

SCIMITAR SYNDROME: A CASE REPORT

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ABSTRACT

Scimitar syndrome is rare congenital disorder, characterized by an abnormal rightsided pulmonary venous drainage in the inferior vena cava.

We report the case of a 9-month-old female infant, in whom the diagnosis was suspected on a chest radiograph and the definitive diagnosis was confirmed by a CT study

Keywords: Scimitar, abnormal pulmonary venous return, CT

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CASE HISTORY

A 9-month-old female infant reported with acute dyspnea without fever or cough. On examination, her pulse was regular (140 beats/minute) with 50 breaths/minute.

Respiratory system examination revealed crepitanes on the right side, chest x-ray showed a cardiomegaly with a right basal opacity (**figure 1**)



Figure 1: Chest radiograph showing cardiomegaly, right basal opacity (arrow)

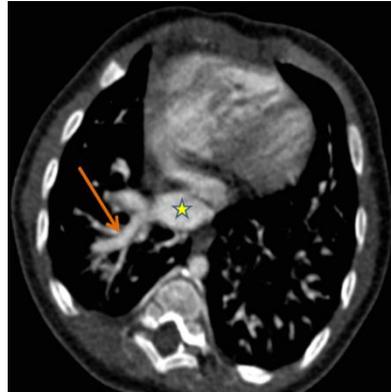


Figure 2: CT thorax showing lower part of the aberrant venous drainage (arrow) to inferior vena cava (*)

Coronal and 3D reconstruction of the CT study (**figure3**) showed typical scimitar-shaped structure running from the middle of the right lung toward diaphragm.

Take into consideration tomography's results, the diagnosis of scimitar syndrome was made and the treatment has been started.

CT angiogram showed an aberrant right venous drainage to inferior vena cava (**figure2**), Left pulmonary vein ending into left atrium is well visualized and the corresponding pulmonary vein on the right side is absent.

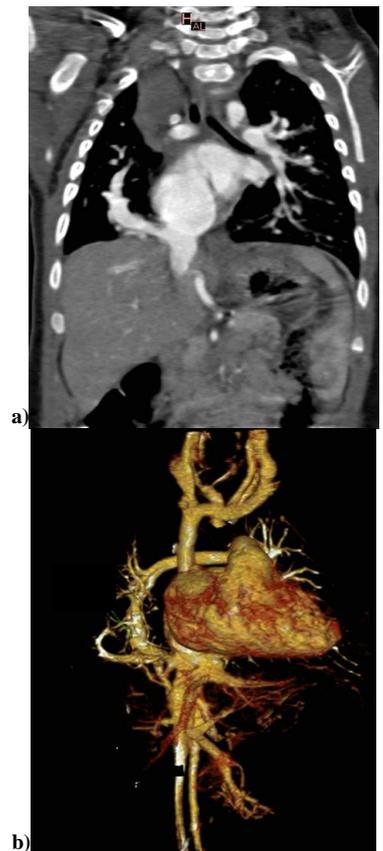


Figure 3: Coronal (a) and 3D reconstruction (b) showing a typical scimitar-shaped structure

DISCUSSION

Scimitar syndrome is a disease characterized by a right abnormal pulmonary venous return located most often in the inferior vena cava where the aberrant pulmonary vein describes a path realizing the appearance in Turkish sword known as "scimitar" (Figure 4).



Figure 4 : scimitar

It is essentially a complex combination of lung malformations and partial anomalous pulmonary venous return also grouped under the term Halasz syndrome known for more than a century, it includes hypoplasia of the right lung, dextroposition of the heart, hypoplasia of the right

pulmonary artery (RPA), and anomalous systemic arterial supply from the aorta to the right lung [1,2] This syndrome predominates in the female sex and its prevalence is between 1/100 000 and 1/33 000 live births. Clinically, this syndrome is manifested by dyspnea, asthenia or repeated pulmonary infections, and sometimes massive hemoptysis favored by the systemic vascularization of the sequestered lung [3]. This is most commonly caused by pulmonary hypertension due to cardiac and/or right lung anomalies.

The association of an abnormal pulmonary venous return to pulmonary sequestration is described in 50% of patients with this syndrome [4]. The aberrant systemic artery usually originates from the lower part of the descending thoracic aorta or the initial part of the abdominal aorta [5]

In our case, the aberrant systemic artery originates from the celiac trunk feeding a sequestration of the right pulmonary base (Pryce I type) Figure 5

The treatment is surgical and outcome is dependent on the nature and severity of the anomalies [6]. It's based on a reimplantation of the pulmonary vein in the left atrium associated in some cases with a surgery of the pulmonary sequestration. Heart failure may also be caused because of a large arterial supply from the abdominal aorta to a sequestered lobe. In these situations, cardiac catheterization may be used to embolise the aberrant pulmonary blood supply.

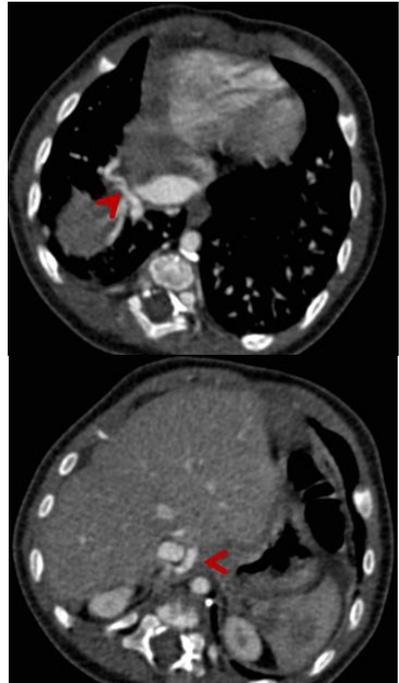


Figure 5: the aberrant systemic artery (head of arrow) originates from the celiac trunk

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