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CASE REPORT

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A rare case of scimitar syndrome with pulmonary arterial hypertension in an adult female

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Abstract

Scimitar syndrome is a rare congenital anomaly characterized by partial or total anomalous pulmonary venous drainage of the right lung to the inferior vena cava. We report a case of a 67-year-old female who presented with cough and dyspnea and was diagnosed with scimitar syndrome and pulmonary arterial hypertension based on comprehensive imaging and hemodynamic evaluation. This case highlights the importance of considering scimitar syndrome as a cause of pulmonary hypertension even in adult patients.

KEYWORDS

computed tomography angiography, pulmonary arterial hypertension, scimitar syndrome

INTRODUCTION

Scimitar syndrome is a rare congenital anomaly with an estimated prevalence of 1–3 per 100,000 live births.[1] It is characterized by partial or total anomalous pulmonary venous drainage of the right lung to the inferior vena cava, forming a scimitar-shaped shadow on chest radiographs. The anomalous pulmonary vein often drains into the inferior vena cava just below the diaphragm. The syndrome is commonly associated with hypoplasia of the right lung and signs of pulmonary hypertension.[2]

Scimitar syndrome can be complicated by pulmonary arterial hypertension (PAH).[3] PAH results from increased pulmonary blood flow due to the left-to-right shunt created by the anomalous venous drainage. The pathophysiology behind this combination is not well understood. There are limited reports on the diagnosis and management of scimitar syndrome with PAH in adult patients. We present a rare case of an adult female diagnosed with scimitar syndrome and PAH.

CASE PRESENTATION

A 67-year-old female presented to our hospital with a 1-month history of dry cough and progressive shortness of breath. She had no other respiratory or cardiac symptoms. Her past medical history was unremarkable with no prior diagnosis of congenital heart disease. She was a lifelong nonsmoker and had no exposure to environmental toxins.

On examination, her vital signs were normal but oxygen saturation was 89% on room air. Chest auscultation revealed decreased breath sounds on the right side.

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The rest of the clinical examination was normal. Routine blood tests showed no significant abnormalities.

Chest X-ray showed a smooth curving structure projecting across the right pulmonary hilum toward the lung base. (Figure 1). Subsequent computed tomography angiography of the pulmonary arteries showed enlargement of the pulmonary artery and its branches, and the right pulmonary veins were not visualized in their expected location (Figure 2). There are multiple ways to demonstrate dilation of the right pulmonary veins, and abnormal drainage into the right atrium (Figure 3). Cardiac ultrasound demonstrated an enlarged right ventricle (Figure 4a), prominent pulmonary artery dilation (Figure 4b), with tricuspid regurgitation jet velocity measured at 3.4 m/s, suggestive of PAH. Right ventricular contrast echocardiography demonstrated retrograde flow from the right pulmonary vein into the right atrium via the superior vena cava, consistent with the findings of scimitar syndrome (Figure 4c). No intracardiac shunting was seen, ruling out atrial or ventricular septal defects. Right heart catheterization demonstrated a mean pulmonary artery pressure of 40 mmHg, confirming the diagnosis of PAH.

Based on integrated findings from multiple diagnostic modalities, a diagnosis of scimitar syndrome with PAH was made. The patient was started on sildenafil 20 mg three times daily. She reported improvement in her symptoms at follow-up visits.

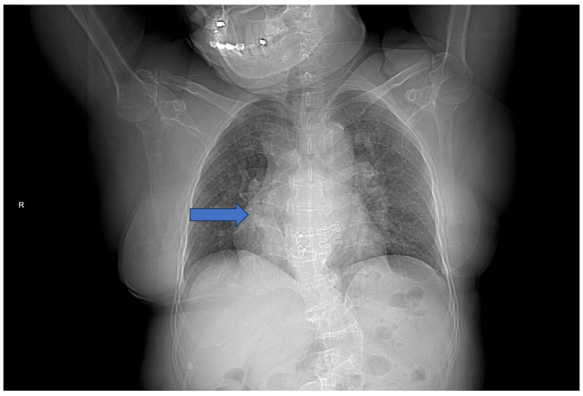

FIGURE 1 Chest X-ray findings: A smooth curving structure projecting across the right pulmonary hilum toward the base of the lung.

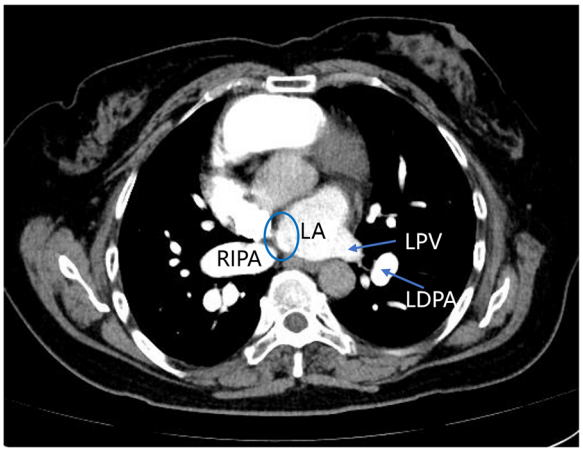

FIGURE 2 Computed tomography angiography imaging: The image reveals an enlargement of the pulmonary artery and its branches. Notably, the right pulmonary veins are not visualized in their expected location (circled). LA, left atrium; LDPA, left descending pulmonary artery; LPV, left pulmonary vein; RIPA, right interlobar pulmonary artery.

DISCUSSION

Our case highlights two important learning points. Firstly, scimitar syndrome should be considered in the differential diagnosis even in adult patients presenting with respiratory complaints. Although more commonly diagnosed in childhood, some cases escape early detection and remain undiagnosed until later in life.[4]

Second, scimitar syndrome can be complicated by pulmonary arterial PAH, though the true incidence is unclear. Previous studies have reported the incidence of PAH in scimitar syndrome ranges from 10% to 50%.[2,4–8] The incidence may differ between adult and pediatric populations. Reports indicate PAH complicates approximately 10%–30% of adult scimitar syndrome cases[4] whereas 30%–50% of infantile scimitar syndrome patients develop PAH.[9,10] The presence of the left-to-right shunt allows increased pulmonary blood flow which causes endothelial dysfunction and remodeling of distal pulmonary arterioles, resulting in PAH.[7]

The pathophysiology behind the development of PAH in scimitar syndrome is not fully understood, but several potential mechanisms can be inferred. The anomalous pulmonary venous drainage in scimitar syndrome results in a left-to-right shunt, which increases blood flow to the pulmonary arteries. This increased blood flow can cause endothelial dysfunction and remodeling of the pulmonary arterioles, leading to PAH.[9,11] Additionally, the hypoplasia of the right lung and pulmonary artery commonly seen in scimitar syndrome may contribute to the development of PAH by creating an imbalance in blood flow distribution.[12]

The management of scimitar syndrome with PAH poses a therapeutic challenge. It warrants a multidisciplinary approach with input from interventional cardiology, pulmonary medicine, and cardiothoracic surgery. Treatment options include surgical repair of the anomalous pulmonary vein, transcatheter device closure of the shunt, medical

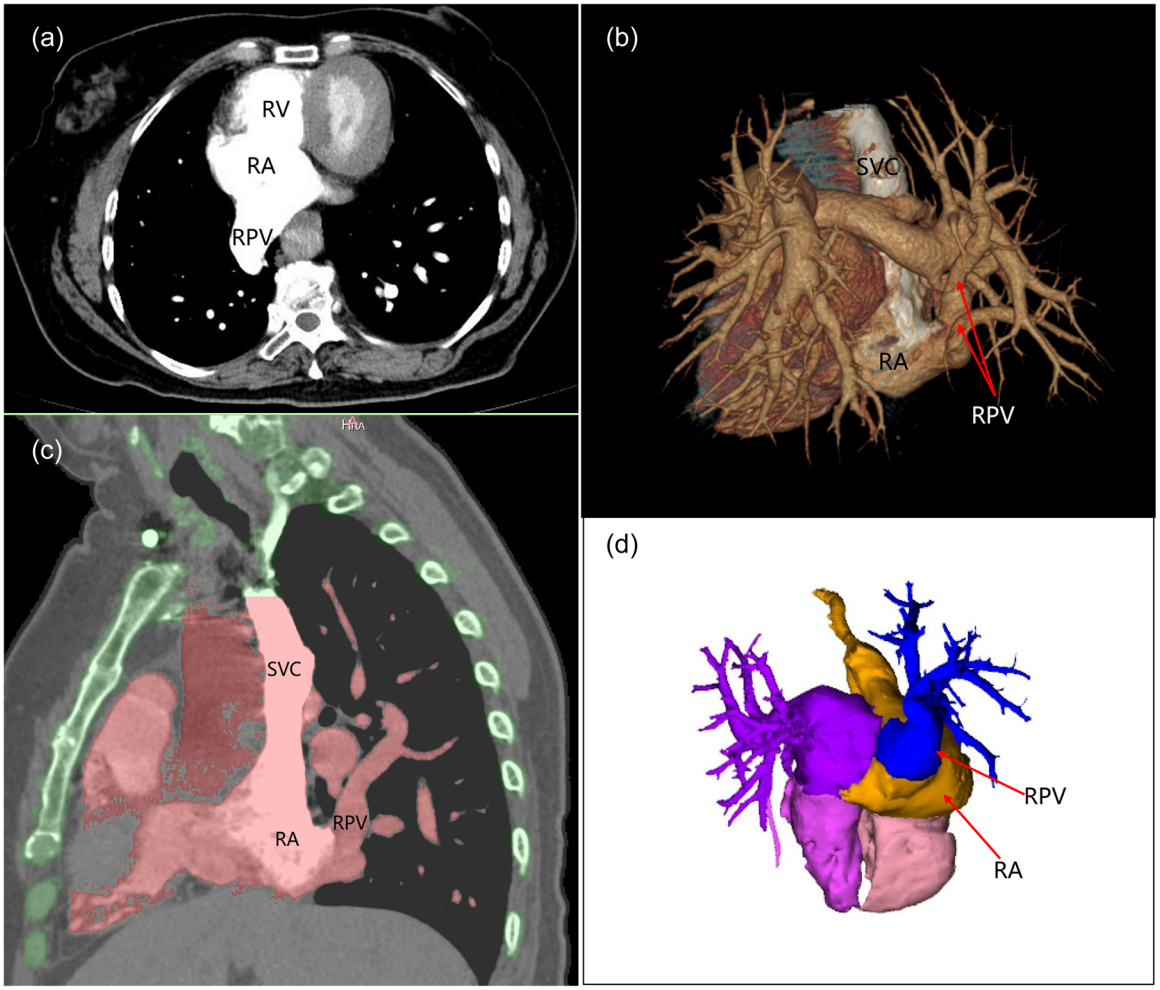

FIGURE 3 (a) CTA imaging shows the right pulmonary vein is dilated, with abnormal reflux into the right atrium. (b–d) Clear visualization of abnormal reflux of the right pulmonary vein into the right atrium is evident in the 3D reconstructed images from CTA, multiplanar reformation reconstructed images from CTA, and vascular reconstruction images using Mimics Research 20.0 software. 3D, three-dimensional; CTA, computed tomography angiography; RA, right atrium; RPV, right pulmonary vein; RV, right ventricle; SVC, superior vena cava.

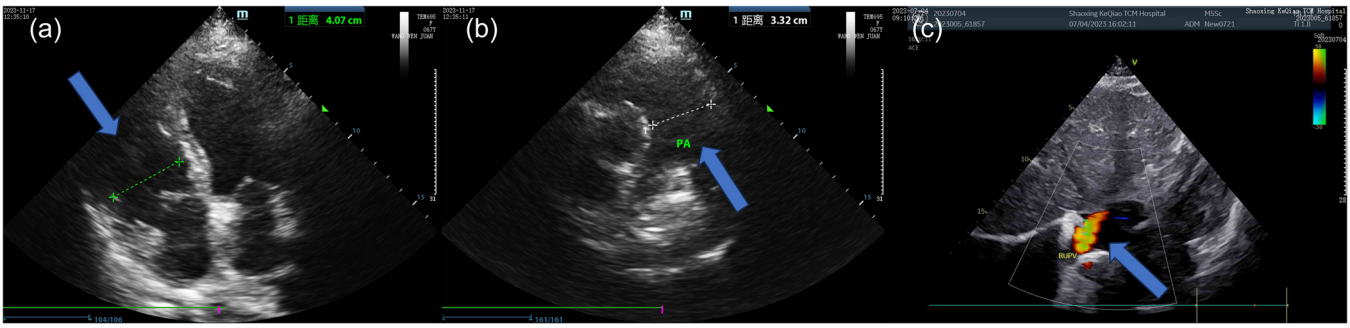

FIGURE 4 (a) Parasternal long axis view shows an enlarged right ventricle (arrow). (b) Suprasternal view depicts dilation of the main pulmonary artery (arrow). (c) Apical four chamber view on right ventricular contrast echocardiography demonstrates retrograde flow from the right pulmonary vein into the right atrium via the superior vena cava (arrow), correlating with scimitar syndrome anatomy.

management of PAH, or lung transplantation in refractory cases.[5,6] The choice depends on the anatomy, shunt size, severity of PAH, and presence of other comorbidities in the patient population.

In conclusion, this case highlights the importance of considering scimitar syndrome as a rare cause of pulmonary hypertension even in adult patients with no prior diagnosis of congenital heart disease. Increased awareness of this

association can help guide appropriate diagnostic workup and management. Further research is warranted to better understand the pathophysiology and improve the management of scimitar syndrome with PAH.

AUTHOR CONTRIBUTIONS

Yang Liu, Weiliang Ruan, and Ziye Li contributed equally to this work. They were involved in patient care, data collection, and manuscript writing. Hua Wang, Shenghai Chen, Yuhong Ding, and Jianfeng Jin were involved in patient care and data collection. All authors read and approved the final manuscript.

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CONFLICT OF INTEREST STATEMENT

The authors declare no conflict of interest.

ETHICS STATEMENT

Not required.

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