

Scimitar syndrome - A rare cause of recurrent pneumonia

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ABSTRACT

Scimitar syndrome is a congenital anomaly characterized by anomalous drainage of the right lung into inferior vena cava. This may be associated with other anomalies in the form of pulmonary hypoplasia, systemic arterial supply of right lung, and congenital heart diseases. We report an infant with recurrent pneumonia who turned out to be a case of scimitar syndrome on further workup. The patient was managed surgically by selective embolization of the artery from celiac trunk to sequestered lung. This case report highlights the fact that scimitar syndrome should be suspected in a patient with recurrent pneumonia with typical chest X-ray findings.

Key words: *Recurrent pneumonia, Scimitar syndrome, Venolobar syndrome*

Scimitar syndrome is a rare congenital anomaly having an incidence of 1–3/100,000 live births [1]. It is characterized by total or partial anomalous drainage of the right lung into inferior vena cava. It forms part of the pulmonary venolobar syndrome, which includes right lung hypoplasia, congenital heart diseases, and abnormal arterial supply to the right lung. Clinical presentation varies from asymptomatic incidental detection in adulthood to recurrent pneumonia in childhood to pulmonary hypertension and congestive cardiac failure in the newborn period [2]. Here, we describe a case of 1-year-old girl with recurrent pneumonia diagnosed as scimitar syndrome without pulmonary hypertension and cardiac defects. We report this case because of its rarity and to highlight the fact that it is one of the rare causes of recurrent pneumonia.

CASE REPORT

A 1-year-old girl presented to the emergency with fever for 2 days and breathing difficulty for 1 day. She had a history of previous three hospital admissions at the age of 4 months, 6 months, and 9 months, respectively, for similar complaints and was managed conservatively each time. She was born of a full-term normal vaginal delivery at home with uneventful pre-, peri-, and post-natal periods. There was no history of the suck-rest-suck cycle, difficulty in feeding, cyanosis, or recurrent infections at other sites such as skin, ear, sinuses, or gastrointestinal tract.

On examination, she was tachypneic with subcostal retractions. Breath sounds were decreased and coarse crepitation was heard on the right side of the chest. Cardiac apex was shifted to the right fourth intercostal space in the right parasternal area. No murmurs were heard. Chest X-ray showed a small right hemithorax with

shifting of heart and trachea to right side and hyperinflation of the left side of the chest. She was started on intravenous antibiotics with which she gradually improved. She was evaluated for the cause of recurrent pneumonia.

Gastroesophageal reflux scan and bronchoscopy came out to be normal. A two-dimensional (2-D) echo showed dextroposition without any shunt physiology. As the chest X-ray findings were suggestive of atelectasis or pulmonary hypoplasia, contrast-enhanced computed tomography (CT) of the chest was done which showed reduced lung volume on the right side with right mediastinal shift along with an anomalous pulmonary vein on the right side draining into the suprahepatic part of inferior vena cava suggestive of scimitar syndrome. CT pulmonary angiography was done to better delineate the pathology, and it revealed drainage of the right pulmonary vein into inferior vena cava and systemic arterial supply of right lower lobe from aorta, hypoplastic right lung, and right pulmonary artery as shown in Fig. 1. Right heart catheterization was subsequently done which showed findings consistent with scimitar syndrome with right lower lobe receiving its blood supply from celiac trunk and right lung having characteristic “fir tree” appearance on venous angiography. The patient was referred for cardio-thoraco-vascular surgery where coil embolization of the blood supplying artery coming from celiac trunk to sequestered lung was done. The patient is in regular follow-up and is doing well with no subsequent episodes of pneumonia after the procedure.

DISCUSSION

Scimitar syndrome is a form of an anomalous pulmonary venous drainage (APVD), first described by Cooper in 1836. APVD is

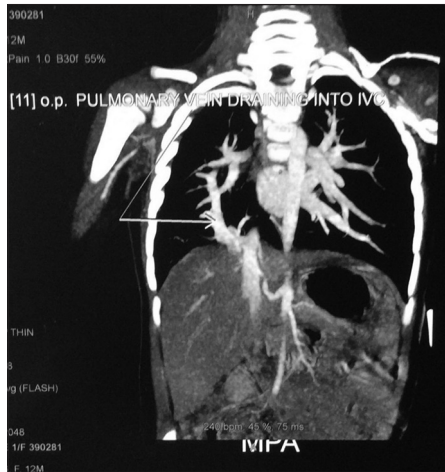


Figure 1: Computed tomography pulmonary angiography

characterized by total or partial anomalous drainage of the right lung into systemic veins, mostly in superior vena cava, inferior vena cava, or right atrium instead of the left atrium. When drainage of pulmonary veins occurs in inferior vena cava, it is known as scimitar syndrome. This APVD leads to right atrium volume overload. The term scimitar syndrome was coined by Naill in 1960 because the anomalous vein along the right heart border as a tubular opacity radiographically resembled a curved Turkish sword or scimitar [3]. This is called as “scimitar sign.” It is more common in females. It is rarely familial and almost never affects the left lung [4].

Scimitar syndrome may be a part of venolobar syndrome which is associated with pulmonary hypoplasia/sequestration, pulmonary artery hypoplasia, supply of right lung from aorta, horseshoe lung, cardiac defects such as atrial septal defect (ASD), ventricular septal defect, patent ductus arteriosus, tetralogy of Fallot, and other anomalies [5]. It is a rare disease with the paucity of data from India and around the world with the five largest case series having a total of 39 cases [6]. Scimitar syndrome has variable presentation depending on different age groups. It presents in infancy with pulmonary hypertension, congestive cardiac failure, and pneumonia. At any age group, it may present with recurrent pneumonia or as an incidental finding on chest X-ray. Our patient was an infant who presented with recurrent pneumonia and did not have pulmonary hypertension and cardiac defects although pulmonary artery hypoplasia with a supply of the right lung from aorta was present.

Physical examination may show decreased breath sounds on the right side with crepitation and mediastinal shift to the right side. Associated ASD may be associated with a fixed split second heart sound but is usually normal. Chest X-ray in scimitar syndrome shows opacified right hemithorax with dextroposition of heart and compensatory hyperinflation of lungs on the opposite side [7]. This X-ray finding is also seen in pulmonary hypoplasia or atelectasis [8]. Our patient had similar X-ray findings. A curved shadow of the anomalous vein descending along the right cardiac border (the scimitar sign) may be seen usually in older children and adults [9].

Scimitar syndrome is usually suspected on the basis of chest X-ray findings. Diagnosis can be further confirmed by contrast

CT of the thorax or CT pulmonary angiography to delineate the associated arterial anomalies. 3-D CT, cardiac-gated magnetic resonance imaging (MRI), cine MRI, and 3-D contrast-enhanced magnetic resonance angiography are newer modalities for diagnosis. Cardiac catheterization remains the gold standard to define the exact anatomical extent of the anomaly which helps in planning the surgical intervention.

The presence of scimitar syndrome is an indication for surgical repair, especially if associated ASD, pulmonary hypertension, pulmonary artery hypoplasia, or scimitar vein stenosis is present [10,11]. Prognosis of patients detected as asymptomatic individuals in adulthood is good. In infants, prognosis depends on the degree of pulmonary hypertension and associated cardiac anomalies and is associated with high mortality. In our patient, selective coil embolization of the blood supplying artery from celiac trunk to the sequestered lung was done

CONCLUSION

This case report highlights the fact that scimitar syndrome should be suspected in a patient with recurrent pneumonia with typical chest X-ray findings. The triad of respiratory distress, right lung hypoplasia, and dextroposition of heart should raise suspicion of scimitar syndrome.

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