Scimitar Syndrome with a Rare Systemic Venous Anomaly Association: Original Image

Nadir Bir Sistemik Venöz Anomali Birlikteliğinde Scimitar Sendromu

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Geliş Tarihi/*Received:* 30.12.2014
Kabul Tarihi/*Accepted:* 01.03.2015

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Key Words:
Scimitar syndrome; vena cava, inferior; multidetector computed tomography

Anahtar Kelimeler:
Scimitar sendromu; vena kava, inferior; çok kesitli bilgisayarlı tomografi

Turkiye Klinikleri J Cardiovasc Sci 2015;27(2):73-5

doi: 10.5336/cardiosci.2014-43221

The Scimitar syndrome is an uncommon and congenital abnormality of the cardiopulmonary system. In the Scimitar syndrome, blood returning to the inferior vena cava (IVC) from the right lung through the pulmonary vein is directed to an area of the immediately above or below the hemidiaphragm.[1,2] Scimitar syndrome associated with the interruption of the IVC is extremely unusual.[3] The incidence of IVC interruption alone is 0.3%; however, for individuals with congenital heart diseases, this incidence may be as high as 2%.[3]

Radiographically, the appearance of Scimitar syndrome is often characteristic. The syndrome may be diagnosed using ultrasonography, magnetic resonance imaging and computed tomography (CT).[4] We presented a patient who was diagnosed with Scimitar syndrome based on different radiological images. In conclusion, this unusual case highlights that the multidetector CT (MDCT) can be an adequate tool for the clarification of complex thoracic venous anomalies.

A one-year-old girl was referred to our clinic due to repeated infections affecting her respiratory tract. The patient's chest radiograph revealed the presence of dextrocardia and right lung hypoplasia, along with compensatory emphysema in the left lung (Figure 1A). Echocardiography indicated a right partial abnormal pulmonary venous connection anomaly, a low venosum type defect in the atrial septum, and a perimembranous type defect in the ventricular septum. The catheter angiography revealed that the IVC was interrupted, and that the interruption continued into the azygos and hemiazygos veins. A Scimitar vein was identified, linking to the IVC's hepatic area (Figure 1B). Engorged hemiazygos vein connected to the azygos vein with multiple venous collaterals at the thoracic vertebral level, and finally drained into the superior vena cava (Figure 1C). The patient un-

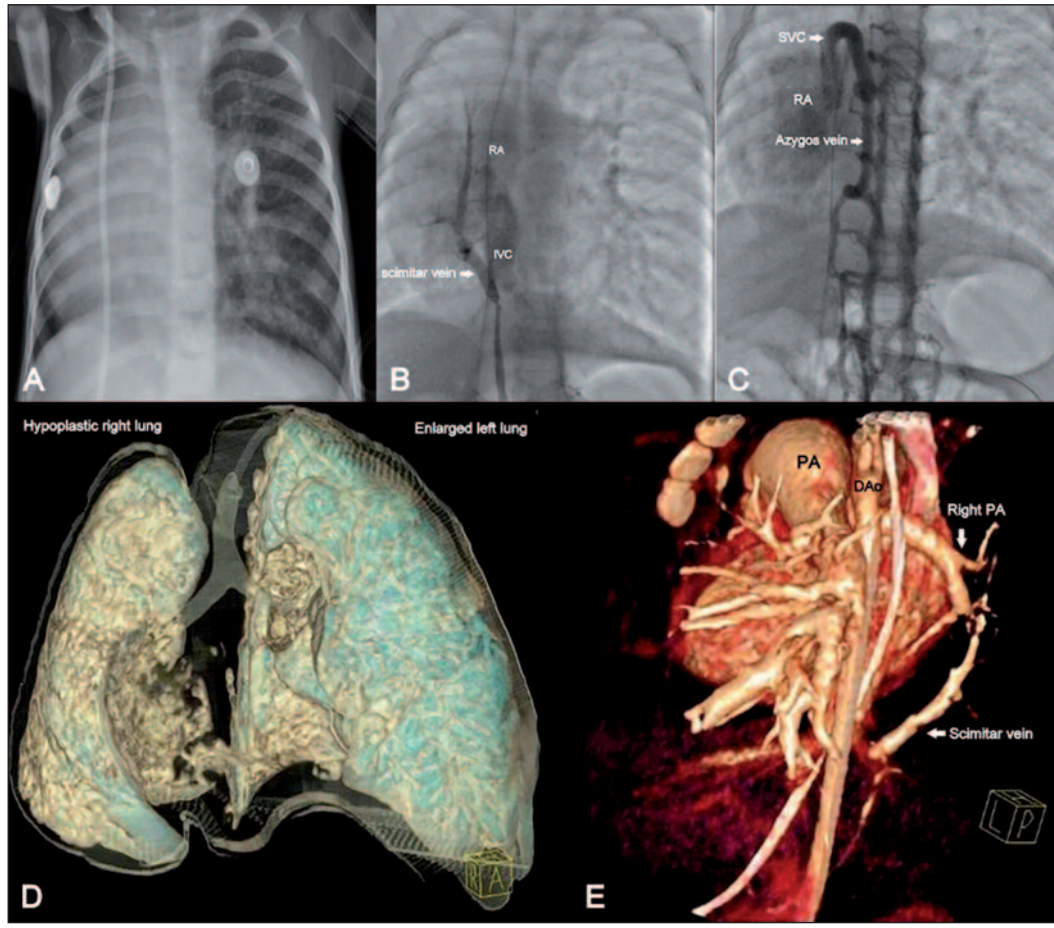

FIGURE 1A-E: Evaluation of radiological images showed that the right lung was small in size with compensatory emphysema of the left lung and the filling of the anomalous Scimitar vein and systemic venous structures.

IVC: Inferior vena cava; SVC: Superior vena cava; PA: Pulmonary artery; RA: Right atrium; DAo: Descending aorta.

derwent examination with the MDCT display, which identified a mild hypoplasia in the right lower lung (Figure 1D), and a Scimitar vein draining into the suprahepatic IVC (Figure 1E). The patient was scheduled for elective surgery following the treatment of her infection.

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