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Baffle thrombosis in an adult with remote prior scimitar vein repair mimicking massive pulmonary embolism☆

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ABSTRACT

A 58-year-old man with a history of Scimitar syndrome diagnosed and surgically repaired in early adulthood presented multiple times to the emergency department complaining dyspnea, chest pain, and hemoptysis. Asymmetric pulmonary arterial flow rates between left and right lungs resulted in an apparent filling defect on computed tomographic pulmonary arteriography, which was repeatedly misdiagnosed clinically and radiologically as a massive pulmonary embolus. This case highlights the importance of understanding the pathophysiology and post-surgical complications of repaired congenital cardiovascular disease. Delayed phase acquisitions are often necessary to characterize the physiology of repaired congenital cardiovascular disease with associated shunts.

1. Introduction

Scimitar syndrome describes a constellation of findings, most typically anomalous right pulmonary vein connecting to the right cardiac circulation [usually via the inferior vena cava (IVC)], right-sided pulmonary hypoplasia, and systemic arterial collaterals supplying the right lung. Various additional cardiopulmonary variations and associations have been described including atrial septal defect (ASD) and ventricular septal defect (VSD), pulmonary artery (PA) hypoplasia, diaphragmatic hernias, abnormal bronchopulmonary anatomy and sequestrations, and vertebral and genitourinary abnormalities [1,2].

The radiographic appearances of Scimitar syndrome are well known, and its accompanying asymmetric perfusion on conventional angiography is documented [3]; however, the associated appearance of abnormal pulmonary arterial blood flow on computed tomography pulmonary angiography (CTPA) has not previously been described. We report a case of post-operative scimitar repair and asymmetric pulmonary arterial perfusion with delayed right pulmonary blood flow, repeatedly misinterpreted as massive pulmonary embolus (PE).

2. Case report

We describe the most recent clinical episodes of a 58-year-old man, who presented to the emergency department (ED) of a metropolitan teaching hospital complaining recurrent chest pain, hemoptysis, and dyspnea on four occasions with multiple intervening cardiac and respiratory outpatient clinics, before his diagnosis was confirmed on the fourth episode when he was transferred to a specialist cardiothoracic center for evaluation. His course was complicated by a misinterpretation of clinical findings and, in retrospect, misdiagnosis of PE during an unrelated presentation many years earlier.

2.1. First encounter

The patient described a past history of single vessel ischemic heart disease treated with right coronary arterial stent, paroxysmal atrial fibrillation, pulmonary hypertension, PE and elective repair of Scimitar syndrome (right pulmonary veins redirected to left atrium via intra-atrial and trans-septal baffle) discovered incidentally at age 29 years.

A triage chest radiograph (not shown) was performed on arrival. It demonstrated a previous median sternotomy but no discernable scimitar vein remnant. Reduced right hemithoracic volume was misinterpreted as a pulmonary effusion and vague right-sided parenchymal opacities were described. Initial history provided to the radiologist did not include the prior history of scimitar vein repair but had been documented in the clinical notes, which were not requested by the reporting radiologist.

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A CTPA study (Somatom AS+64 slice; Siemens Corporation, Munich, Germany) demonstrated a large proximal right PA filling defect (Fig. 1a), right supraclavicular and mediastinal lymphadenopathy, right-sided interlobular septal opacities, and mosaic and nodular ground glass attenuation. A diagnosis of large right PA embolus was made with right pulmonary parenchymal changes attributed to the hemodynamic effects of large volume proximal obstruction.

As the patient was therapeutically anticoagulated for prior PE at presentation, concern for occult malignancy prompted whole body imaging (not shown), but no cancer was detected. Bilateral lower limb venous Doppler (not shown) was negative for deep venous thrombosis, but an IVC filter was inserted and unfortunately complicated by a femoral insertion site arteriovenous fistula (treated conservatively). Six-week follow-up CTPA (not shown) was unchanged.

2.2. Second encounter (5 months)

A second ED presentation for recurrence of chest pain and hemoptysis prompted another CTPA, which demonstrated improved yet persistent asymmetric contrast opacification of the right PA, stable appearing lymphadenopathy, and parenchymal changes (Fig. 1b). Despite this, and after apparent direct comparison with the initial CTPA, it was concluded that the PE had resolved.

2.3. Third encounter (11 months)

Ongoing symptoms yielded another "positive" CTPA (not shown) with a diagnosis of refractory PE. Persisting right-sided interstitial changes raised concern for lymphangitic carcinomatosis and a combined positron emission tomography (PET)/computed tomography (CT) study was performed (not shown) to exclude PA sarcoma; it demonstrated intense FDG uptake within the lymphadenopathy, mild uptake in the right lung, and no uptake in the right PA. A fine needle aspiration of an enlarged FDG avid supraclavicular lymph node yielded only inflammatory cells.

2.4. Fourth encounter (18 months)

Suspicion of an alternative diagnosis after another apparently positive CTPA (not shown) prompted a contemporaneous CMR (Magnatom Aera; Siemens Corporation, Munich, Germany). Axial T1 post-contrast sequence demonstrated no PA filling defect (Fig. 2a). The multi-phase dynamic magnetic resonance pulmonary angiography (MRPA) resolved the ongoing diagnostic dilemma.

Early-phase MRPA (Fig. 2b) confirmed asymmetric PA perfusion hemodynamics resulting in delayed contrast flow artifact in the right main PA, mimicking a large proximal PE. Late-phase MRPA sequences (70 s) demonstrate that delayed acquisition permitted contrast to opacify the more slowly filling right PA system (Fig. 2c), accounting for the apparent resolution of PE (Fig. 1b), which in retrospect was due to a late CTPA acquisition (note the opacification of the descending thoracic aorta). No flow through the intra-atrial baffle could be demonstrated.

3. Outcome

The patient was diagnosed with baffle occlusion and right pulmonary venous hypertension that was a significant contributor to the reduced right pulmonary arterial flow. The IVC filter was removed without incident and anticoagulation was ceased. At the time of writing, the patient reports no subsequent episodes of hemoptysis and as such has not yet required embolization of his arterial collaterals. His chest pain and dyspnea persist but have improved.

4. Discussion

Clinical manifestations of Scimitar syndrome are variably dependant on the extent of left-to-right shunt, presence of ASD or VSD, valvular abnormalities, and degree of pulmonary hypertension [4]. Perfusion of the right lung is commonly reduced in Scimitar syndrome [5], in both pre-operative and post-operative states [6]. Perfusion abnormality is likely the result of hypoplastic lung, a small right PA or pulmonary venous obstruction, and while surgery can correct the vascular shunt, underlying pulmonary architectural abnormalities persist. These background abnormalities can result in lymphatic congestion, which would explain the persistent parenchymal findings in the right lung in this case (Fig. 1c) rather than the erroneous suspicion of lymphangitic carcinomatosis.

A literature search (PubMed/MEDLINE, CINAHL, and Google Scholar) yielded only one publication with images demonstrating such asymmetric perfusion on (conventional) pulmonary angiography in the setting of Scimitar syndrome [3].

In almost every case, the CT studies were read by different radiologists and the history of previous PE may have prejudicially resulted in an element of *confirmation bias*. Interestingly, the previous diagnosis of PE was made many years earlier by interstate intensive care physicians during an admission for a severe community-acquired pneumonia. The conclusion was drawn despite a negative CTPA (presumably performed on an older slower machine) and was based on asymmetric pulmonary arterial pressures during invasive monitoring. Unfortunately, this history, potential bias, and misinterpretation resulted in a total of six CTPAs, one CT chest/abdomen/pelvis, and one PET/CT performed during the described timeframe, with an estimated effective dose of 83.4 mSv[1] [7,8].

CTPA has replaced conventional pulmonary angiography to become the gold standard in the diagnosis of PE but has inherent limitations. While it is essentially non-invasive and can provide excellent anatomic detail, it does not, under typical circumstances,

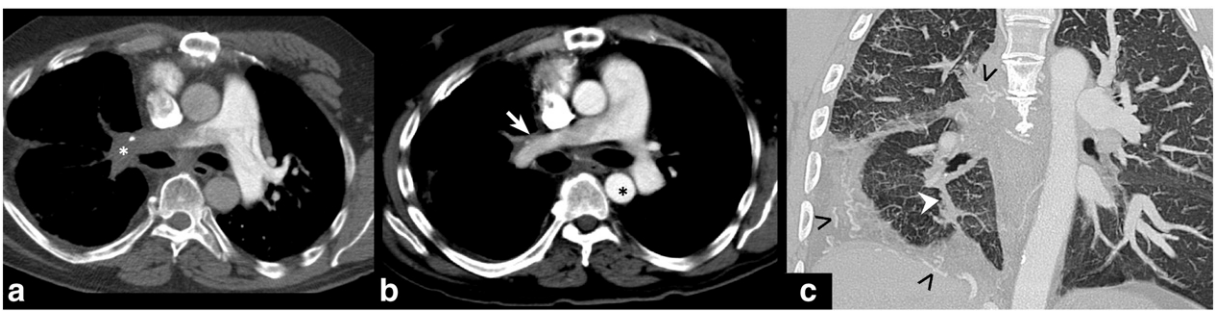

Fig. 1. (a) Large right PA filling defect (white asterisk), with relatively dense contrast opacification of the left pulmonary arterial tree. (b) Improved contrast opacification of the right PA (white arrow); however, disparity between left and right pulmonary flow remains. Dense contrast opacification of the descending aorta (black asterisk) reflects unintentional late acquisition. (c) Network of arterial (bronchial and hepatic) collaterals supplying the right lung (open black arrowheads). Post-surgical remnant of the scimitar vein (white arrowhead).