Scimitar Syndrome
Comprehensive, Noninvasive Assessment With Cardiovascular Magnetic Resonance Imaging

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Scimitar syndrome is a rare vascular anomaly whereby a partial anomalous pulmonary venous drainage to the inferior vena cava results in left-to-right shunt. Scimitar syndrome can be associated with congenital cardiovascular defects (dextrocardia, atrial septal defects, and right pulmonary artery hypoplasia), pulmonary anomalies (hypoplasia, sequestration), and tracheobronchial anomalies. Thus, it can manifest with heart failure and recurrent pneumonia.\(^1\),\(^2\) Diagnosis is obtained during childhood, when symptoms related to relevant shunt and other cardiovascular and pulmonary anomalies are present. When asymptomatic, scimitar syndrome can be accidentally discovered in adulthood.\(^3\) Standard chest x-ray evaluation is usually pathognomonic (Figure 1), although it is mandatory that a cross-sectional imaging technique be used to confirm the diagnosis and to obtain comprehensive assessment of any associated anomalies.

Both computed tomography and magnetic resonance imaging (MRI) allow accurate diagnosis. In our experience, MRI is preferred because it allows the patient to avoid exposure to ionizing radiation and it does not require administration of contrast agent. Fast imaging with steady-state precession (TrueFISP) bright blood sequences in standard planes (Figure 2) provides excellent anatomic visualization; cine magnetic resonance sequences (balanced steady-state free precession technique) allow fast and reliable assessment of cardiac function and excellent depiction of associated cardiac anomalies; and flow maps acquired by means of phase-contrast sequences permit the demonstration of flow within the anomalous vessel as well as precise quantification of the left-to-right shunt (Figure 3 and Data Supplement Movie).

In the present case, we obtained chest x-ray images of a 36-year-old woman before she underwent minor maxillofacial surgery (Figure 1). Her past clinical history was unremarkable, and ECG results were normal. On the basis of MRI findings, the final diagnosis of scimitar syndrome was made (Figures 2 and 3 and Movie). The patient was asymptomatic, and the pulmonary-to-systemic blood flow ratio was 1.14; thus surgery was not indicated. Nonetheless, because cine MRI showed slight increases in right ventricle volume (telere-diastolic 111 mL/m^2, telerestolic 60 mL/m^2) the patient will be followed clinically and echocardiographically.

In conclusion, we think cardiovascular MRI without administration of contrast medium is the preferred technique for diagnosing scimitar syndrome, as it allows for comprehensive assessment of the anomalous vessel and heart and thus provides sufficient information for appropriate surgical or conservative management.

Figure 1. A 36-year-old, healthy woman with an unremarkable past clinical history and normal ECG. Chest x-ray imaging, performed before minor maxillofacial surgery, shows linear opacity in the right lung (arrows) consistent with a vascular structure (a scimitar sign).
Disclosures

None.

References


Figure 2. Multislice, single-phase, fast imaging with steady-state precession (TrueFISP) sequence on coronal (A and B) and axial (C and D) planes shows an anomalous vessel in the right lung (arrows) connected to the inferior vena cava.

Figure 3. Phase-contrast sequence (A) shows that flow within the anomalous vessel (arrow) has the same craniocaudal direction as flow within descending aorta (arrowhead). Analysis of the flow curve in the anomalous vessel (B). Values are negative because flow was encoded in the caudocranial direction (peak velocity 42 cm/s, average flow 1.3 L/min).
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