Total transcatheter correction of scimitar variant having dual pulmonary venous drainage

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ABSTRACT

Scimitar syndrome is a clinical triad of anomalous pulmonary venous drainage, lung hypoplasia, and anomalous aortic blood supply to the lung segment. When there is dual pulmonary venous drainage both to inferior vena cava and left atrium, it is called scimitar variant. A young child presenting with recurrent chest infections, dextroposition of the heart, and scimitar shadow on chest X-ray was identified to have scimitar variant after a detailed evaluation and managed successfully by catheter interventions. This report discusses the embryogenesis and angiographic findings of scimitar variants, indications for interventions, and issues in its management.

Keywords: Catheter intervention, coil closure, double-arched pulmonary veins, dual pulmonary venous drainage, Scimitar syndrome, scimitar variant

INTRODUCTION

Classical Scimitar or venolobar syndrome is characterized by anomalous pulmonary venous drainage to the inferior vena cava that radiologically presents as a "Turkish sword."[1] A few patients who have an additional dual drainage to left atrium are termed "Scimitar variants."[2] Surgical treatment in young symptomatic patients is challenging due to pulmonary hypertension, reduced lung vascular bed, small pulmonary veins, and difficult access to aortic collaterals.[1] Even though clinical results are good, catheter closure of aortic collaterals only partially corrects the hemodynamics but leaves the left-to-right shunt through the anomalous pulmonary veins.[3] A young symptomatic child with scimitar variant and dual drainage presented with lung hypoplasia, dextroposition, and aortic collaterals and was managed nonsurgically with correction of both hemodynamic problems.

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CASE REPORT

A 2-year-old girl presented with recurrent wheezy chest infections since infancy. Clinical examination showed a dextroposed heart with no other significant cardiac findings. The chest radiograph revealed dextroposition, an abnormal scimitar shadow along the right heart border without cardiomegaly [Figure 1a]. Right heart chambers were mildly dilated, and an abnormal pulmonary vein was draining into the inferior caval vein with mild turbulence [Figure 1b]. There were no associated intracardiac anomalies. Catheter hemodynamics showed a shunt ratio of 1.11:1, pulmonary artery pressures were 25/6 (mean 16) mmHg, and indexed pulmonary vascular resistance was 1.6 Wood units. An abdominal aortic large collateral with systemic pressures (75/45, mean 68 mmHg) perfused the lower lobe of the right lung, and

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venous phase demonstrated the double-arched scimitar vein to inferior vena cava [Figure 2]. This was closed with multiple coils [Video 1]. Right pulmonary arteriogram showed dual drainage of the right lung both to left atrium and to inferior vena cava with a connecting vein between the two draining veins [Figure 3 and Video 2]. The scimitar vein was accessed from femoral venous catheter and closed at the vena caval junction using multiple coils, which rerouted the venous drainage of the right lung to the left atrium [Figure 4 and Video 3]. On low-dose aspirin, she remained free of the wheezy respiratory illnesses with normal size of cardiac chambers on echocardiography at 1-year follow-up.

DISCUSSION

Scimitar syndrome is caused by the disordered embryonic development of the entire lung bed, resulting in anomalous venous drainage to the vitelline veins, lung hypoplasia, and anomalous aortic blood supply to the lungs.[1] Infantile form with pulmonary hypertension and heart failure often has poor outcomes, but adult form is associated with normal pulmonary artery shunt and clinically tolerated left-to-right pretricuspid shunt.[3]

Interventions are needed if (i) significant left-to-right shunt (uncommon due to lung hypoplasia that permits preferential contralateral lung flows), (ii) heart failure and pulmonary hypertension which are often not only due to aortic collaterals to the lungs but also due to pulmonary venous narrowing or lung hypoplasia, (iii) recurrent lung infections, or (iv) associated cardiac defects. Surgically, rerouted hypoplastic veins may narrow or clot; lobectomy may compromise lung vascular bed; surgical access of abdominal aortic collaterals may be challenging.[1]

Closure of aortic collaterals leads to resolution of pulmonary hypertension and heart failure, reduces pulmonary venous pressures, and improves symptoms.[2] Fick oximetry fails to identify a significant shunt if the inferior vena caval sampling for mixed venous blood includes the pulmonary veins, and the aortopulmonary collaterals perfusing the lungs distally at precapillary level are missed in the main pulmonary artery sample.

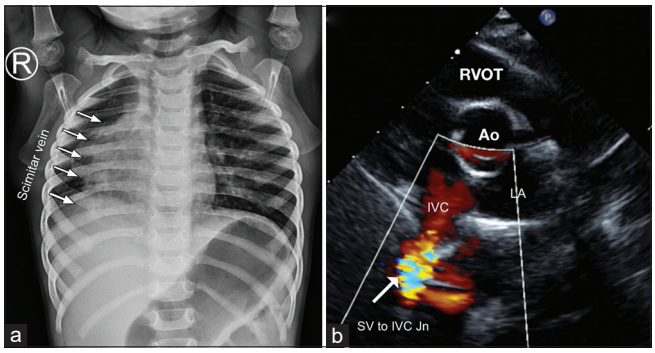

Figure 1: Chest X-ray (a) shows dextroposition of heart, normal cardiothoracic ratio, minimal plethora, and scimitar vein shadow (arrows). Parasternal short axis view of echocardiogram (b) demonstrates a turbulent color flow into the inferior vena cava close to the right atrial junction from the scimitar vein. Ao: Aortic root, LA: Left atrium, RVOT: Right ventricular outflow tract

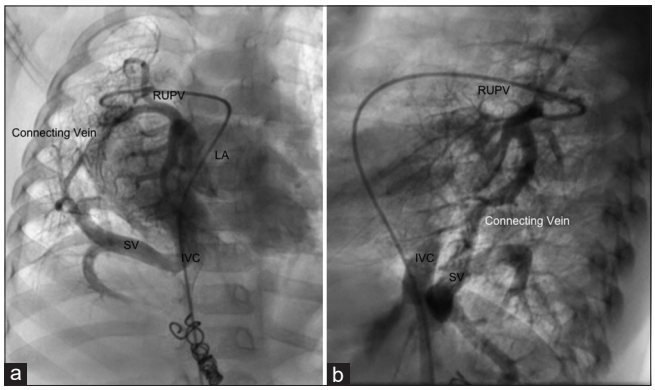

Figure 3: The venous phase of the right pulmonary arteriography in anteroposterior (a) and lateral (b) view demonstrates dual drainage of the right upper pulmonary vein to left atrium and inferior vena cava through scimitar vein and they were communicating through a connecting vein

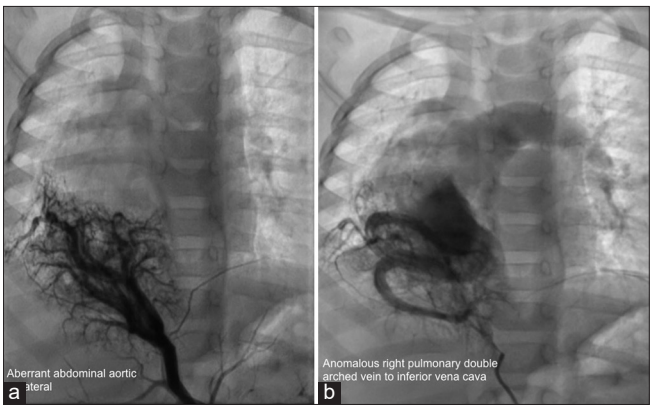

Figure 2: Selective angiogram in the aortic collateral (a) shows large flows into the lower lobe of the right lung. Venous phase (b) demonstrates a double-arched scimitar vein that drains to the inferior vena cava and fills the right cardiac chambers

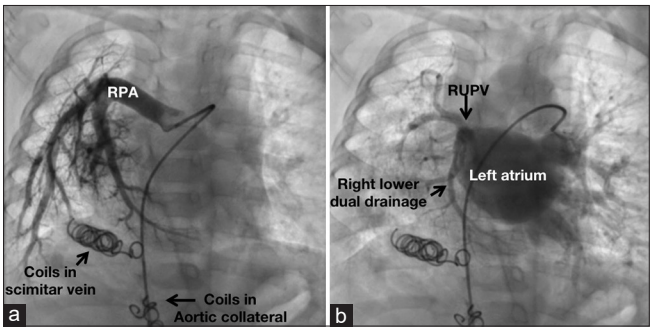

Figure 4: Another right pulmonary arteriogram performed after coil closure of scimitar vein entry shows the right lung vascular distribution in arterial (a) and venous (b) phases. The left atrial filling was complete and unobstructed

This explains the very small left-to-right shunt in our patient. The pulmonary hyperperfusion from aortic collateral may cause segmental hypertension and is unrecognized from central pulmonary artery pressures. Transcatheter closure arrests the torrential blood flow through large abdominal collaterals in Scimitar syndrome.

During embryogenesis, the developing pulmonary venous channels may initially be connected to both inferior vena cava and left atrium.[2] Obliteration of the left atrial connection leads to "classical Scimitar syndrome." Persistence of both the connections results in "Scimitar variant."[2,4] If the inferior vena caval connection gets obliterated in later stages of development, the tortuous double-arched venous channel courses as a "meandering pulmonary vein" toward the left atrium.[5] In the presence of dual drainage, there is preferential flow to the right atrium due to improved compliance of the right ventricle. Closure of the anomalous drainage reroutes the venous return.[4] The right-sided chamber dilatation in asymptomatic adult survivors will form a rationale for closing the anomalous venous channel in the presence of dual drainage.[4]

After closing the anomalous vein drainage with coils, a right pulmonary arteriography demonstrated rerouting of the venous drainage toward the left atrium through the connections to the right upper pulmonary veins. If patients with dual drainage have a stenotic inferior vena caval entry site, the alternative venous pathway to the left atrium progressively enlarges and takes over the right lung drainage, thereby making it a "congenitally palliated scimitar variant."[6] The absence of associated heart defects also facilitated catheter interventions in our patient.

A careful imaging including invasive pulmonary arteriography is warranted in every patient with anomalous pulmonary venous connection to identify dual drainage and adequacy of the two drainage limbs and to decide on a transcatheter solution.[7-10] In patients with dual drainage and a smaller vein connecting to left atrium, a balloon occlusion of the scimitar vein may indicate the tolerance of transcatheter occlusion of the anomalous vein.[10] If anomalous veins are long and tortuous or hypoplastic, transcatheter closure may prove to be a simpler solution than surgical rerouting that carries the risk of pulmonary vein stenosis or occlusion.[10]

CONCLUSIONS

Scimitar variants are associated with dual pulmonary vein drainage to both inferior vena cava and left atrium. Demonstration of a communicating vein between the two could offer a transcatheter solution to arrest the pretricuspid left-to-right shunt by closing the anomalous inferior vena caval entry with devices or coils. Awareness about such dual drainage in many patients may open up more catheter-based solutions.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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