

Scimitar Sign

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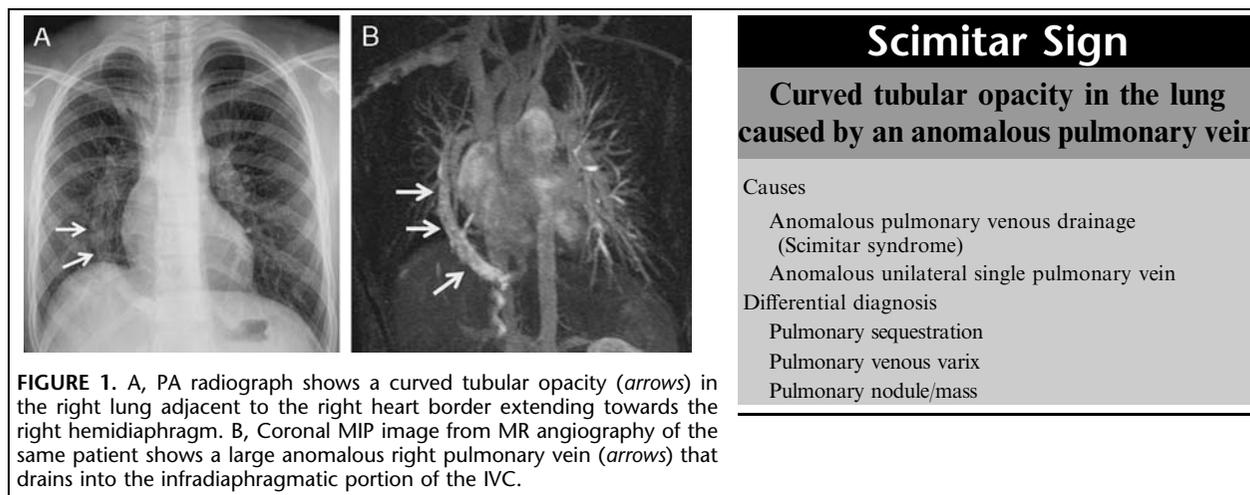


FIGURE 1. A, PA radiograph shows a curved tubular opacity (arrows) in the right lung adjacent to the right heart border extending towards the right hemidiaphragm. B, Coronal MIP image from MR angiography of the same patient shows a large anomalous right pulmonary vein (arrows) that drains into the infradiaphragmatic portion of the IVC.

Appearance: The scimitar sign refers to a curved, tubular opacity adjacent to the right of the heart and extending toward the diaphragm seen on a frontal radiograph. Scimitar refers to a Turkish/Persian sword that has a curved blade.

Explanation: The scimitar sign has classically been described as a pathognomonic radiographic finding in scimitar syndrome (also hypogenetic lung or venolobar syndrome), which is characterized by anomalous pulmonary vein, pulmonary hypoplasia, cardiac dextroposition, pulmonary artery hypoplasia, and systemic arterial supply to the lung. Occasionally, not all these features are present in an individual patient. The scimitar sign is caused by the anomalous pulmonary vein, which drains all or most of the right lung and runs adjacent to the right heart border, from the level of the hilum to the diaphragm, progressively widening (Fig. 1A). The vein usually traverses the diaphragm and drains into the IVC, but may also drain into a hepatic or portal vein or the right atrium.¹ (All references cited in this article can be found at <http://links.lww.com/JTI/A45>.)

However, the scimitar sign can also occur with another entity, the anomalous single pulmonary vein (ASPV), in which the entire lung is drained by a single pulmonary vein with an anomalous course, but draining normally into the left atrium.² ASPV has been described by several terms, including anomalous unilateral single pulmonary vein, meandering pulmonary vein, scimitar variant, pseudo-scimitar syndrome, or pulmonary varix, all of which have only minor variations in their respective imaging features. One study determined that 14 of the 32 total reported cases of ASPV presented with a scimitar sign on chest radiography.³

Discussion: Distinguishing scimitar syndrome from an ASPV is clinically important, since the anomalous venous drainage in scimitar syndrome results in a left-to-right shunt, which when large or if associated with pulmonary hypertension, may require treatment with surgery or embolization. ASPV is not associated with a shunt and, hence, does not require further invasive investigation or management.

Both these entities are almost exclusively seen in the right lung. Only 2 cases of scimitar syndrome and 8 cases of ASPV have been reported on the left, and 2 reported cases of ASPV have been bilateral.³ Both are associated with right lung hypoplasia, cardiac dextroposition and systemic arterial supply. Anomalies of bronchial and pulmonary arterial branching, lobes and fissures are commonly seen. Horse-shoe or cross-over lung has been reported in both entities. Atrial septal defect and other congenital defects may also be associated with scimitar syndrome.¹ Only a few cases have been reported where both of these entities coexist.^{4,5} Occasionally, ASPV may also have branches that drain into the IVC.³ Due to the similarities in these entities, it has been speculated that they result from a similar embryological defect. While scimitar syndrome is believed to be caused by failure of integration of primitive pulmonary venous plexus with the common pulmonary vein, resulting in communication with cardinal or umbilical veins, ASPV is believed to be due to hypoplasia or atresia of one of the pulmonary veins before pulmonary segmentation. As a result, the drainage of the entire lung occurs through the remaining vein.³ An alternative explanation is that a scimitar vein has a dual embryologic connection to the IVC and the left atrium. Obliteration of the connection to the left atrium results in scimitar syndrome, while complete/partial obliteration of the connection to the IVC results in ASPV.⁶

CT or MR angiography is used for characterizing and distinguishing these anomalies (Fig. 1B). MRI also aids therapy by quantifying the amount of shunting through the anomalous vein and associated defects. The differential diagnosis for a scimitar sign includes a pulmonary sequestration, pulmonary venous varix, and a pulmonary nodule/mass.

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