 in blood samples. Therefore, further investigations into other alleles may be warranted in the future.

病例報告 113_C178

異體造血幹細胞移植後弓形蟲病：病例研究與診斷挑戰

Toxoplasmosis Post-Allogeneic Hematopoietic Stem Cell Transplantation: A Case Study and Diagnostic Challenges

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Introduction

Toxoplasmosis is a rare but potentially fatal complication in patients undergoing allogeneic hematopoietic stem cell transplantation (allo-HSCT). Common symptoms include fever, pneumonia, encephalitis, and retinochoroiditis. Due to severe immunosuppression following transplantation, distinguishing toxoplasmosis from other infectious complications and initiating timely treatment can be challenging.

Case Report

A 57-year-old man developed toxoplasmosis 360 days after undergoing allo-HSCT. The patient had a history of exposure to pet cats, which served as a significant risk factor. On day 174 post-transplant, he experienced symptoms including low-grade fever, left-hand tremors, limb weakness, and bilateral retinochoroiditis. Concurrently, he presented with grade 2 skin acute graft-versus-host disease (aGVHD), and infections by cytomegalovirus (CMV) and Salmonella.

Initial imaging, including a brain CT performed in December 2022, showed no abnormalities. Due to concerns about myotonic syndrome, EEGs were performed between January and April 2023, which revealed persistent slow waves indicating systemic brain dysfunction. Tremors showed improvement with antiepileptic treatment; however, gait instability continued to deteriorate.

A repeat MRI of the brain on April 29, 2023, revealed multiple calcifications and contrast-enhancing lesions in the cerebral hemispheres and brainstem. These findings initially suggested drug-induced toxic encephalitis. However, worsening neurological symptoms and a fever spike of 39.0°C on day 356 post-transplant warranted further investigation.

Based on the history of pet exposure, neurological symptoms, and imaging results, a diagnosis of toxoplasmosis was considered. Serological testing revealed positive Toxoplasma IgG and IgM, supporting the diagnosis. Following consultation with an infectious disease specialist, treatment with trimethoprim/sulfamethoxazole (TMP/SMX) and pyrimethamine began on May 27, 2023.

Discussion

The primary risk factor in this case was the patient's contact with pet cats, combined with the immunosuppression from allo-HSCT. The clinical presentation of toxoplasmosis often overlaps with other infectious conditions, delaying diagnosis. In this case, collaboration with a multidisciplinary team enabled accurate identification and treatment of toxoplasmosis, resulting in symptom management and stabilization.

Conclusion

Diagnosing toxoplasmosis in allo-HSCT patients is complex due to the nonspecific nature of its

symptoms and their similarity to other infections. Early recognition is crucial, particularly in patients presenting with intermittent fever and central nervous system symptoms. A high index of suspicion, especially in the presence of risk factors such as pet exposure, is key to improving outcomes for these patients.

病例報告 113_C179

長期留置人工血管病患之微桿菌菌血症病例報告

Case of *Microbacterium* species Bacteremia in a Patient with Long-term Indwelling Chemo Port

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Introduction

Microbacterium spp. is a genus that has been increasingly identified in clinical settings, though human infections remain rare. These bacteria are commonly found in environmental sources such as soil and water, but infections in humans are typically linked to immunocompromised individuals. Most reported cases occur in patients with long-term indwelling devices, such as chemo ports, which act as entry points for these pathogens. Diagnostic advancements such as MALDI-TOF MS and 16S rRNA sequencing, have greatly improved the accuracy of identifying rare organisms like *Microbacterium* spp., allowing for better understanding of their clinical relevance.

Case Report

A 65-year-old male with a history of esophageal cancer, undergoing long-term steroid therapy for psoriasis, presented to the hospital with symptoms of fever and chills. Blood cultures from both the chemo port and peripheral blood samples yielded gram-positive bacilli, which were later identified as *Microbacterium paraoxydans* using MALDI-TOF MS. After confirming chemo port-related bloodstream infection, the chemo port was removed, and a 14-day course of vancomycin followed by levofloxacin was administered. The patient recovered fully without additional complications.

Discussion

M. paraoxydans is a rare pathogen in human infections, with only a handful of cases documented in the literature. Most of these infections occur in patients who are immunocompromised or have long-term indwelling devices, such as central lines or chemo ports. Although the clinical data on *Microbacterium* spp. remain limited, the infection is typically responsive to antibiotics such as vancomycin, gentamicin, and levofloxacin. In this case, timely removal of the infected chemo port and appropriate antibiotic therapy resulted in a favorable outcome. This case highlights the importance of considering rare infections in patients with compromised immune systems and indwelling medical devices, as early intervention is crucial to prevent complications.

Conclusion

This case underscores the need for heightened awareness of infections caused by *Microbacterium* spp., particularly in patients with long-term indwelling devices. Due to the scarcity of data on treatment protocols and resistance patterns, further research is essential to optimize diagnostic approaches and therapeutic strategies. More studies are required to better understand the pathogenicity, clinical presentation, and effective management of *Microbacterium* spp. infections in humans, which will contribute to improved patient outcomes and healthcare practices.

病例報告 113_C180

一例反覆性無色桿菌菌血症的基因體分析

A case report of recurrent *Achromobacter* bloodstream infection patient: genetic analysis of an unusual pathogen in Taiwan

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Introduction

The genus *Achromobacter*, with over 20 recognized species, is an emerging opportunistic pathogen, which can cause pneumonia, bacteremia, and infections in various sites. Vulnerable populations include patients with medical devices, underlying conditions, such as malignancies, and healthcare exposures. This study investigates the genetic and phenotypic changes among *Achromobacter* isolates from a patient with recurrent *Achromobacter* bloodstream infections (BSIs).

Case Report

We reported a 61-year-old woman with a history of ovarian cancer. The patient developed fever during chemotherapy infusion via a chemo port, and *Achromobacter denitrificans* (isolate 1) was detected in the blood culture by matrix assisted laser desorption ionization-time of flight mass spectrometry (MALDI-TOF MS). One month later, the patient again experienced chills during chemotherapy infusion, with *Achromobacter* species (isolate 2) detected in the blood culture by MALDI-TOF MS. The chemo port was subsequently removed. Whole-genome sequencing (WGS) was performed on the *Achromobacter* isolates. Comparison of species identification by MALDI-TOF MS and WGS revealed discrepancy, and WGS identified a novel *Achromobacter* species. Isolates 1 and 2 exhibited a maximum average nucleotide identity (ANI) of 86% against all known *Achromobacter* type strains, falling below the 95% species demarcation threshold. Besides, analysis of non-synonymous single nucleotide polymorphisms (SNPs) between isolates revealed missense mutations in *rapA*. Furthermore, isolate 2 exhibited increased minimum inhibitory concentration (MICs) for cefepime, ceftriaxone, ciprofloxacin, and tigecycline compared to isolate 1. This change coincided with the T76S mutation in *rapA*, suggesting a potential role for this mutation in the development of resistance.

Discussion

The characteristics of our BSI patient aligned with prior reports, showing a predominance of *Achromobacter* infections in individuals with underlying malignancies and central venous catheter implants. However, traditional identification methods, such as MALDI-TOF MS, may lack the resolution to differentiate closely related species. Genome sequencing reveals within-host pathogen evolution, where longitudinal isolate SNPs identified missense mutations in genes like *rapA*. *rapA* is an RNA polymerase-associated protein. *rapA* gene down-regulated *yhcQ*, which encodes a putative multidrug resistance pump. Mutants in the *rapA* gene exhibited greater sensitivity to beta-lactam, norfloxacin, chloramphenicol, and gentamicin. Genomic adaptations, including SNPs, suggest within-host evolution contributing to resistance and persistence within

the host.

Conclusion

Our study demonstrates the utility of WGS for accurate species identification, uncovering novel lineage, and tracking within-host evolution of *Achromobacter* species in recurrent infections. The identification of the correlation between SNPs and resistance changes provides valuable insights into the molecular mechanisms underlying antibiotic resistance in this emerging pathogen. These findings underscore the importance of WGS-based surveillance to monitor the spread of resistant strains and inform the development of effective strategies to combat *Achromobacter* infections.

病例報告 113_C181

以高劑量抗蛇毒血清治療赤尾青竹絲咬傷後罕見凝血病變：一病例報告

High-Dose Antivenom Therapy for an Unusual Coagulopathy Following Trimeresurus stejnegeri stejnegeri Bite Envenoming: A Case Report

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Introduction

Trimeresurus stejnegeri stejnegeri is one of the major venomous snakes in Taiwan, causing more than half of snakebite envenomings each year. Its bite usually leads to localized symptoms such as swelling, pain, blistering, and wound necrosis; standard doses of antivenom (1-2 vials) are usually sufficient to treat such envenomings. Systemic effects, like coagulopathy, are uncommon but potentially life-threatening. Although antivenom is the mainstay of treatment for snakebite envenomings, its effectiveness in treating coagulopathy, such as seriously low fibrinogen levels induced by *T. s. stejnegeri* bite envenoming, is still uncertain. In our previous observations, standard doses of antivenom have been of little effective in treating this type of coagulopathy. This report describes how high doses of antivenom were used to treat an unusual and severe blood clotting problem caused by a *T. s. stejnegeri* bite.

Case Report

A 50-year-old previously healthy female was bitten on her right middle finger by a *T. s. stejnegeri*. Two hours later, her whole hand became severely swollen, with the swelling extending 10 cm above the wrist. Laboratory tests showed progressively declining of fibrinogen levels (lowest at 87.4 mg/dL), a prolonged prothrombin time (INR: 1.29), and high levels of fibrin breakdown products (FDPs, 14.2 μg/mL), confirming the presence of defibrinogenation and coagulopathy. The patient was given 4 vials of antivenom about three hours after the bite along with cryoprecipitate transfusion. Although the swelling and pain improved, the coagulopathy persisted, requiring another 4 vials of antivenom and blood product transfusions. In total, the patient received 8 vials of antivenom and 20 units of cryoprecipitate. Her abnormalities in many coagulation-related markers returned to normal on the fourth day after the bite, and she was discharged after 8 days hospitalization without further complications.

Discussion

T. s. stejnegeri venom contains enzymes that act like thrombin and other proteins that can lead to low fibrinogen levels and deplete clotting factors. This case demonstrated that the usual doses of antivenom may not be enough to treat severe coagulopathy caused by *T. s. stejnegeri* bites. In our previous observations, even though antivenom was given multiple times, large amounts of blood products were still needed to correct the abnormal clotting function. We therefore, assumed that the coagulopathy related to *T. s. stejnegeri* bite required higher dose of antivenom. The antivenom's ability to neutralize the coagulopathy seemed limited, indicating that more frequent checks of clotting levels may be necessary for optimal treatment. A more aggressive antivenom regimen could potentially reduce the need for blood products and lower the associated

transfusion burden.

Conclusion

This case shows the difficulties in treating severe coagulopathy following a *T. s. stejnegeri* bite and emphasizes the potential need for higher dose of antivenom. The amount of antivenom needed was much more than usual, and many blood products were required for the patient to recover. Early use of higher doses of antivenom and more frequent checks of blood clotting markers might be necessary in cases of severe coagulopathy caused by *T. s. stejnegeri* envenomation.