

## Scimitar syndrome: A rare case of recurrent pneumonia

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### Abstract

Scimitar syndrome is a rare, congenital malformation. It is characterized by partial anomalous pulmonary venous connection to inferior vena cava, hypoplastic right lung along with dextroposition of the heart. Depending on the severity of the defect it can present as early as in neonatal period or incidentally later in life. Its incidence is 1-3 per 100,000 patients. We present a case of a two-year-old girl who presented with recurrent fever, cough and was later diagnosed as scimitar syndrome.

**Keywords:** Scimitar syndrome, Dextroposition of heart, Pulmonary hypoplasia.

### Introduction

Scimitar syndrome is a rare condition of right sided pulmonary and vascular malformations. It is also called congenital pulmonary venolobar syndrome. Females are more frequently affected than males. Its incidence is reported to be 1 per 100000. The exact cause or any genetic mutation has not been identified. Scimitar syndrome is a triad of: (1) anomalous vein appearing as curvilinear vascular density along the right heart border (2) hypoplastic right lung or a part of it and (3) heart in dextroposition.

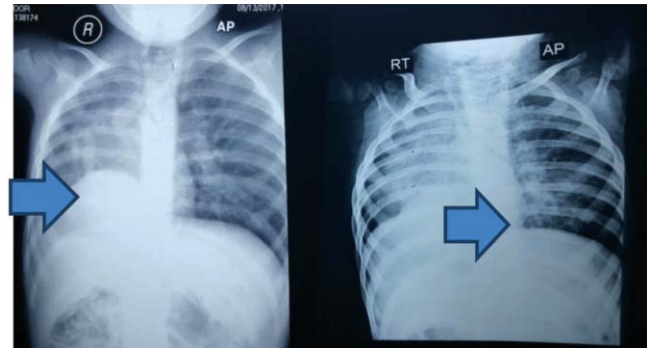
The term Scimitar is derived from Turkish language means a curved Turkish sword or Scimitar because of the radiographic appearance of the anomalous vein, which appears as a tubular density along the right cardiac border. We report the case of a 2-year-old girl diagnosed with this condition. Informed consent was taken from father for this publication. Approval was taken from ethical review committee.

### Case Report

Two years old girl presented in outpatient department of Shifa Falahee Community Health Center, Islamabad on September 2017 with complaint of repeated chest infections. Her symptoms started at five months of age and had been admitted thrice in

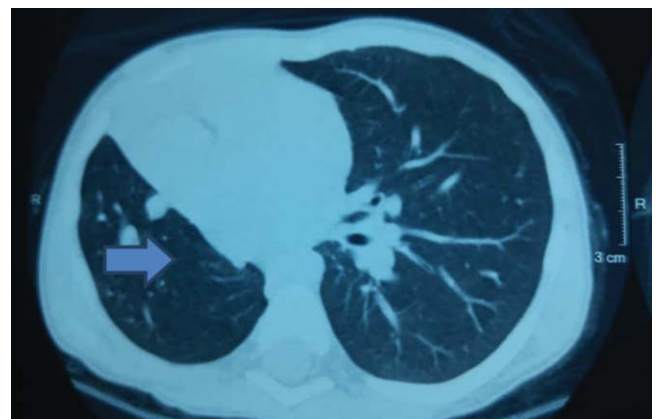
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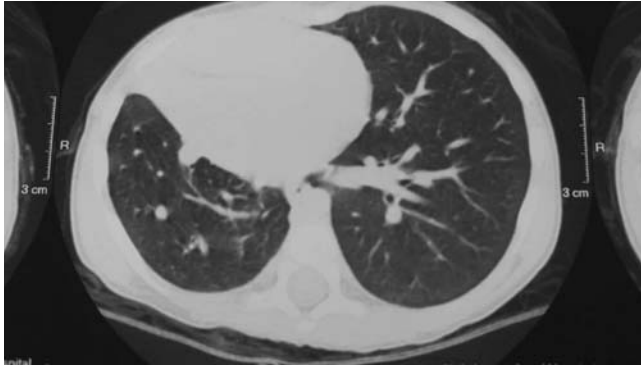


**Figure-1:** Chest X-ray showing scimitar vein appearing as tubular density, dextroposition of heart and lung hypoplasia.

local hospitals for pneumonia. She was born at term via normal delivery and there were no complications at birth. Her parents were non-consanguineous; she has three other siblings who are healthy, including two boys and a girl. On examination, she had no dysmorphism; her height and weight were on the 10th centile. She did not have clubbing; however, her breath sounds were decreased on right side and apex beat was also localized on the right side in 4th intercostal space medial to mid-clavicular line. She was worked up at other hospital and labelled as isolated dextrocardia and previous echo was reported to be normal. She had been extensively investigated previously including immunodeficiency tests.



**Figure-2:** CT scan shows right lung hypoplasia and dextroposition of the heart and scimitar vein.



**Figure-3:** Re-demonstration of defect.

Laboratory evaluation revealed mild normocytic normochromic anemia, her chest x-ray showed reduced lung volume on the right side, mediastinum shift on the right and hyper inflated left lung along with tubular density along right heart border as shown in Figure-1.

High resolution chest CT done showed bronchial atresia of right middle and lower lobe bronchi with simultaneous hypoplasia of right middle and lower lung lobes. Small hypoplastic ipsilateral pulmonary artery, mediastinal shift and compensatory hyperinflation of left lung. A prominent aberrant vessel opening into the suprahepatic IVC just below the diaphragm creating a curvilinear appearance as shown in Figure-2,3. No significant collateral vessels were seen.

Echo showed that the heart was in dextroposition, however, there was no significant dilation of right ventricle and partial anomalous pulmonary venous drainage. Our patient had no pulmonary hypertension at this stage.

The classical triad of hypoplastic lung, anomalous pulmonary venous drainage and dextroposition of the heart confirmed the diagnosis of Scimitar Syndrome.

## Discussion

The Scimitar syndrome is a rare, congenital disorder which is seen more often in girls than boys. The defining abnormality of this condition is a partial anomalous pulmonary venous drainage of a part of or the entire right lung into the inferior vena cava-atrial junction, the inferior vena cava, or the right atrium. Scimitar vein is the name given to the anomalous vein because of its crescent shape on chest radiograph, resembling a Turkish curved scimitar sword. This is demonstrated in the chest radiograph of this patient.<sup>1</sup>

The other important feature of this syndrome is hypoplastic right lung. Majority of cases are associated with a partial or total hypoplastic right lung, which frequently demonstrates lobar anomalies such as a mono- or bilobed.<sup>2</sup> The clinical presentation of this syndrome is variable depending on the underlying defect, similar is the case with age at presentation. With severe defects infants can present with respiratory problems, feeding difficulty, cyanosis and heart failure. Majority of older children who are diagnosed after 1 year of age remain asymptomatic and adults are also diagnosed when characteristic findings are picked up incidentally on chest radiograph.<sup>3</sup>

One of the presentations may be pulmonary hypertension due to a significant left-to-right shunt.<sup>4</sup> It is caused by the anomalous systemic pulmonary venous drainage and associated cardiac defects. Other factors contributing to the development of pulmonary hypertension may include the presence of a systemic arterial supply to the right lung and a stenosis of the anomalous Scimitar vein, which is seen in a minority of patients.

The surgical correction of the defect can be attempted through the following procedures: re-routing the blood flow of the anomalous pulmonary vein to the left atrium, repairing associated cardiac defects, and ligating any anomalous systemic arterial supply to the right lung.<sup>5</sup> In cases where there is associated pulmonary sequestration and recurrent pneumonias, a lobectomy or pneumectomy may be considered. Surgical repair is not required in asymptomatic patients with minimal shunting.<sup>6</sup>

In our patient, condition was fully explained to the family and at present conservative approach was considered. If she gets any further repeated chest infections, then lobectomy will be considered.

## Conclusion

Recurrent pneumonias can have a long list of differential diagnosis depending on history and clinical examination. However, once the common causes like infection, immunodeficiency, cystic fibrosis have been ruled out congenital pulmonary malformations should also be considered. Although rare but when dealing with triad of recurrent pneumonia, right lung hypoplasia and mediastinal shift, physicians should consider possibility of Scimitar syndrome.

**Disclaimer:** None to declare.

**Conflict of Interest:** None to declare.

**Funding Sources:** None to declare.

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