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Case Report

A late presentation of scimitar syndrome in adult ☆,☆☆

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ABSTRACT

Scimitar Syndrome is a rare congenital disorder and a variant of partial anomalous pulmonary venous connection (PAPVC) in which part or even the entire right lung is drained by right pulmonary veins that connect anomalously to the Inferior Vena Cava (IVC). It has various presentations including exertional dyspnea, recurrent chest infection, pulmonary artery hypertension, and hemoptysis. The initial diagnosis of PAPVC may be made by echocardiography and is typically confirmed by magnetic resonance imaging, computed tomography, or cardiac catheterization. We report a 69-year-old man with progressive dyspnea on exertion associated with palpitations of five years of evolution. The patient was diagnosed with cardiomyopathy, pulmonary hypertension and was started on treatment with anticoagulation, digoxin, and metoprolol for his atrial fibrillation. Despite the treatment, the dyspnea did not improve. The patient underwent cardiac catheterization, where the anomalous venous drainage was confirmed. Scimitar syndrome was corrected by surgical intervention with complete resolution of symptoms.

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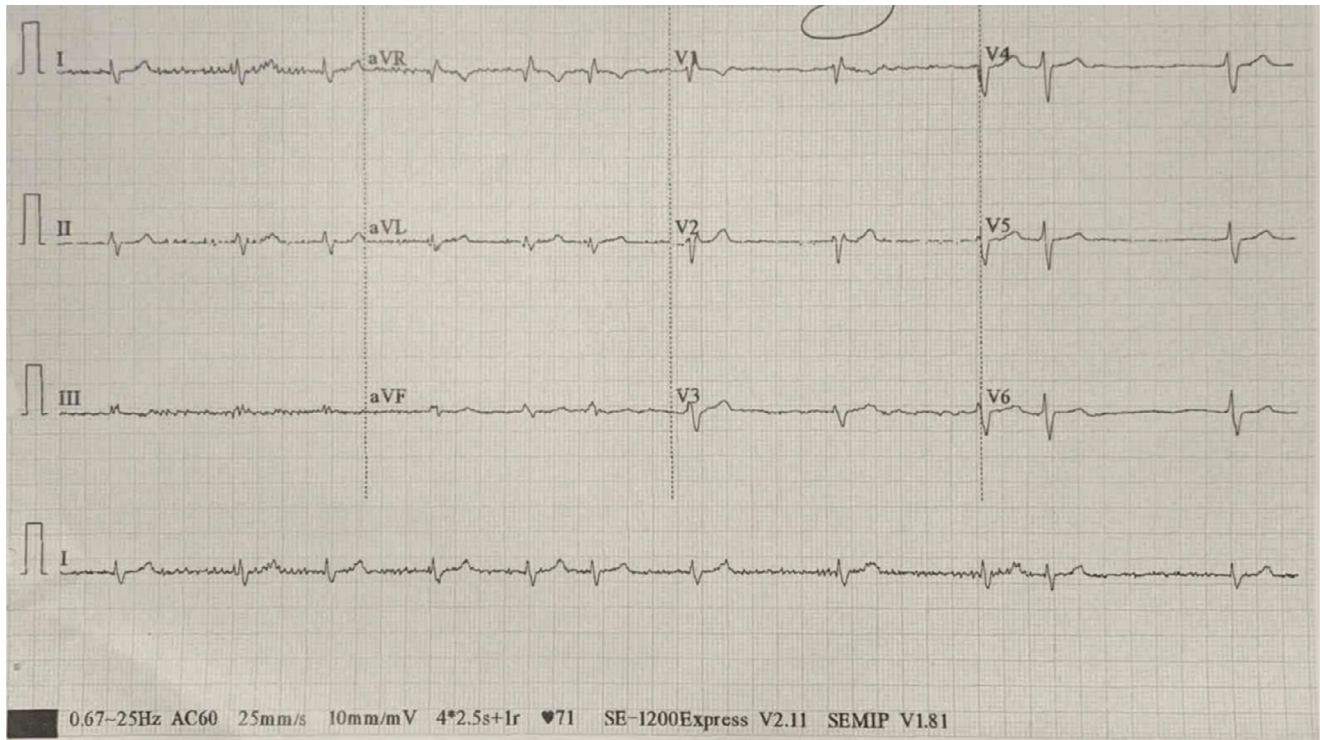


Fig. 1 – Patient’s initial EKG: atrial fibrillation (A-fib) with adequate ventricular response, incomplete right bundle branch block, right ventricular enlargement, right axis deviation, low voltage, and poor R progression.

Introduction

This patient presented with scimitar syndrome, a rare combination of partial anomalous pulmonary venous return, right lung hypoplasia, and dextroposition of the heart.

Partial anomalous pulmonary venous return, a congenital disorder, results in an abnormal return from the pulmonary veins to the right side of the heart resulting in oxygenated blood from the pulmonary vein reaching a systemic vein before reaching the right atrium. Resulting in a left-to-right shunt [1].

Scimitar syndrome is an uncommon congenital disorder and a rare partial anomalous pulmonary venous return variant commonly diagnosed on infants in which part of or the entire right lung is drained by the pulmonary vein into the inferior vena cava, giving the curvilinear dimension the appearance of a Turkish sword (Scimitar) [2–4].

The syndrome is associated with hypoplasia of the right lung and pulmonary artery, dextroposition of the heart, and anomalous systemic arterial supply to the ipsilateral lung [2,5]. Without the characteristically curved anomalous right pulmonary vein, the pathology cannot be defined, and a different etiology must be searched [2].



Fig. 2 – Cardiac catheterization showing anomalous venous drainage.

Table 1 – Cardiac study results.

Echocardiography	Right heart catheterization	Left heart catheterization	Saturation run
Severe RAE	RAP: 16 mm Hg	BP: 150/90 mm Hg	SVC: 83.3%
Massive RVE	RVP: 39/10 mm Hg	MAP: 110 mm Hg	RA: 86.1%
Severe TR	PAP: 39/14 mm Hg	AO: 150 mm Hg	PA: 86.7%
Bubble Test Negative	MPAP: 22 mm Hg	LVP: 149 mm Hg	Ao: 98.2%
RVSP: 30 mm Hg	PCWP: 20 mm Hg	EDP: 30 mm Hg	PV: 90.5%
EF: 60%	CO: 8.02 L/min	EF: 60%	Qp/Qs: 3.8
	CI: 4.2L/min/m ²	Free of COD	Systemic and pulmonary blood flow

RAE, right atrial enlargement; RAP, right atrial pressure; BP, blood pressure; SVC, superior vena cava; RVE, right ventricular enlargement; RVP, right ventricular pressure; MAP, mean arterial pressure; RA, right atrial; TR, tricuspid regurgitation; PAP, pulmonary arterial pressure; AO, aortic outflow; PA, pulmonary artery; MPAP, mean pulmonary artery pressure; LVP, left ventricular pressure; Ao, aortic; PCWP, pulmonary capillary wedge pressure; EDP, end diastolic pressure; PV, pulmonary venous; CO, cardiac output; EJ, ejection fraction; Qp/Qs, pulmonary-systemic flow ratio; CI, cardiac index; COD, Coronary Obstructive Disease.

**Fig. 3 – The right pulmonary veins draining in the inferior vena cava.**

Case presentation

We report a 69-year-old man with a past medical history of arterial hypertension, atrial fibrillation, hypothyroidism, and gastroesophageal reflux disease who presents with worsening acute over chronic progressive exertional dyspnea for 5 years and palpitations. Physical examination was remarkable for an irregular heart rhythm, with no murmur, no jugular venous distention, and no hepatojugular reflex. Lungs were clear on auscultation with no crackles or decreased breath sounds and no cyanosis or peripheral edema. The patient denied dizziness, headache, blurred vision, cough, chest pain, paroxysmal nocturnal dyspnea, weight loss, syncope, or any toxic habit.

Electrocardiogram was performed in the emergency department, which showed atrial fibrillation with adequate ventricular response, incomplete right bundle branch block, right ventricular enlargement, right axis deviation, low voltage, and poor R progression (Fig. 1). A transesophageal echocardiogram was performed showing severe tricuspid regurgitation, right ventricular enlargement with anomalous venous drainage and pulmonary hypertension. (Table 1)

The patient underwent cardiac catheterization, where the anomalous venous drainage was confirmed (Fig. 2). A chest CT angiography was performed, showing a right pulmonary vein drainage into the inferior vena cava (Figure 3, 4, 5 and 6). Given the catheterization results and CT angiography, cardiothoracic surgery was successfully performed to correct the anoma-

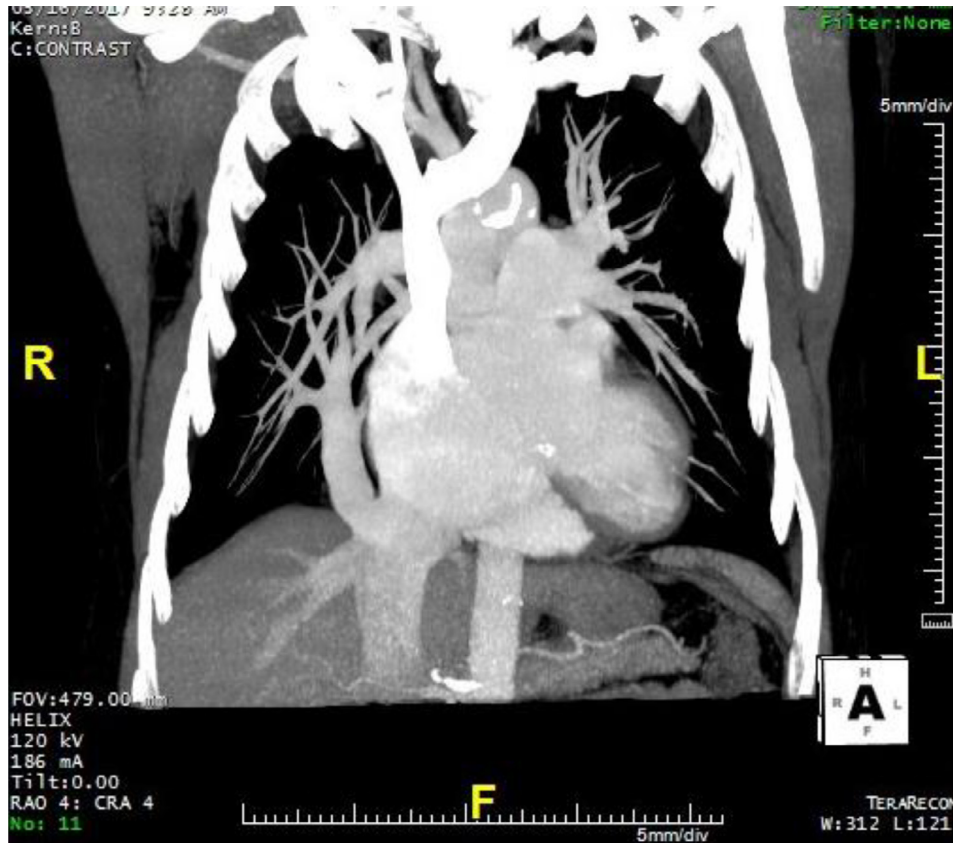


Fig. 4 – CT of the thorax, right pulmonary vein draining into inferior vena cava.

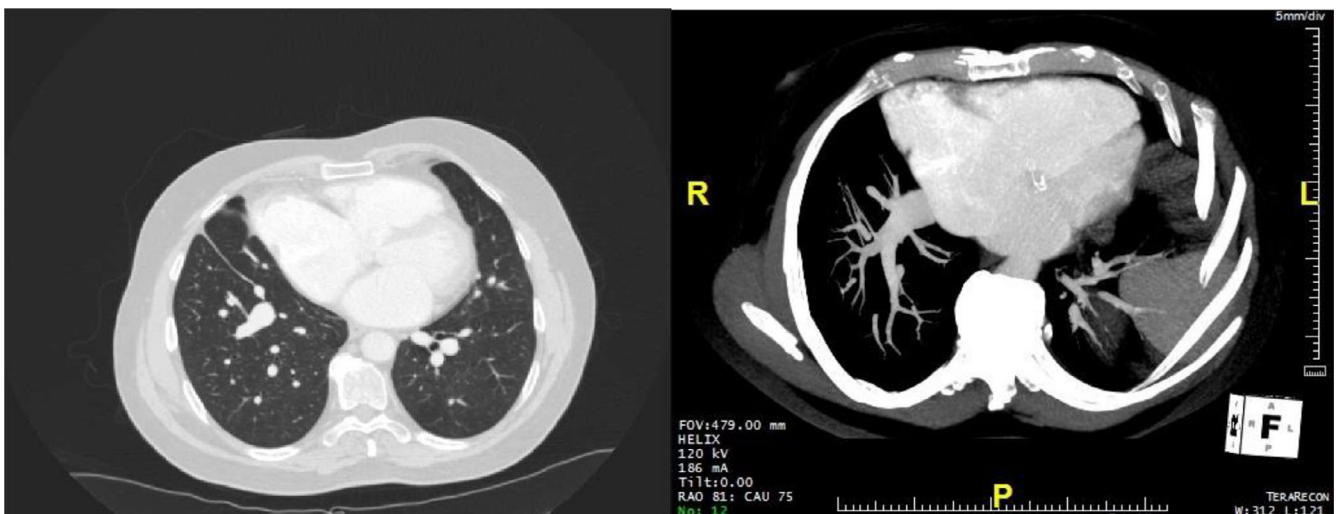


Fig. 5 – Axial CT image through the heart showing the four cardiac chamber size.

lous venous drainage and the tricuspid valve repaired. The postoperative transesophageal echocardiogram confirmed a wide patent path between the Scimitar vein and the left atrium and a wide-open inferior vena cava draining into the right atrium. The tricuspid valve had only mild residual regurgitation.

Discussion

In most infantile patients, Scimitar syndrome is identified as an isolated lesion with a benign outcome. Nonetheless, the prognosis is poor when found in association with cardiac de-

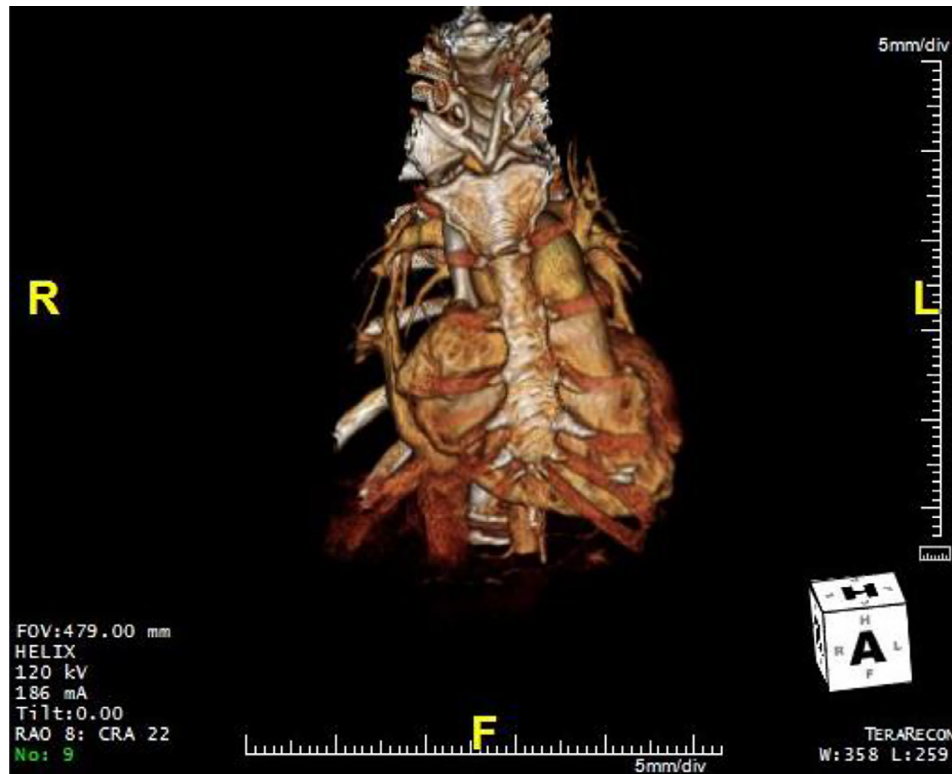


Fig. 6 – 3D CT reconstruction, right pulmonary vein draining into the IVC.

fects and pulmonary arterial hypertension an increased risk of congestive heart failure [6]. The adult presentation of the syndrome is often overlooked clinically as patients are often asymptomatic.

Diagnosis work-up starts with relies heavily on advanced imaging studies and cardiac catheterization can aid in diagnosing. A plain chest X-ray can show the shadow of the descending right pulmonary vein, dextroposition of the heart, and hypogenesis of the lung while a chest CT scan can provide detailed information about the anatomy of the bronchial trees, vasculature, and cardiac chambers.

Our patient debuted with symptoms as an adult, with a left to right shunt. If this shunt is not repaired, pulmonary vascular remodeling can occur resulting in the development or worsening of the pulmonary arterial hypertension [1]. Due to the patient undiagnosed Scimitar syndrome with no previous intervention, severe worsening of his shunt and pulmonary condition including newly diagnosed pulmonary arterial hypertension surgery was performed.

Atrial fibrillation in the patient can be explained mostly in part due to the atrium enlargement and the development of arterial pulmonary hypertension due to the left to right shunt discovered upon admission.

In conclusion, Scimitar syndrome is a rare congenital pathology and must not be overlooked as a benign condition. Most commonly seen in infants, physicians must be aware of the adulthood presentation in previously asymptomatic patients. Associated cardiac symptoms and pulmonary hypertension play a key role in long-term outcome, management, and mortality as pulmonary hypertension increases mortality rate in patients. In addition, advanced imaging studies along-

side associated cardiac and pulmonary symptoms monitoring can help in deciding when is appropriate to use surgery as a definitive treatment as asymptomatic patients with small left to right shunts do not need intervention.

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