

The sword and the shunt: Scimitar syndrome

Sir,

A 50-year-old female was presented to the emergency department with worsening shortness of breath for the last 5 days. Medical history was unremarkable other than hypertension and obstructive sleep apnea. Chest radiography [Figure 1] revealed elevated right hemidiaphragm with a mass-like opacity in the right lower zone obscuring cardiac silhouette. Furthermore, a vertically oriented linear opacity in the right lower zone was noted (arrow).

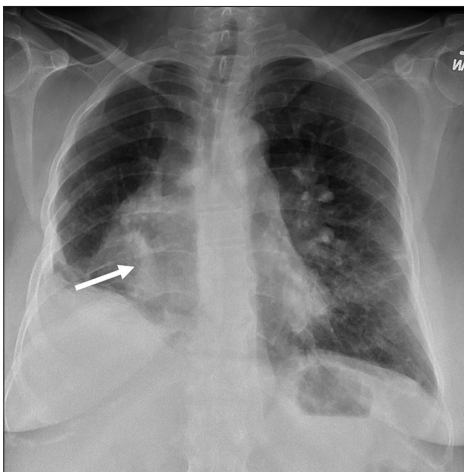


Figure 1: Posteroanterior chest radiograph demonstrates elevated right hemidiaphragm, mass-like opacity in the right lower zone obscuring cardiac silhouette. Furthermore, a vertically oriented linear opacity in the right lower zone was noted (arrow)

the chest demonstrated that right-sided pulmonary veins were draining in a vertical vein which emptying into inferior vena cava (IVC) [Figures 2 and 3]. Right-sided cardiac chambers and main pulmonary artery were dilated. Hypoplastic right lung and elevated right hemidiaphragm were also present. The constellation of the findings is consistent with Scimitar syndrome. The patient underwent a right heart catheterization that showed a significant left-to-right shunt with a Qp/Qs ratio of 1.8. Nonemergent surgical repair was planned.

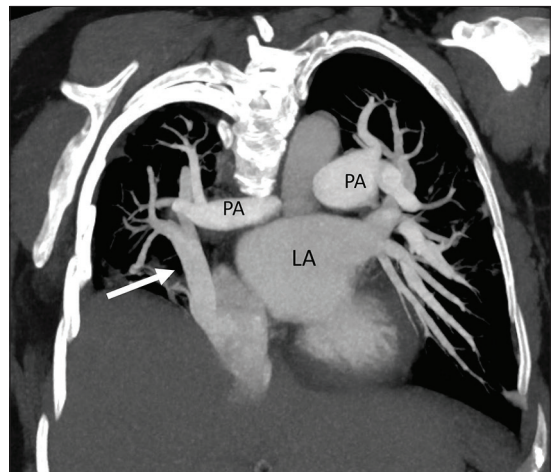


Figure 2: Coronal oblique maximum intensity projection computed tomography angiography image demonstrates that all of the right-sided pulmonary veins are draining into the inferior vena cava via a vertical vein (arrow), creating a left-to-right shunt. Note the normal communication between left atrium and left-sided pulmonary veins

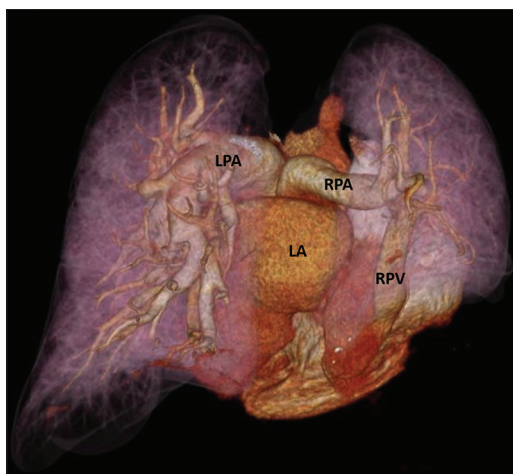


Figure 3: 3D volume-rendered computed tomography angiography image (posterior view) shows the absence of communication between left atrium and right-sided pulmonary veins. The right-sided pulmonary veins are emptying into the inferior vena cava. Also note the hypoplastic right lung. LA: Left atrium, LPA: Left pulmonary artery, RPA: Right pulmonary artery, RPV: Right pulmonary veins

Scimitar syndrome, also known as hypogenetic lung syndrome, is a congenital partial anomalous pulmonary venous return, which results in left-to-right shunt and eventually pulmonary hypertension and right heart failure. Most commonly, the pulmonary veins of the right lower lobe empty in a vertical vein which drains into the IVC. Although the anomalous vein might be seen on radiography as a tubular opacity parallel to the right heart border creating a shape of Turkish sword (scimitar), CTA provides accurate identification of the pulmonary venous anatomy and draining vein that is crucial for the planning of surgical treatment.^[1,2]

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Conflicts of interest

There are no conflicts of interest.

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