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Images in Congenital Cardiac Disease

Evaluation of scimitar syndrome by multislice computed tomography

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Keywords: Pulmonary venous drainage; partial anomaly; imaging modalities

A 12 YEAR-OLD BOY WAS REFERRED FOR INVESTIgation of a cardiac murmur. The electrocardiogram depicted right axis deviation, showing an increased R to S ratio in V4R, with deep S waves in leads V3R through V3. A chest radiography showed the heart to be predominantly located in the right side of the chest, with the cardiac apex directed to the right, and presence of a crescent-like shadow at the lower right lung, the so-called scimitar sign (Fig. 1).

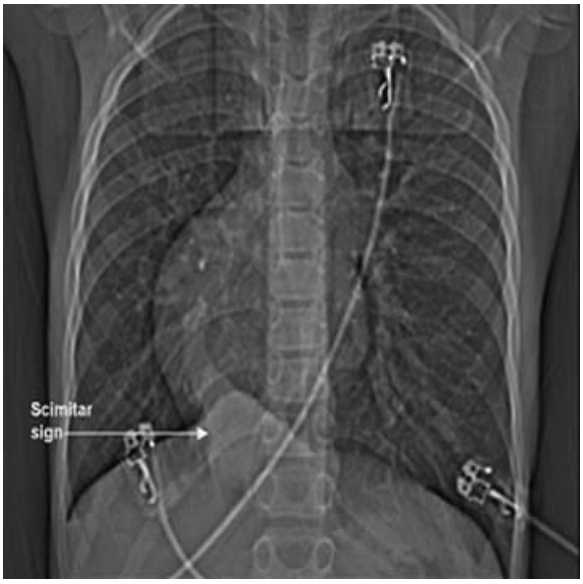

Figure 1.

Echocardiography revealed usual atrial arrangement, enlargement of the chambers of the right heart, and anomalous connection of the right pulmonary veins to the inferior caval vein. Computed tomography confirmed the diagnosis. A venous collector, the scimitar vein, was indentified receiving the right pulmonary veins, and shown to drain into the inferior caval vein just below the diaphragm (Figs. 2, 3). At cardiac catheterization, a significant left-to-right shunt was documented,

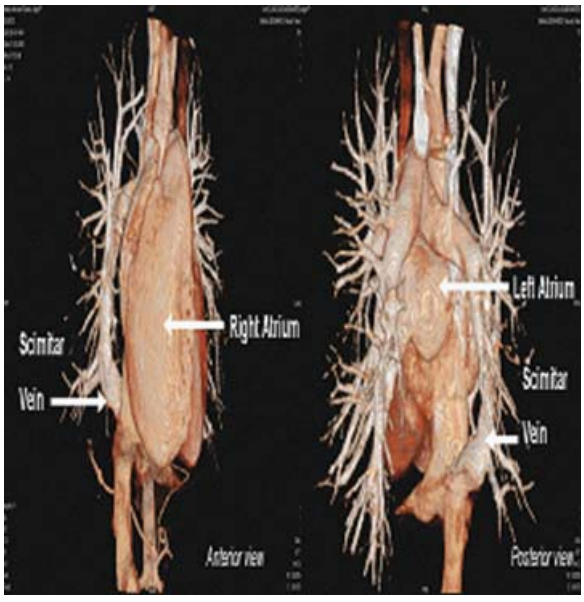

Figure 2.

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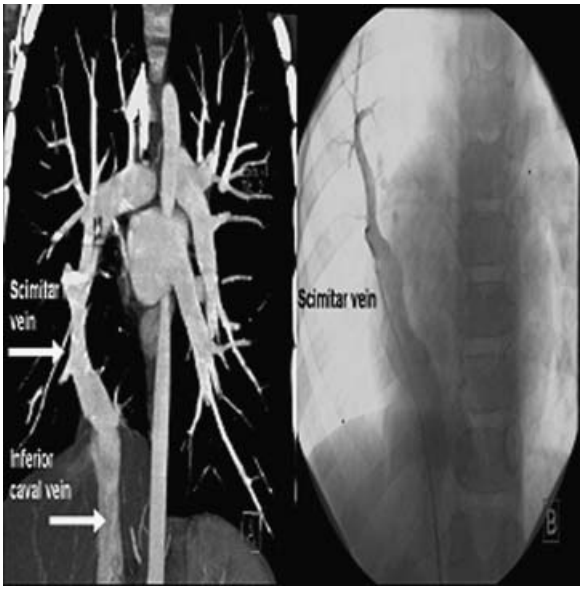

Figure 3.

albeit with normal pulmonary arterial pressures were documented. The patient was submitted to surgical repair, and enjoyed an uneventful recovery.

The scimitar syndrome is a rare malformation involving anomalous drainage of the right pulmonary veins. Chest radiography typically shows an opacity mimicking a scimitar sword. Multiple associated anomalies have been described. Respiratory infections and dyspnoea are frequent in children. Pulmonary hypertension may occur later. Echocardiography usually demonstrates the collector vein, but demonstration of intrapulmonary vessels, aortopulmonary collateral arteries, bronchial morphology, or sequestration may require additional studies.[1]

Cardiac catheterization was performed in our case to calculate shunts and pulmonary vascular resistance, but is unnecessary in most patients. Although magnetic resonance could be used, computed tomography was performed not only to define the exact anatomy of the pulmonary veins, but also to exclude associated pulmonary anomalies, such as hypoplasia or lobar sequestration.

Reference

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