Scimitar Syndrome

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A 38-year-old woman presented with dyspnea and fatigue for 1 week. Physical examination was unremarkable. Laboratory tests were unrevealing. Chest X-ray showed dextrocardia and a right lung mass measuring 5×4 cm (Fig. 1). Computed tomography (CT) of the chest revealed an accessory lobe of the liver extending into the chest cavity through a defect in the posterior right hemi-diaphragm, and the right pulmonary vein draining into the infradiaphragmatic inferior vena cava (IVC) instead of the left atrium (Fig. 2), consistent with Scimitar syndrome.

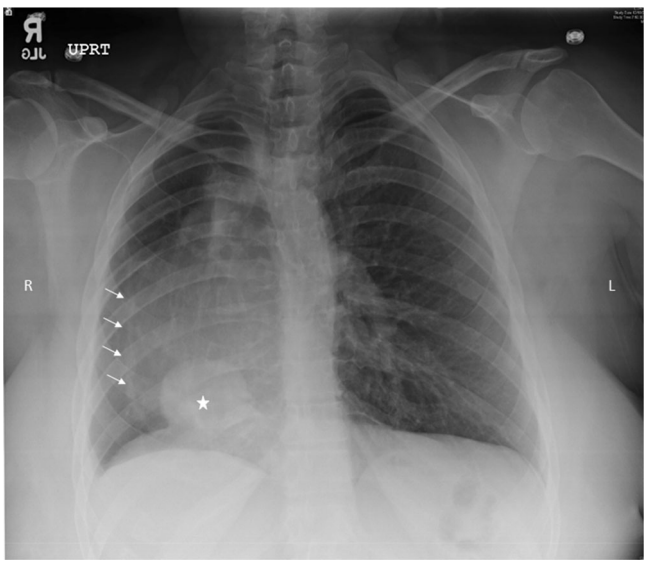

Figure 1 Chest x-ray showing the scimitar sign (arrows), dextrocardia, hypoplastic right lung and well-circumscribed mass at the right lower lung base (star).

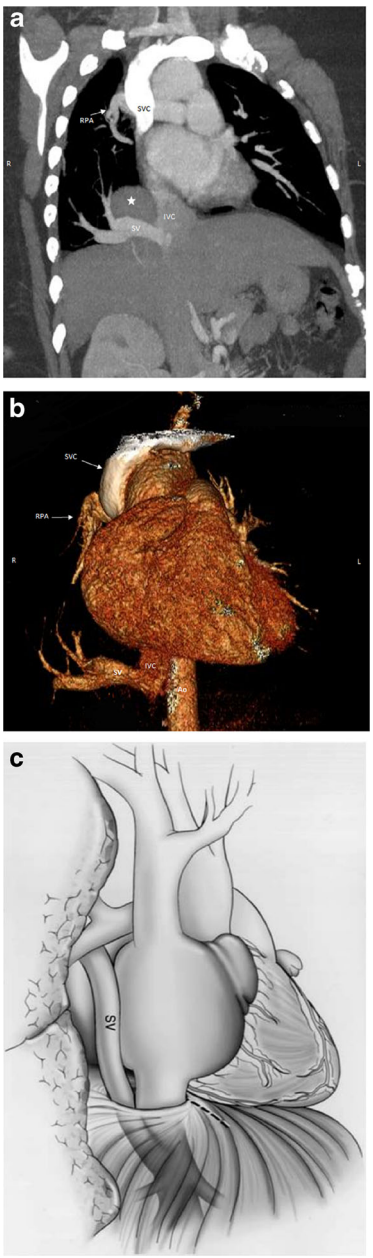

Figure 2 Panel a Chest CT scan with contrast in coronal section demonstrates the scimitar vein (SV), a large anomalous right pulmonary vein coursing into the infradiaphragmatic inferior vena cava (IVC). An accessory liver lobe is noted at the base of right hemithorax (star). Panel b 3 D volume rendered CT scan image shows the scimitar vein draining into the inferior vena cava. Panel c Posterior view of heart with scimitar vain (SV) draining below the diaphragm. IVC; inferior vena cava, SVC; superior vena cava, SV; scimitar vein, RPA; right pulmonary artery, Ao; aorta.

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Scimitar, venolobar, or hypogenetic lung syndrome is a rare congenital anomaly of pulmonary venous return,[1] in which the right pulmonary vein drains anomalously into the IVC. The curved course of the anomalous right pulmonary vein gives the characteristic appearance of a Turkish sword "scimitar" on chest X-ray. A variety of congenital defects are commonly associated with this specific type of anomalous pulmonary venous return.[2] Infants usually present with failure to thrive, pulmonary hypertension or heart failure symptoms. Adult patients typically remain asymptomatic and are incidentally discovered on routine chest radiograph. Some adult patients may present with symptoms of fatigue, dyspnea, or recurrent pneumonia. However, most patients with isolated Scimitar syndrome have excellent prognosis.[3]

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Conflict of Interest: *The authors declare that they do not have a conflict of interest.*

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