

Scimitar syndrome: imaging by telecardiography, heart catheterization and angiography

Scimitar sendromu: Telekardiyografi, kalp kateterizasyonu ve anjiyografi ile görüntüleme

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Scimitar syndrome is a rare congenital anomaly. This syndrome is characterized by partial or complete anomalous pulmonary venous drainage of the right lung to the inferior vena cava (1,2). On the chest radiograph, the vein produces a vascular shadow to the right of the heart that descends toward the diaphragm, resembling a scimitar, which is a short curved Turkish sword. Scimitar syndrome is often associated with hypoplasia of the right lung, anomalous systemic vascular supply of the right lung from the aorta, dextrocardia and bronchial anomalies. A 2-year-old girl presented with a non-resolving right lower lobe infiltrate.

The chest radiograph showed a typical scimitar-shaped shadow in the right lung (Fig. 1).

Cardiac catheterization confirmed that nearly all venous return from the right lung was directed via the curved scimitar vein to the inferior vena cava just above the diaphragm (Fig. 2,3), the left pulmonary veins drained normally into the left atrium and atrial septal defect. Although the classic components of the scimitar syndrome are abnormal venous drainage of the affected lung, dextrocardia, and systemic arterial supply to the affected lung, there is only an anomalous connection of the pulmonary vein with



Figure 1. Chest radiograph shows abnormal draining vein overlying the right lower lobe

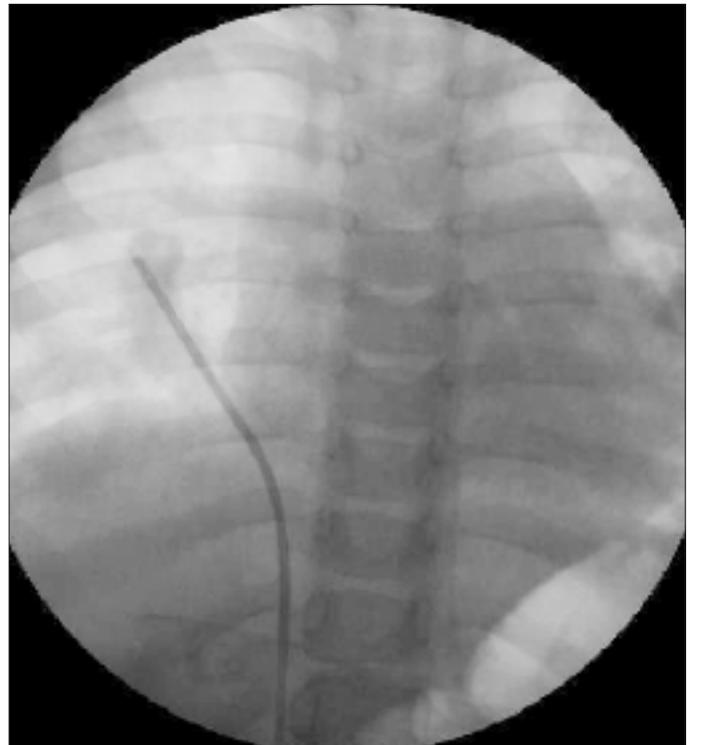


Figure 2. Cine image demonstrates that catheter is directed via the inferior vena cava to the scimitar vein just above the diaphragm



Figure 3. Re-circulation phase of the right pulmonary artery angiogram demonstrating anomalous pulmonary venous drainage to the inferior vena cava

the inferior vena cava without additional anomalies except atrial septal defect in our case.

The scimitar syndrome should be considered in the presence of an atypical right paracardial shadow on the postero-anterior view of chest radiography.

References

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