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Images in Congenital Cardiac Disease

"Double whammy": anomalous pulmonary and systemic venous drainage in a patient with scimitar syndrome

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Abstract Anomalously draining right pulmonary veins are expected with scimitar syndrome, but systemic venous abnormalities are rare. We present an unusual case of a female patient with scimitar and an interrupted inferior vena cava.

Keywords: Scimitar syndrome; systemic venous anomaly; pulmonary venous anomaly; adult congenital heart disease

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Scimitar syndrome consists of anomalous drainage of right-sided pulmonary veins into the inferior vena cava, a relatively hypoplastic right lung, and a hypoplastic right pulmonary artery with or without pulmonary sequestration. Abnormal systemic venous drainage is rare.

A 35-year-old female with a history of fatigue and exercise intolerance underwent chest X-ray that showed an abnormal density consistent with scimitar syndrome (Fig 1). Cardiac magnetic resonance demonstrated anomalous right upper and middle pulmonary veins that joined into a common confluence and drained into the right atrium/hepatic vein junction. Caudally, the inferior vena cava was interrupted and continued via the azygous to the right superior vena cava (Fig 2; Supplementary 2b movie). The estimated Qp:Qs by velocity encoded cine was 2:1.

At surgical repair, the site of scimitar confluence drainage was too low to facilitate an intracardiac baffle. The vein was transected, extended via a Gore-Tex tube graft, and sewn into the back wall of the left atrium (Fig 3). The anomalous azygous drainage was untouched. Weeks later, the post-operative cardiac computed tomography illustrated the patency of the graft (Fig 4).

There was one study that examined 122 adult scimitar cases that showed only one patient with

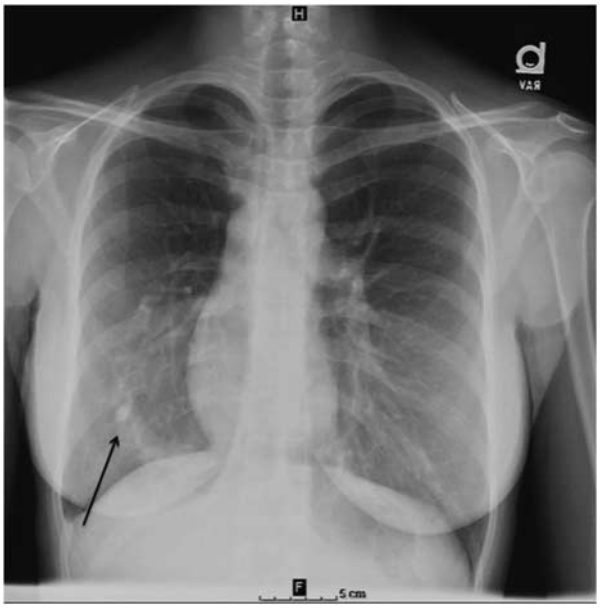

Figure 1.
Initial chest X-ray. Chest X-ray showing the curvilinear density in the lower right chest consistent with a scimitar vein (arrow).

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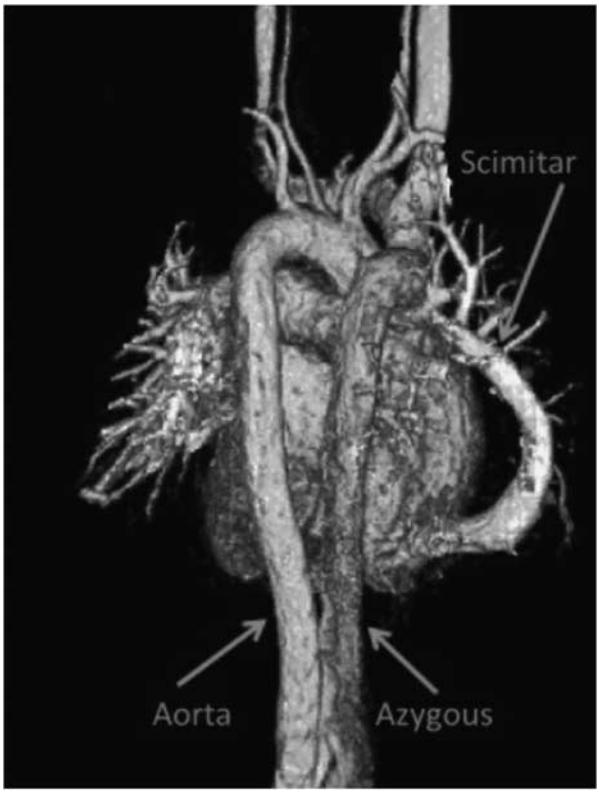

Figure 2.
3D MRI reconstruction. Posterior view of 3D MRI reconstruction of the cardiac anatomy, demonstrating the anomalous drainage of the scimitar vein into the right atrium/hepatic junction. The IVC is interrupted and drains via the posterior azygous. IVC = inferior vena cava; MRI = magnetic resonance imaging.

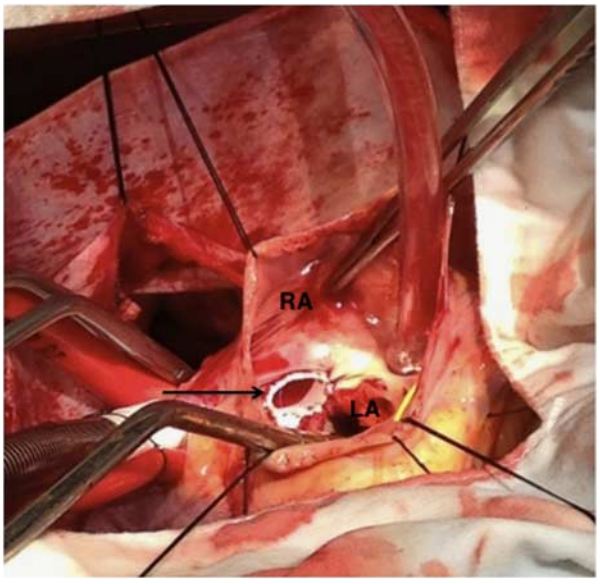

Figure 3.
Intra-operative photo. Intra-operative photo with the atrium opened and the atrial septum removed. The white ring of Gore-Tex is the distal end of the scimitar vein baffle (arrow) entering the left atrium (LA) with the right atrium (RA) above. An ASD patch will then be placed to septate the atrium.

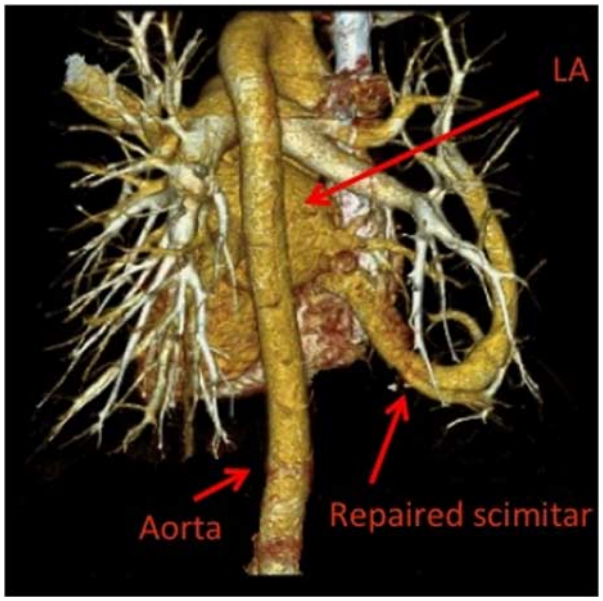

Figure 4.
Post-operative evaluation of repaired scimitar vein. Post-operative 3D computed tomography reconstruction, illustrating the patent pathway of the redirected scimitar vein in to the left atrium (LA).

interrupted inferior vena cava.[1] Rare case reports of scimitar syndrome associated with inferior vena cava interruption suggest that the anomalous pulmonary vein can drain cranially into the hepatic venous confluence, or caudally into the azygous causing significant dilation. Our case represents a rare form of scimitar syndrome with both pulmonary and systemic venous abnormalities.

Supplementary materials

For supplementary material referred to in this article, please visit http://dx.doi.org/10.1017/S1047951113000553

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