Case Report

A rare cause of recurrent pneumonia: Scimitar syndrome

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Abstract

Scimitar syndrome is characterized by partial or total anomalous pulmonary venous return from the right lung along with hypoplasia of the lung. This syndrome has varied presentations, from an asymptomatic state to severe pulmonary hypertension and/or heart failure. Newer diagnostic modalities have improved our understanding of this rare syndrome and hence providing newer treatment options to be tried. Here, we present a case of a 4-year-old child with recurrent pneumonia with Scimitar syndrome. We are reporting this case in view of the rarity of this syndrome and its presentation as recurrent pneumonia.

Key words: *Anomalous pulmonary venous return, Lung hypoplasia, Recurrent pneumonia, Scimitar syndrome*

Recurrent pneumonia often proves to be a diagnostic challenge to the pediatrician due to its numerous causes. Scimitar syndrome is a very rare cause of recurrent pneumonia [1]. The name Scimitar syndrome is derived from the curvilinear vascular pattern created on a chest radiogram by the anomalous pulmonary vein coursing along the right side of the heart towards the diaphragm, which resembles a "Scimitar" or Turkish Sword. The term venolobar syndrome is used when scimitar syndrome is associated with certain other conditions such as right pulmonary hypoplasia, dextroposition of the heart, right pulmonary artery hypoplasia, pulmonary sequestration and anomalous communication between the right lower lobe of the lung, and infradiaphragmatic segment of the aorta [1,2]. Scimitar syndrome has varied presentations, from an asymptomatic state to severe pulmonary hypertension and/or heart failure [3-5]. A high index of suspicion is needed to diagnose scimitar syndrome in any child with recurrent or persistent pneumonia. Here, we present a child diagnosed to have scimitar syndrome who presented to us with a history of recurrent pneumonia.

CASE REPORT

A 4-year-old female child, first born to a third-degree consanguineous marriage was brought with complaints of repeated episodes of lower respiratory tract infections. Child had been hospitalized on four occasions in various hospitals and treated with intravenous antibiotics, the records of which were not available. Antenatal and neonatal history was non-contributory.

On physical examination, the child had respiratory distress. Heart rate was 110/min, respiratory rate - 40/min, temperature was 39°C, Blood pressure 78/52 mm Hg. The child's weight was at 10th centile (13 kg) and height was <10th centile (92 cm). Respiratory system examination revealed small right hemithorax and reduced intensity of breath sounds on the right side. There were crepitations on the right side of the chest. Cardiac examination revealed shift of the apex beat to the right side in fourth intercostal space in right parasternal area without any murmur.

Blood investigations revealed elevated total counts with neutrophilia. Chest X-ray (Fig. 1) showed small right hemithorax with shift of trachea and heart toward right side. Right lung field was more opaque than left due to increased background reticulations. His electrocardiogram was normal and two-dimensional echocardiogram revealed dilatation of the right sided chambers with intact septum. Main pulmonary artery and left pulmonary artery was well seen, but right pulmonary artery could not be visualized. Computed tomography (CT) chest (Fig. 2) revealed a hypoplastic right lung with the shift of the trachea and heart toward the right side. The right inferior pulmonary artery was seen draining into the inferior vena cava at the level of the diaphragm just above the inferior vena cava-hepatic vein junction. A diagnosis of scimitar syndrome was made. The need for surgical intervention was explained to the parents. However, they declined the same in view of financial constraints. Child was treated symptomatically with IV antibiotics and antipyretics.

DISCUSSION

Scimitar syndrome was first described in 1836 by George Cooper during an autopsy of a 10 month old infant. The

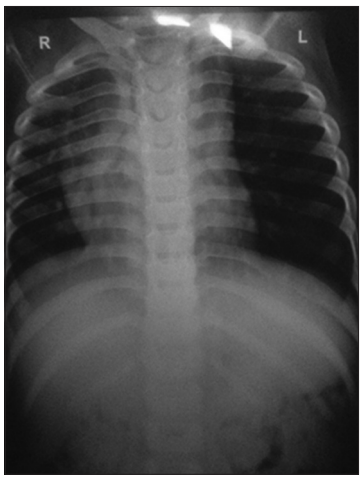

Figure 1: Chest X-ray showing small right lung volume, trachea and cardia pushed to the right side

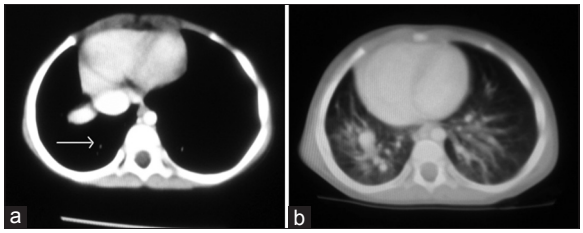

Figure 2: Computed tomography chest showing the right inferior pulmonary artery draining into the inferior vena cava (IVC) at the level of the diaphragm just above the IVC-hepatic vein junction and the hypoplastic right lung

incidence of scimitar syndrome is 1-3/100,000 live births [1,6]. However, its true incidence may be much higher in view of some asymptomatic cases. The disease often presents in early infancy but may present later in childhood or rarely in adults. In infants it usually presents with tachypnea, congestive heart failure, and pulmonary hypertension. It may lead to recurrent lower respiratory tract infections as in our case. Recurrent episodes of wheeze have also been described in the literature. Hemoptysis may be the presenting symptom rarely [7]. Scimitar syndrome is associated with congenital heart disease, mostly ostium secundum type atrial septal defect. In some cases, ventricular septal defect, patent ductus arteriosus, aortic stenosis, aortic arch anomalies, Shone complex, and Fallot's tetralogy can be seen. Other anomalies associated with this syndrome are sequestration of the lower lobe of the right lung, horseshoe lung and anomalous perfusion from the abdominal aorta [5,8-10].

Diagnosis is made with the help of echocardiography, characteristic chest X-ray, CT chest findings. Echocardiography delineates both the Scimitar vein as well as any systemic arterial supply to the right lung. Congenital heart defects are associated with Scimitar syndrome in 75% of infants with severe symptoms, atrial septal defect (ASD) being the most common congenital heart disease. The flow velocity pattern in the Scimitar vein differs from the normal pulmonary venous flow. It is monophasic extending throughout the cardiac cycle with no reverse flow at atrial contraction in the Scimitar vein. CT or magnetic resonance angiography provides excellent visualization of vascular anatomy of this complex congenital defect noninvasively [11].

Cardiac catheterization and angiography are extremely useful tools to confirm the diagnosis and clarify the exact anatomy and degree of pulmonary hypertension. Radionuclide perfusion scan can reveal abnormal arterial supply while ventilation scan can identify bronchial connection in a patient with Scimitar syndrome. In sequestration, aeration can occur through pores of Kohn or through anomalous fistulous connection. Scintigraphic screening is helpful to rule out significant vascular shunting [12].

Treatment primarily involves surgical correction of the anatomical anomaly. However, the most effective surgical method has not been clearly identified. Surgical options include 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 atrium. An intra-atrial patch may be used to create a tunnel, redirecting flow from the anomalous pulmonary vein to the left atrium through an ASD [13]. Uthaman et al. described coil embolization of Scimitar vein and artery, followed by definitive surgery later in life as a method of treatment [14]. Pneumonectomy is required in certain cases. Baskar et al. performed pneumonectomies for three of the six patients in their case series, citing low weight to be the reason for opting out of reimplantation surgery [10].

Pulmonary hypertension is a problem seen in infants and older children with scimitar syndrome [15]. Pulmonary artery pressure was significantly reduced by pulmonary venous stenting in combination with coiling of aberrant vascular supply to the sequestered lung segment in a report by Awasthy et al. [16]. Further experience with such procedures could be useful as it suggests that interventional procedures such as pulmonary venous stenting could be an emergent palliative treatment to relieve pulmonary hypertension in children with Scimitar syndrome.

The age of presentation and the presence of associated anomalies are important in predicting the outcome. In general, presentation in infancy and presence of heart failure are poor prognostic factors.

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