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Case Report of Type A Aortic Dissection Complicated with Bilateral Extensive Ppulmonary Hemorrhage and Pulmonary Artery Intramural Hematoma

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Abstract

Aortic dissection with extensive involvement of other branching vessels is a serious concern and has a grave prognosis. Although its occurrence is rare, vessel dissection can extend from the ascending aorta to the pulmonary artery because they share a common vessel adventitia and this can result in pulmonary artery intramural hematoma or pulmonary artery dissection, with blood leakage into the pulmonary alveoli. Herein, we report a case of Stanford type A aortic dissection with suspected involvement of the pulmonary artery, which contributed to an extensive pulmonary hemorrhage as revealed by computed tomography. Furthermore, left ventriculography revealed subsequent enhancement of the bilateral pulmonary arteries, thus yielding additional support to the notion that this complication can occur in patients with type A aortic dissection. In addition to a dissected vessel flap and intramural hematoma over the pulmonary artery, extensive pulmonary hemorrhage that results from unstoppable blood leakage into vulnerable lung alveoli may be an indirect but significant clue for potential concomitant pulmonary artery intramural hematoma or dissection.

Key Words: aortic dissection; left ventriculography; pulmonary artery intramural hematoma; pulmonary hemorrhage

Background

Aortic dissection with extensive involvement of other branching vessels, regardless of whether they are type A or B, is rare and often leads to coronary, cerebral, visceral or extremity malperfusion resulting in end organ failure[1,2]. Although aortic dissection involving the pulmonary artery is less common than dissection involving other main branching vessels, its prognosis is worse.

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Concomitant aortic artery and pulmonary artery dissection is typically identified when a dissected vessel flap or a periarterial hematoma is detected in computed tomography images[3]; such images can also be used to determine the dissected entry site and the extent of dissection[4]. However, an aortic dissection can extend further into distal pulmonary branches and even the alveoli, resulting in pulmonary hemorrhage[3]. Therefore, clinicians should remain alert to the possibility of extensive pulmonary hemorrhage in cases of suspected pulmonary artery intramural hematoma (PA-IMH) and dissection.

In addition to computed tomography ,echocardiography which is readily available, has been recommended for the timely identification of intimal flaps over the aorta and the pulmonary artery[5,6]. However, the dissected portion over the distal right and left branching pulmonary artery vessels may be difficult to identify in this manner. Aortography and angiography can also be used to evaluate dissections because then can reveal the passage of the contrast agent into the pulmonary arterial trees[7]. Blood leakage into the common arterial adventitial, leading to substantial pulmonary hemorrhage, is also an indirect but key clue for diagnosing an aortic dissection.

Aortic dissection with concomitant pulmonary artery dissection is a rare but fatal condition with a high incidence of mortality. Although the optimal surgical approach for treating an isolated pulmonary artery dissection remains subject to debate and conclusive guidelines for treatment are lacking[4,6,8], emergent surgical repair of an aortic dissection with concomitant pulmonary artery dissection has been reported to be lifesaving.

Case presentation

A 52-year-old woman with a history of hypertension, suddenly collapsed. Bystander cardiopulmonary resuscitation was performed, and direct defibrillation was administered once for a shockable rhythm by using an automated external defibrillator. Point-of-Care Ultrasound at the emergent department revealed a negative result of peritoneal etiology; bowel gas obstructed imaging of the abdominal aorta. The initial rhythm at the emergency department was pulseless electrical activity. Cardiopulmonary cerebral resuscitation was performed for 1 hour, with three episodes of ventricular fibrillation after cardioconversion; subsequently extracorporeal membrane oxygenation support (ECMO) was arranged.

The laboratory data upon admission revealed an extreme hemoconcentration with elevated hemoglobin (20.7 g/dL) and hematocrit (62.3%) levels but a decreased fibrinogen level (<80 mg/dL), which indicated a massive hemorrhage. Coagulopathy was also detected, with a decreased platelet concentration of 81 × 103/μL, prolonged activated partial thromboplastin time (115.1 seconds), and prolonged prothrombin time (18.8 seconds).

The patient's artery blood gas values indicated respiratory acidosis with CO_2 retention and decreased oxygen levels (pH = 7.057, PCO_2 = 80.8 mmHg, PO_2 = 46.0 mmHg, HCO_3 = 23.0 mmol/L, BE-b = -9.7 mmol/L). Biochemistry revealed inadequate tissue perfusion (lactate = 175.7 mg/dL) and elevated cardiac enzyme levels (troponin I = 19.5582 ng/mL, creatinine phosphokinase = 3406 U/L). Other abnormal findings included impaired hepatic (AST = 584 U/L, ALT = 484 U/L) and renal function (creatinine = 1.10 mg/dL, eGFR = 52.16). The patient's electrolyte levels, including those of sodium and potassium, were within the normal range (Na = 145 mmol/L, K = 4.9 mmol/L).

Although the electrocardiogram did not reveal ST segment elevation, emergent coronary angiography, with intra-aortic Balloon Pump insertion, was performed because acute coronary syndrome with ventricular fibrillation rhythm and cardiogenic shock was suspected. The angiography revealed

diffuse atherosclerosis with 80% stenotic lesions over the right coronary and left anterior descending arteries.

On the basis of left ventriculography, which was performed to evaluate left ventricular ejection fraction, with subsequent enhancement of bilateral pulmonary artery trees, aortic dissection was suspected, (Figure 1). Aortic dissection was then verified through computed tomography, which revealed the following:

1. Stanford type A aortic dissection with abnormal air accumulation over the ascending aorta

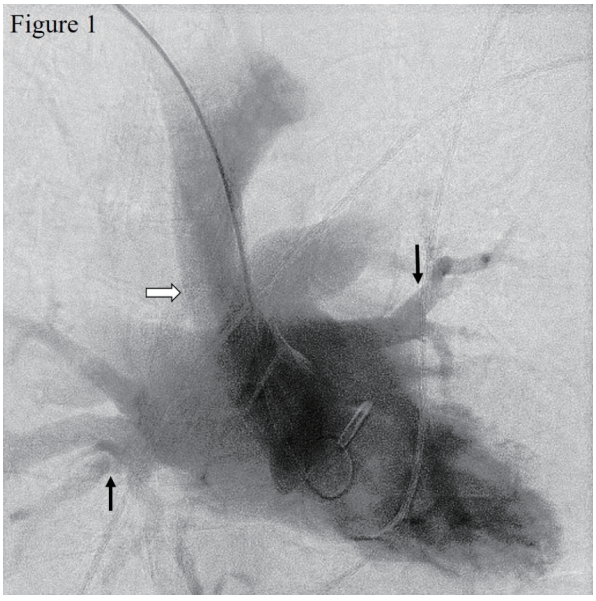

Figure 1. Left ventriculography
White arrow: Aortic dissection with dissected intimal flap
Black arrow: Bilateral pulmonary artery tree enhancement

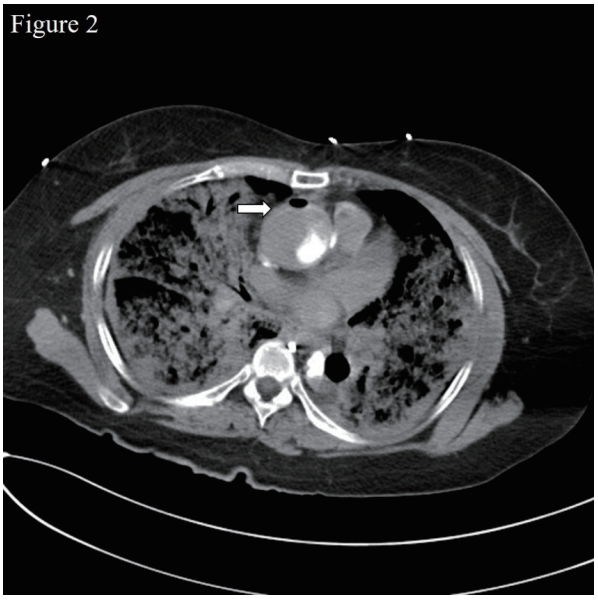

Figure 2. Computed tomography images
White arrow: Stanford type A aorta dissection with formation of large false lumen and abnormal air accumulation

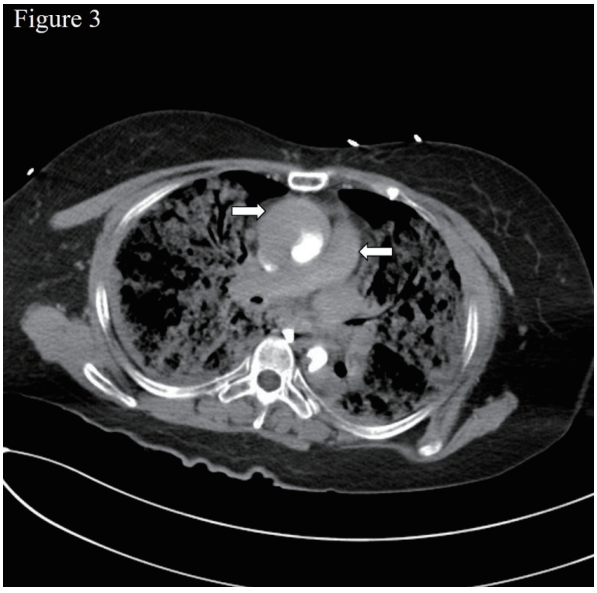

Figure 3. Computed tomography images
White arrow: Hyperdense attenuation surrounding the ascending aorta and pulmonary artery

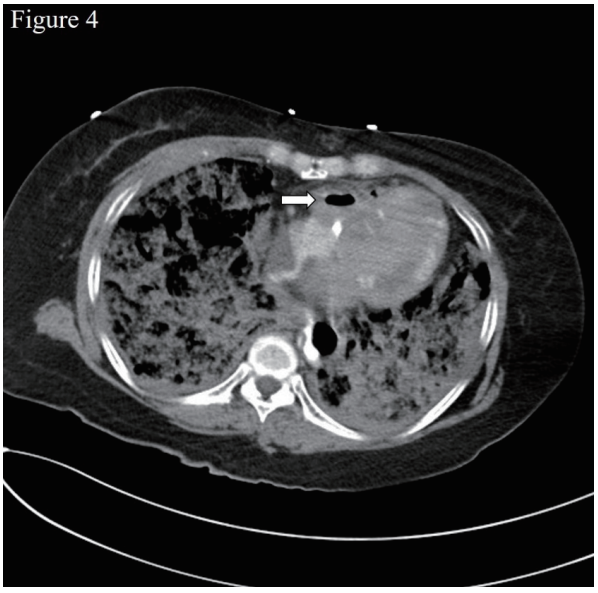

Figure 4. Computed tomography images
White arrow: Abnormal air accumulation over the right atrium and right ventricle

(Figure 2), with hyperdense attenuation surrounding the ascending aorta and pulmonary artery (Figure 3) and air accumulation over the right atrium and right ventricle (Figure 4);
2. Involvement of the brachiocephalic artery and the right and left common carotid arteries, and slow blood flow in the right renal artery
3. Marked brain swelling indicating acute ischemic-hypoxic encephalopathy;
4. Extensive bilateral lung opacity (Figures 2-4).

The computed tomography finding of hyperdense attenuation surrounding the ascending aorta and pulmonary artery and extensive bilateral lung opacities were notable; blood had leaked into the lung alveoli, following the path of least resistance. The patient died a few hours later due to hemorrhagic shock and profound cardiogenic despite ECMO support.

Discussion

In type A aortic dissection, the complication of concomitant or extended dissection along the pulmonary artery is rare, but can occur when blood leaks into the vessel adventitia shared by the ascending aorta and pulmonary artery trunk, causing an IMH[3,9,10]. Hodges et al. reported that an IMH of the aortic artery the can be identified by crescentic or circular vessel wall thickening (≥5 mm), with wall attenuation between 39 and 72 Hounsfield units on computed tomography images[11]; a PA-IMH can be similarly identified. Although extended dissection of the pulmonary artery has most frequently been reported with type A aortic dissection, a rare case of type B aortic dissection was also recorded[12].

A diagnosis of pulmonary artery dissection can be made when a dissected intimal flap in the pulmonary artery is identified through echocardiography or contrast-enhanced computed tomography angiography. When hyper-attenuated crescentic or circumferential thickening along the wall of the aorta or pulmonary artery is revealed on computed tomography, IMH can be established[13]. The cause of PA-IMH is presumed to be blood extravasation into the common aortopulmonary adventitia secondary to aortic wall rupture[3]. A review article by Sueyoshi et al. recommended that hemorrhages be classified as category 1 (IMH of the pulmonary artery or blood localized around the pulmonary artery), category 2 (IMH extending into the interlobular septa) or category 3 (IMH extending into the alveoli), with category 3 being the most severe and typically involving poor prognosis and a high risk of death[13].

The most common presentation of aortic dissections involving other aortic branching vessels is an IMH with a dissected pulmonary artery intimal flap[10,11,14,15]. Other rare cases have been reported where the aortic dissection was associated with a rupture extending through the patent ductus arteriosus[16] and the aortopulmonary artery fistula[17]. In another rare case, the mediastinal hematoma compressed the pulmonary arteries, mimicking a massive pulmonary embolus[18]. In addition, as in our case, cases involving pulmonary infiltration or hemorrhage have been reported[19,20]. In one unique case, aortic dissection with pulmonary infiltration mimicked coronavirus pneumonia[21]. These cases should alert clinicians to the possibility of concomitant pulmonary artery hematoma and dissection.

Certain genetic diseases such as Turner syndrome, Marfan syndrome and Ehlers-Danlos syndrome, or structural abnormalities, such as a bicuspid aortic valve or aortic coarctation, may predispose a patient to having fragile blood vessel. The only conceivable risk factor for aortic dissection in our patient, however, was hypertension. Aortic atherosclerosis and arteritis are other possible causes in the case of the middle-aged woman. Despite small aortic aneurysm dissection potentially being related to an underlying autoimmune disease[22], in this case, the associated autoimmune biomarkers could not be collected in time.

Conclusions

Although cases of isolated pulmonary infiltration with hematomas around the ascending aorta indicating pulmonary artery dissection have been documented, no case of isolated pulmonary infiltration with extensive bilateral pulmonary hemorrhage has been reported. In this case, we observed no evidence of a dissected flap over the pulmonary artery. However, the extensive involvement of other vessels, including the brachiocephalic artery, right common carotid artery, and left common carotid artery, slow blood flow in the right renal artery, and the abnormal presentation with aortic IMH formation and hyperdense attenuation of the pulmonary artery gave the impression of an aortic dissection with pulmonary artery involvement extending to the lung alveoli. Moreover, computed tomography images revealed hyperdense attenuation surrounding the pulmonary artery and extensive bilateral pulmonary hemorrhage, which further led to the suspicion of concomitant pulmonary artery hematoma, potentially resulting in unstoppable blood leakage into vulnerable lung alveoli.

Mechanic cardiopulmonary cerebral resuscitation might also induce pulmonary hemorrhage. Left ventriculography revealed the contrast enhancement of the bilateral pulmonary arteries, which further increased the likelihood of this complication in our patient with type A aortic dissection. The abnormal air retained in the right ventricle, right atrium, and ascending aorta was inferred to have originated from the distal pulmonary artery and to have been induced by positive mechanical ventilation and cardiopulmonary cerebral resuscitation, providing another clue of aortic dissection with pulmonary artery involvement.

If only non-contrast-enhanced computed tomography is available, the presentation of PA-IMH and extensive pulmonary hemorrhage may be an indirect but significant indicator of aortic dissection with pulmonary artery involvement. Clinicians should thus be aware that computed tomography, before an angiography, may be necessary for assessing an uncertain cause of cardiac arrest to avoid delayed diagnosis and management.

List of abbreviations

PA-IMH: Pulmonary artery intramural hematoma
ECMO: Extracorporeal membrane oxygenation

Availability of data and materials

All relevant data supporting the conclusions of this article are included within the article.

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Ethics declarations

Ethics approval and consent to participate

The study was approved by Ethics committee of Shin Kong Wu Ho-Su Memorial Hospital, and the patient's family gave written consent to the study's description of the patient's case.

Consent for publication

Written informed consent for publication was obtained from the patient's family.

Competing interests

The authors declare that they have no competing interests.

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案例報告：Type A 主動脈剝離併發雙側廣泛性肺出血及肺動脈血管壁內血腫

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摘要

主動脈血管剝離擴及其他週邊重要分支血管的情況雖然並不常見，但預後卻十分不良。由於主動脈與肺動脈被包覆在同一層血管結締組織外膜，當發生主動脈血管剝離時，剝離的範圍可能擴及至肺動脈，導致肺動脈血管壁內血腫，甚至肺泡出血。我們於此文介紹一個少見的 type A 主動脈剝離案例，併發肺動脈血管壁內血腫與廣泛性雙側肺泡出血 (據電腦斷層影像分類為第 3 類)。後續左心室造影亦顯示不正常的雙側肺動脈顯影。因此，當發生主動脈血管剝離時，肺動脈血管壁內血腫與廣泛性肺泡出血是一個重要的線索，間接顯示了肺動脈血管的受損甚至剝離。