

Case Report

Scimitar Sign and Scimitar Syndrome

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Introduction

Scimitar syndrome is a rare congenital cardiovascular anomaly with an incidence of 1-3 per 1,00,000 live births. The radiological description "Scimitar sign" may occur in isolation which is more frequently encountered in clinical practice. It can also be part of the full blown Scimitar syndrome wherein other congenital anomalies are also present. This case report is intended to highlight Scimitar syndrome with particular emphasis on the radiological characterization of the individual components of the syndrome.

Key words: Scimitar syndrome, Scimitar sign

Case report

A 35 year old lady presented to our outpatient clinic with symptoms suggestive of asthma. Clinical examination of the respiratory system revealed the presence of polyphonic wheezes over both lung fields. On routine evaluation she was found to have an abnormal chest radiograph (Fig 1). There was evidence of volume loss in the right hemithorax with ipsilateral shift of mediastinum and a linear opacity parallel to the right heart border. With a suspicion of Scimitar sign, she was further evaluated with a contrast enhanced computed tomogram of the chest which revealed the presence of the following abnormalities - hypoplasia of the right lung, dextroposition of the heart, hypoplastic right descending pulmonary artery and an enlarged and elongated anomalous pulmonary vein having tributaries from the hypoplastic right upper lobe, middle lobe, lower lobe and the right inferior pulmonary vein and finally joining the inferior vena cava at the diaphragmatic hiatus level. (Fig 2). This was better delineated by the technique of Maximum Intensity Projection, which is a method of three dimensional reconstruction using the computed tomogram images. (Fig 3). The left pulmonary artery is of normal caliber (Fig 4) whereas the right

pulmonary artery is hypoplastic (Fig 5). These constellation of radiological signs are consistent with a diagnosis of Scimitar syndrome, a rare but well known congenital cardiovascular anomaly.

Discussion

The radiological sign, coined by Naill in 1960, is called Scimitar sign because the characteristically curved anomalous right pulmonary vein that drains into the inferior vena cava resembles the curved Middle Eastern (Turkish) sword called "Scimitar". A variety of congenital thoracic abnormalities are associated with this specific type of partial anomalous pulmonary venous return first described by Cooper in 1836^{1,2}. Associated anomalies are variable and include 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 lung³. This rare anomaly has an incidence of approximately 1 to 3 per 100,000 live births⁴; the true incidence may be higher because many patients are asymptomatic.

Scimitar syndrome overlaps with pulmonary sequestration and the term "venolobar syndrome" has been



Fig 1: Chest X ray PA view depicting the anomalous vein (Scimitar sign) and hypoplastic right lung



Fig 2: Contrast enhanced CT scan clearly delineating the anomalous course of the inferior pulmonary vein



Fig 3: Maximum intensity projection image which gives a three dimensional view of the anomalous course of the inferior pulmonary vein



Fig 4: Left pulmonary artery which is normal in caliber

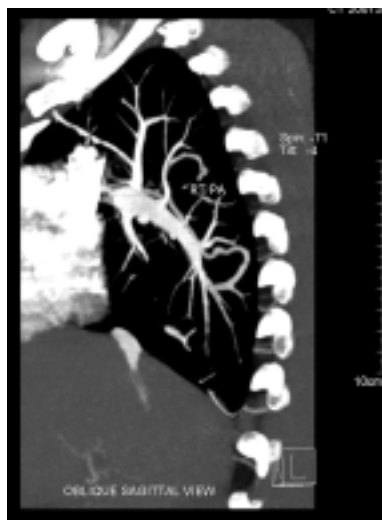


Fig 5: Hypoplastic right pulmonary artery

coined to include these associated pulmonary and vascular malformations⁵. This condition is associated with:

1. Partial agenesis or hypoplasia of the right lung with bronchial isomerism.
2. Diverticulum or hypoplasia of the right bronchial system.
3. Hypoplasia or agenesis of the right pulmonary artery. This may cause mediastinal shift to the right side and Scimitar vein may be difficult to appreciate or even completely obscured.
4. Abnormal systemic blood supply to at least part of the right lung, most frequently the posterior basal segment of the lower lobe, usually arising from the infra diaphragmatic descending aorta.
5. Dextroposition of the heart due to right lung hypoplasia with mediastinal shift.
6. Accessory diaphragm, eventration or partial absence of the diaphragm.
7. Phrenic cyst.
8. Horseshoe lung.
9. Esophageal and gastric lung.
10. Absence of the pericardium.
11. Other congenital cardiac malformations (25% of cases) including ASD, ventricular septal defect, coarctation of the aorta, tetralogy of Fallot, pulmonary stenosis, absent inferior vena cava with azygous continuation to superior venacava

Three-dimensional computed tomography (CT) and cardiac-gated magnetic resonance imaging (MRI) are useful in visualizing the anomalous pulmonary vein. Scimitar syndrome has been reported most widely in adults and older children and is usually found during a workup for dyspnea, fatigue, recurrent respiratory infections, or as an incidental finding on a routine chest radiograph⁴. This adult form of Scimitar syndrome usually is not associated with pulmonary hypertension and typically has mild symptoms and a benign prognosis.

Most patients require only medical follow up. The indications for surgical repair include the presence of Scimitar syndrome, especially in association with ASD, pulmonary hypertension, or stenosis of the anomalous vein. Several methods with cardiopulmonary bypass have been recommended to repair this anomaly, including direct anastomosis of the Scimitar vein to the left atrium, or division with reimplantation of the anomalous pulmonary vein into the right atrium with baffle insertion to redirect the flow into the left atrium⁶.

References

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