

Adult-form scimitar syndrome in a 64-year old woman

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A 64-year old female, current smoker of approximately 44 pack-years, was referred to us due to fever, dry cough, and shortness of breath on exertion. Physical examination revealed fever, tachypnea, tachycardia, and an oxygen saturation of 90% on room air (SpO₂: 90%, FiO₂: 21%). Laboratory evaluation showed a mild elevation of the white blood cell count (14.000/mm³) and C-reactive protein (9mg/dl). A chest radiograph demonstrated consolidation of the left lung base, as well as an anomalous pulmonary vein on the right side (Figure 1). Based on these radiographic findings, the Scimitar syndrome was suspected. The patient underwent a CT-angiography (CTPA) in order to confirm the suggested diagnosis, which demonstrated a right-sided volume loss of the lung parenchyma, a shift of the heart and mediastinum to the right, and also anomalous ejection of the right lower pulmonary vein (which was dilated, Figure 2) to inferior vena cava. Increased cardiothoracic index and severe dilation of the azygos vein terminating in the superior vena cava were also noted.

Scimitar syndrome is a rare congenital malformation of the right lung characterized by an abnormal right sided pulmonary drainage into the inferior vena cava, hypoplasia of the right lung, with cardiac malformation (dextroposition of the heart or dextrocardia) as well as abnormal artery supply¹. It usually presents as recurrent lung infection, exertional dyspnea, whereas pulmonary artery hypertension and hemoptysis are uncommon symptoms of the syndrome in adult life^{2,3}. Symptomatology in infants includes cyanosis, pulmonary artery hypertension, and cardiac defects often requiring surgical intervention with a rather high mortality risk¹. Diagnosis of Scimitar syndrome is confirmed with chest radiography, CT-angiography and echocardiography.

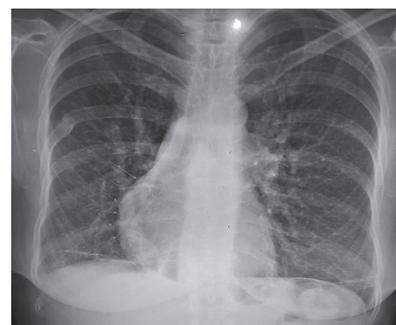


FIGURE 1. Curvilinear pattern created in the chest radiograph by the right pulmonary vein that drains to the inferior vena cava. This radiographic density often has the shape of a scimitar (scimitar sign, red arrows), a type of curved blade sword.

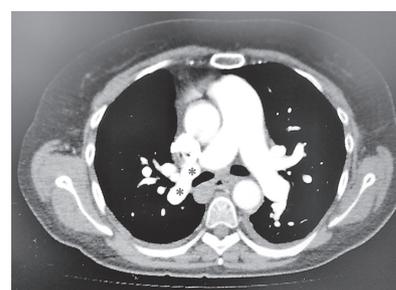


FIGURE 2. CT thorax showing right lung hypoplasia and the dilated and anomalous right pulmonary vein (red asterisks).

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