

The phantom of the chest

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Answer 4: Scimitar vein

Scimitar syndrome is a rare congenital anomaly characterized by the triad of anomalous pulmonary venous drainage, hypoplastic right lung and dextroposition of the heart (1). However Scimitar vein can be seen in isolation. The anomalous vein usually drains into the upper part of the inferior vena cava (IVC) and rarely into the right atrium or portal vein. Hemodynamically, there is an acyanotic left to right shunt. The lung is frequently perfused by the aorta, but the bronchial tree is still connected and thus the lung is not sequestered. It can sometimes be detected in chest X-ray in asymptomatic patients. The Scimitar vein is visible as a curvilinear density along the right heart border, resembling the curved Turkish sword that gives the condition its name (2). About 70% of the patients with Scimitar syndrome have some cardiac anomaly, including atrial or ventricular septal defect, coarctation of the aorta, hypoplastic left heart, total anomalous pulmonary venous connection and patent ductus arteriosus (3). In symptomatic patients, surgical intervention is usually required, but in asymptomatic patients, close follow-up with echocardiography in terms of right heart chamber dilatation seems to be enough.

In this case, the vascular structure spotted in dynamic perfusion and functional cardiac and thorax MRI was interpreted as Scimitar vein, 1.5 cm in diameter and draining mid and lower lobes of the right lung into the suprahepatic segment of the IVC in the basal portion of the right lung (Fig. 4, 5). Besides, other cardiac anomalies like atrial or ventricular septal defect were not seen. Two-left pulmonary veins and one right pulmonary vein draining into the left atrium were seen. No further tests or intervention was planned because the patient was asymptomatic, the right heart chambers were dilated only slightly and the PAP was normal. Further, she was allowed to become pregnant.

Vascular connections can be similar in pulmonary sequestration and Scimitar syndrome. Sequestration is associated with focally increased lung echogenicity and often leads to an abnormal heart position (contralateral mediastinal shift); these findings are absent in Scimitar syndrome/vein. In our patient, no pulmonary pathology can be seen in the chest X-ray; besides, in MRI, abnormality is seen in the venous system and not the arterial system.

The features of chest X-ray in patients with pulmonary hypertension include dilation of the descending branch of the main pulmonary artery, oligemic upper lung fields and dilation of the right heart chambers. In our patient, although slight dilation of the right heart chambers is present, sPAP measured by TTE is within normal range and the chest X-ray features described above are not present.

Palla's sign, seen in chest X-ray of patients with pulmonary thromboembolism, is described as enlargement of the right descending pulmonary artery; it is seen as a solid, sausage-like structure extending from the hilus to the periphery. This condition, which usually develops in patients with risk factors such as

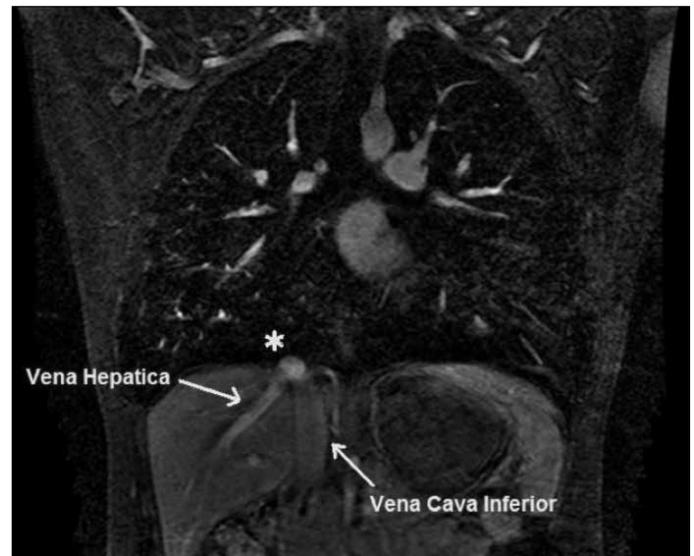


Figure 4. MRI in coronal section shows the Scimitar vein draining into the hepatic vein (asterisk) in the suprahepatic segment and its association with the hepatic vein

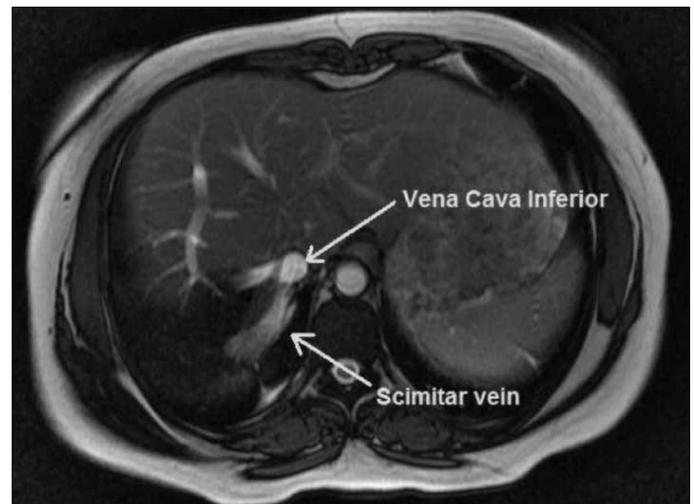


Figure 5. MRI in horizontal section shows the course of the Scimitar vein and its drainage into the inferior vena cava

oral contraceptive use, long distance travel or presence of a malignancy, was not considered in our patient, who wants to become pregnant, on clinical or radiographic grounds.

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References

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2. Halasz NA, Halloran KH, Liebow AA. Bronchial and arterial anomalies with drainage of the right lung into the inferior vena cava. *Circulation* 1956; 14: 826-46. [\[CrossRef\]](#)
3. Najm HK, Williams WG, Coles JG, Rebeyka IM, Freedom RM. Scimitar syndrome: twenty years' experience and results of repair. *J Thorac Cardiovasc Surg* 1996; 112: 1161-8. [\[CrossRef\]](#)