

Type A acute aortic dissection mimicking pulmonary embolism: intramural hematoma of the pulmonary arteries causing right heart pressure overload and hemoptysis

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ABSTRACT

Aim To report on an atypical complication of type A acute aortic dissection (TAAAD) mimicking pulmonary embolism with right heart pressure overload and hemoptysis due to intramural hematoma of the pulmonary arteries extending into the pulmonary interstitium.

Methods A 66-years old male presented with sudden chest pain, dyspnoea and nausea. Electrocardiogram (ECG), point of care ultrasound (POCUS) of the heart and tripple rule out computed tomography (CT) were performed to differentiate the cause of acute chest pain.

Results ECG showed no ischemia. POCUS of the heart showed aneurysmal ascending aorta (4,5 cm), suspected intimal flap, mild aortic regurgitation, minimal pericardial effusion, right heart dilatation with impaired function. CT confirmed TAAAD with extensive intramural hematoma around the central pulmonary arteries propagating to intrapulmonary branches. Additionally, there was dilatation of the right ventricle and right atrium indicative of right heart pressure overload along mild haemorrhagic pericardial effusion anterior to the right ventricle. No evidence of pulmonary embolism was found. Emergency surgery included the replacement of the supracoronary ascending aorta and aortic hemiarch.

Conclusion TAAAD is a rare but life-threatening cause of chest pain often mimicking other conditions such as pulmonary embolism. Pulmonary complications including intramural hematoma of the pulmonic trunk due to its shared adventitia with ascending aorta are rare but clinically significant, causing lumen reduction or occlusion of pulmonary arteries, leading to right heart pressure overload. Timely differentiation is critical as anticoagulation is contraindicated.

Keywords: aortic rupture, aortopulmonary adventitia, chest pain, intramural hematoma, pulmonary artery disease, thoracic aortic syndromes

INTRODUCTION

Acute aortic syndrome represents an emergency condition that includes aortic intramural hematoma, penetrating atherosclerotic ulcer and classic aortic dissection (1). Aortic dissection is characterized by tearing of tunica intima and blood extravasation into the tunica media, creating a dissection flap that divides the true lumen from a newly formed false lumen which can compromise blood flow to vital organs (2). Aortic dissection occurs at an estimated incidence of 6 cases per 100,000 individuals annually, with a mean patient age of 62±14 years and a higher prevalence in males (68.2%) (3,4).

The most prevalent pre-morbid risk factor for acute aortic dissection is hypertension, particularly hypertension resistant to medication. Other risk factors include pre-existing aortic dis-

eases or aortic valve diseases, family history of aortic diseases, history of cardiac surgery, cigarette smoking, direct blunt chest trauma and the use of intravenous drugs (e.g. cocaine and amphetamines) (5).

In type A acute aortic dissection mortality rate without treatment increases by 1% to 2% per hour within the first 24 to 48 hours, reaching nearly 50% by the end of the first week. With prompt surgical repair in-hospital mortality rate is around 15% to 30%, depending on factors such as patient age, presence of comorbidities, and intraoperative complications (6,7).

Aortic dissection typically presents with sudden, severe chest or back pain and may lead to catastrophic complications such as aortic rupture, cardiac tamponade, or organ ischemia (8).

In overt free rupture all the layers of the aortic wall are disrupted which leads to massive haemorrhage and death before a patient arrives to hospital. In contained aortic ruptures perivascular hematoma is sealed off by periaortic structures such as pleura, pericardium and retroperitoneal space, leading to a relatively hemodynamically stable patient (9). Haemorrhage from the ruptured wall of the ascending aorta can extend along the central

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pulmonary arteries, resulting in an intramural hematoma (PA-IMH) due to their shared aortopulmonary adventitia (10).

The aim of this study was to present a rare case of TAAAD that mimicked pulmonary embolism by causing right heart pressure overload and hemoptysis due to intramural hematoma of the pulmonary arteries extending into pulmonary interstitium. This unusual presentation demonstrates that this complication can closely mimic other life-threatening conditions posing significant diagnostic challenges.

PATIENTS AND METHODS

Patients and study design

A 66-year-old male patient with history of poorly controlled arterial hypertension, hyperlipoproteinemia, smoking and bilateral hip-replacement was transferred to our emergency department from a regional hospital due to sudden chest pain with suspected acute coronary syndrome. Chest pain was described as severe, localized to the anterior chest wall without propagation, with a sudden onset while gardening and performing light lifting (max. 10 kg). It was accompanied by dyspnoea and nausea.

On initial presentation the patient's blood pressure was 170/100mmHg measured on both arms, heart rate was 80 bpm, respiratory rate was 14 breaths/min with peripheral oxygen saturation of 96%. Body temperature was 36.5 °C. The patient was 185 cm tall and weighted 90 kg (BMI 26kg/m²). Physical examination revealed pale skin with diaphoresis, no pathological heart or lung sounds.

Methods

Following the patient-centric algorithms outlined in the American Heart Association's Guideline for the Evaluation and Diagnosis of Chest Pain (11) Electrocardiogram (ECG), point of care ultrasound (POCUS) of the heart (transthoracic echocardiography) (Philips XC50, Netherlands) and triple rule out protocol CT (ECG gated multislice computed tomography angiography) were performed.

RESULTS

The ECG revealed sinus rhythm with no signs of acute ischemia. During the recording, the patient reported worsening nausea and experienced hemoptysis, coughing up approximately 60 mL of bright red blood. The POCUS of the heart revealed an aneurysmal ascending aorta measuring up to 4.5 cm with a suspected intimal flap, mild aortic regurgitation, minimal pericardial effusion and right heart dilatation with impaired function. The left ventricle ejection fraction and the function of the remaining valves were normal. The CT confirmed acute type A aortic dissection with an intimal flap originating just above the aortic bulb, extending 50 mm and terminating at the level of the innominate artery. An extensive intramural hematoma surrounding the central pulmonary arteries and propagating to the peripheral intrapulmonary branches was observed. There was a dilatation of the right ventricle and the right atrium indicative of the right heart pressure overload along with a mild haemorrhagic pericardial effusion anterior to the right ventricle. The supraaortic vascular branches and coronary artery branches were unaffected and no evidence of pulmonary embolism was found (Figure 1).

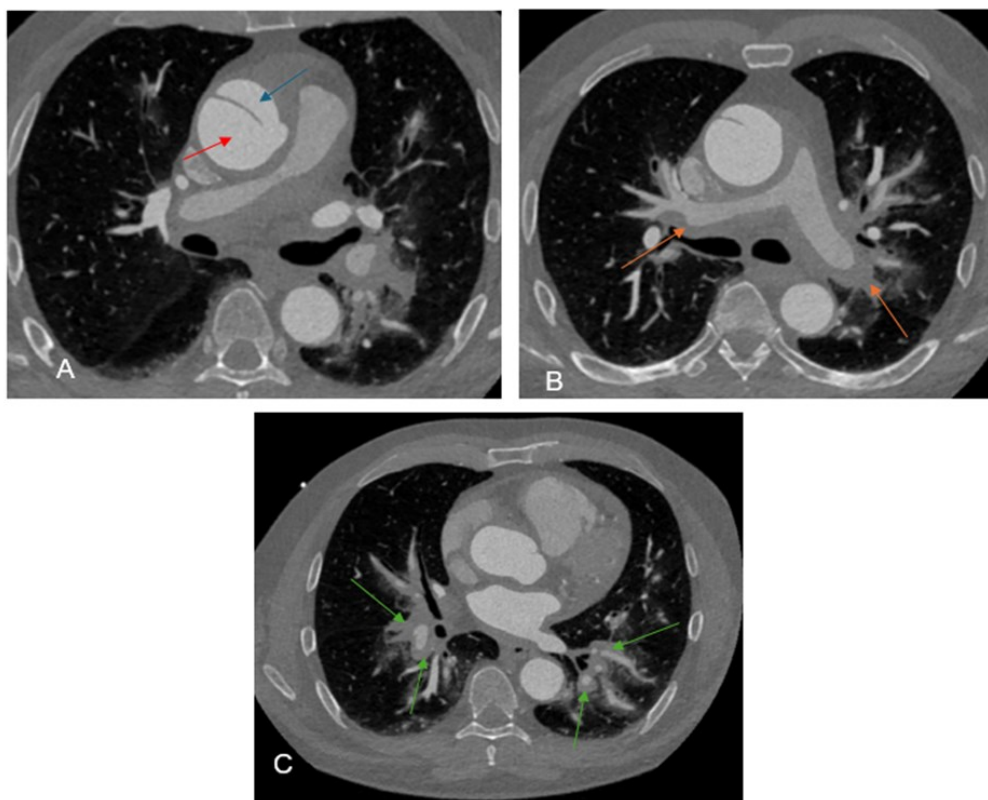


Figure 1. Cardiac computer tomography (CT) angiography - Triple rule out protocol. A) Blue arrow: false lumen, red arrow: true lumen; B) Orange arrows: intramural hematoma around the main pulmonary artery and right and left pulmonary arteries with lumen reduction of the vessels; C) Green arrows: propagation of the hematoma into the wall of distal intrapulmonary branches (Rhön-Klinikum Campus Bad Neustadt, Germany, 2024)

Emergency cardiothoracic consultation was conducted, and the patient was transferred to the intensive care unit. Within hours of admission, urgent surgery was performed including replacement of the supracoronary ascending aorta, as well as aortic hemiarch reconstruction.

Intraoperative transoesophageal echocardiography confirmed the finding observed on initial transthoracic echocardiography (Figure 2).

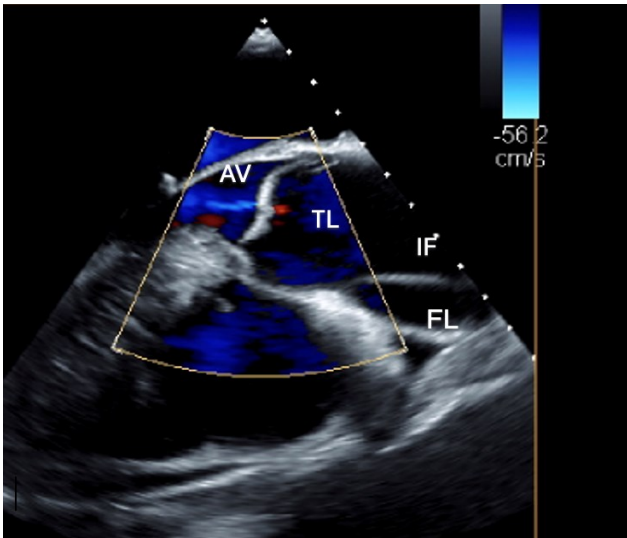


Figure 2. Intraoperative transoesophageal echocardiography (TEE) showing mild aortic regurgitation, true lumen (TL), intimal flap (IF) and false lumen (FL) (Rhön-Klinikum Campus Bad Neustadt, Germany, 2024)

In the immediate postoperative period, the patient developed respiratory failure, which was managed with antibiotics due to elevated infection parameters. Additionally, the patient experienced postoperative delirium which was treated with antipsychotics. The patient was discharged from the intensive care unit after 7 days and left the hospital 17 days post-surgery.

The initial preoperative diameter of the right ventricle of 62 mm, as measured on cardiac CT decreased to 47 mm by the 13 postoperative day, indicated reduction in intrapulmonary pressure after surgery (Figure 3). The follow-up CT angiography of the aorta conducted two months postoperatively revealed complete resorption of the PA-IMH suggesting natural hematoma absorption following surgical aortic repair (Figure 4).

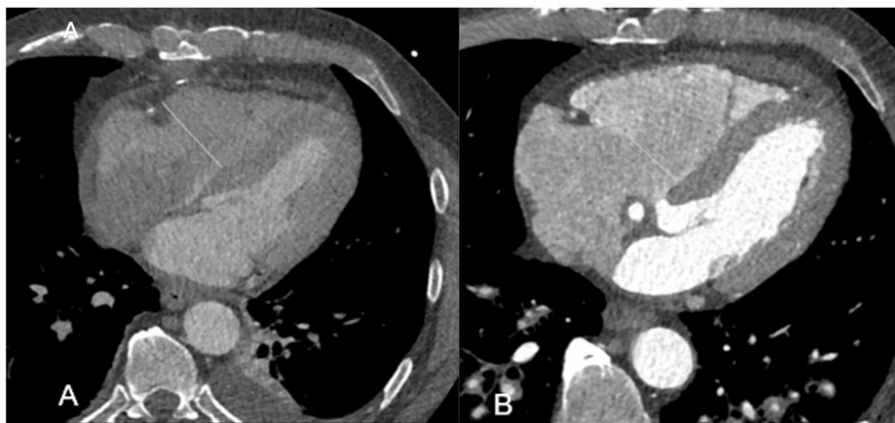


Figure 3. A) Cardiac computer tomography angiography - Triple rule out protocol showing preoperative diameter of the right ventricle of 62mm. B) CT angiography of the aorta on the 13th postoperative day: showing reduced diameter of the right ventricle to 47 mm (Rhön-Klinikum Campus Bad Neustadt, Germany, 2024)

DISCUSSION

Studies have shown that PA-IMH occurs in 9–16% of patients with Stanford type A aortic dissection, manifesting as haemorrhage extending along the pulmonary artery (12). The mechanism of haemorrhage propagation along the pulmonary arteries in aortic dissection becomes evident when correlated with the anatomical relationship of the great vessels of the heart. The ascending aorta and pulmonary trunk have a shared adventitia at the root of the great vessels, extending in the adventitial space of the main pulmonary artery and thereafter into the adventitial space of the left and right pulmonary arteries (13). When a dissection occurs in the posterior aortic wall portion, rupture of the media leads to blood extravasation between the common adventitia and media of the main pulmonary arteries and may further extend into the peribronchovascular interstitium and/or alveoli (10,13). The blood may narrow the lumen of the main PA because the intrapulmonary artery pressure is low (one-tenth of the flow resistance of the systemic circulation) (12). Right pulmonary artery is more affected due to its anatomical correlation (runs horizontally and dorsally) to the ascending aortic root. The main pulmonary vessels are thin-walled and prone to rupture into the alveoli or bronchial airway when subjected to increased arterial pressure, giving rise to hemoptysis (10). Increased intrapulmonary pressure can result in the right heart pressure overload, potentially mimicking pulmonary thromboembolism on imaging. However, it is crucial to differentiate between the two, as anticoagulation or thrombolytic therapy is contraindicated in such cases. Laboratory testing plays a minor role in establishing the diagnosis, as D-dimer levels can be elevated in both pulmonary thromboembolism and acute aortic dissection. However, higher D-dimer values, particularly those exceeding 1,600 ng/mL (normal: ≤ 500 ng/mL), significantly increase the likelihood of an acute aortic syndrome (14).

Patients with TAAAD are typically considered for emergency open surgical repair (15). In cases of PA-IMH due to TAAAD, no specific treatment strategies have been described in the literature. In the presented case, follow-up CT angiography of the aorta performed two months postoperatively revealed complete resorption of the PA-IMH, suggesting natural hematoma absorption after surgical aortic repair. Lizhi reported a similar observation, further demonstrating that the presence of

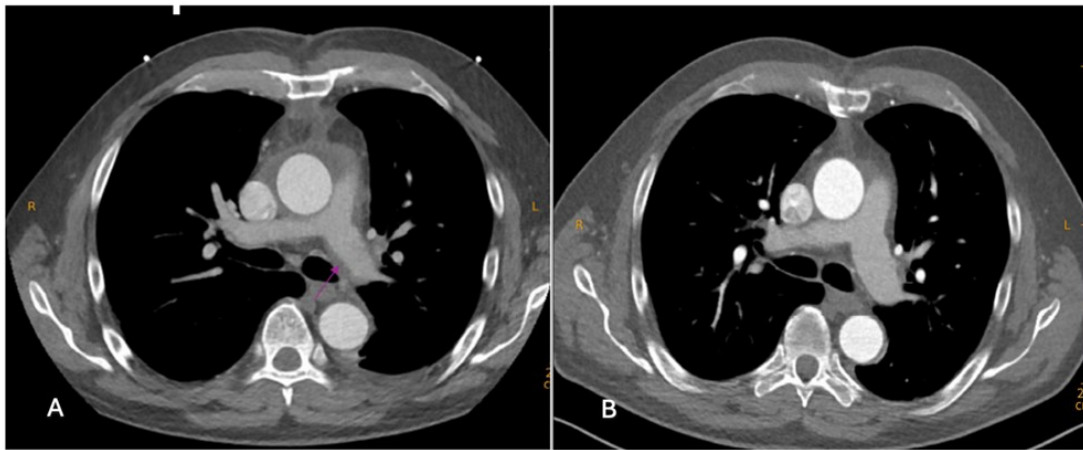


Figure 4. CT angiography of the aorta. **A)** 13th postoperative day: showing an initial resorption of the pulmonary arteries intramural hematoma (PA IMH); **B)** complete resorption of the PA IMH two months after surgery (Rhön-Klinikum Campus Bad Neustadt, Germany, 2024)

PA-IMH due to TAAAD significantly increased early postoperative mortality rates, particularly among older patients (>70 years). The risk was amplified by the extent of PA-IMH; however, it did not influence long-term prognosis when compared to patients without PA-IMH (16).

Although initially described by Buja et al. in 1972 (17), numerous unanswered questions still remain regarding the clinical outcomes of PA-IMH caused by TAAAD, particularly its short- and long-term effects on pulmonary circulation. Prospective multi-centre clinical trials and registries are urgently needed to provide pragmatic insights and guide management strategies.

In conclusion, physicians must maintain a high index of suspicion for atypical presentations of aortic dissection and promptly consider tomographic imaging and transfer to tertiary care, as the mortality rate associated with aortic dissection rises with each passing hour (5). Pulmonary artery intramural hematoma may result from rupture of the posterior wall of the ascending aorta into the shared aortopulmonary adventitia, potentially causing lumen narrowing or occlusion of the pulmonary arteries. This can lead to the right heart pressure overload, mimicking pulmonary thromboembolism on imaging. Timely differentiation is crucial as anticoagulation is contraindicated in such cases.

AUTHORS CONTRIBUTIONS

Conceptualization, E.R.D.; Writing – original draft, E.R.D.; Supervision, S.B.; Data curation, L.L.; Writing – review & editing, D.C. All authors have read and agreed to the published version of the manuscript.

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Conflict of interests: None to declare.

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