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Case Study

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HORSE-SHOE LUNG IN A RARE CASE OF SCIMITAR SYNDROME

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ABSTRACT

Scimitar syndrome or congenital veno-lobar syndrome is characterised by a hypoplastic lung that is drained by an anomalous pulmonary vein into the systemic circulation. It is a type of partial anomalous pulmonary venous return. Horseshoe lung is a rare congenital anomaly and mostly accompanied by scimitar syndrome. Here we present a case of a female child who had scimitar vein with right lung and right pulmonary artery hypoplasia associated with horseshoe lung.

KEYWORDS: Scimitar syndrome, Horse-shoe lung, pulmonary hypoplasia.

INTRODUCTION

The Scimitar syndrome is a rare congenital anomaly that consists in part of total or partial anomalous venous drainage of the right lung to the inferior vena cava (IVC). Horseshoe lung (HL) is usually an associated anomaly, in which the right and left lungs are fused due to the formation of a narrow isthmus of lung parenchymal tissue between the heart and the aorta. Scimitar syndrome is usually associated with other congenital intracardiac defects, most commonly atrial septal defect. Surgical repair may be needed in case of associated abnormalities and complications like pulmonary hypertension.

CASE REPORT

An 8-year-old female child presented to the Paediatrics OPD with complaints of cough with expectoration for 2 weeks and breathlessness on exertion for 4 years. She had a history of recurrent chest infections since childhood.

On general examination, pulse rate was 86 beats/min, BP was 100/68 mm of Hg and respiratory rate was 16/min. On chest examination, there were reduced movements on the right side of chest as compared to left with shifting of apex beat to the right. On auscultation, breath sounds were reduced on the right side of chest with heart sounds audible on the right.

The laboratory investigations revealed no abnormal parameters. Sputum was negative for acid fast bacilli.

She underwent an emergency chest X-ray PA view which revealed volume loss of right lung as evidenced by mediastinal shift towards right, rib crowding and mild elevation of right hemidiaphragm. Scimitar sign was also noted (a curved vascular shadow located to the right of the heart and descending towards diaphragm).

The patient subsequently underwent HRCT lung which showed dextro-position of the heart and volume loss of right lung. Few atelectatic bands are also noted bilaterally which could represent recurrent infections. An isthmus of lung parenchyma was also seen passing between the aorta and heart which is consistent with horseshoe configuration.

On pulmonary angiography, there was hypoplasia of right main pulmonary artery and aplasia of right superior pulmonary vein. An anomalous pulmonary vein/scimitar vein is seen draining into the IVC.

On echocardiography the scimitar vein was seen along with an atrial septal defect.

Based on clinical and radiological findings of right lung hypoplasia with hypoplastic right pulmonary artery, anomalous scimitar vein draining into IVC, dextro-position of heart and horseshoe lung, a diagnosis with Scimitar syndrome associated with horse-shoe lung was made.

The child was given symptomatic treatment with oral antibiotics. Further she was referred to cardiothoracic and vascular surgery department where she was advised regular interval follow-up. As there was no complication in the form of pulmonary hypertension, corrective surgery for Scimitar vein was not done.

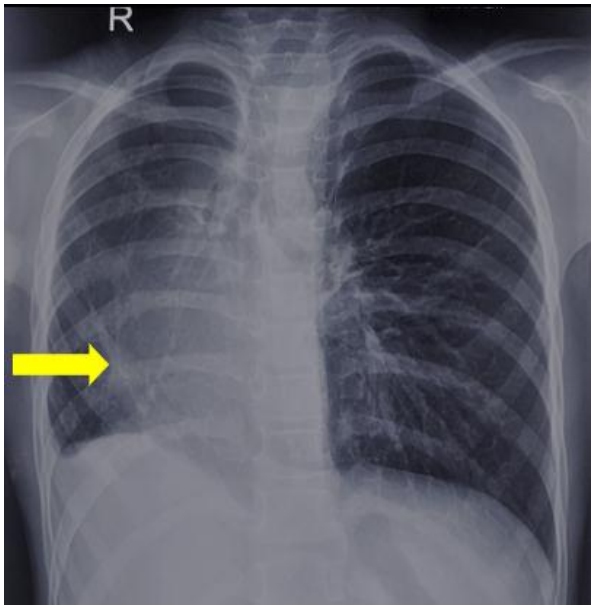

Figure 1: Chest X-ray PA view showing volume loss of right lung as evidenced by mediastinal shift towards right and rib crowding. Scimitar sign (arrow) is seen.

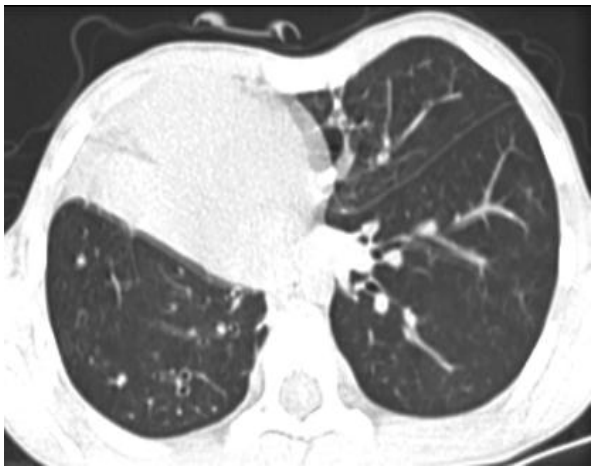

Figure 2: Axial HRCT thorax shows dextro-position of the heart and volume loss of right lung. Few atelectatic bands are also seen bilaterally.

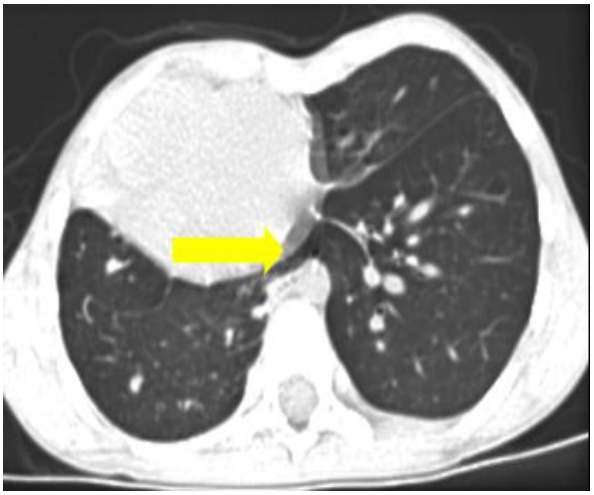

Figure 3: Axial HRCT thorax shows isthmus of lung parenchyma (arrow) passing between aorta and heart. Both lungs are confluent in a horseshoe configuration.

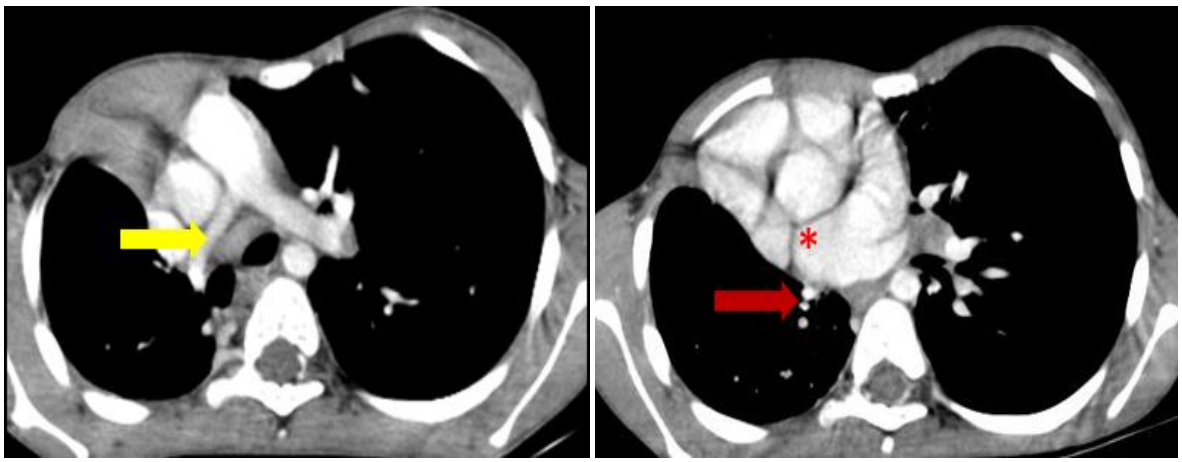

Figure 4(a) and (b) axial angiography images. Image(a) shows right main pulmonary artery hypoplasia (arrow). Image (b) shows right superior pulmonary vein aplasia (*). Right inferior pulmonary vein is normal (arrow).

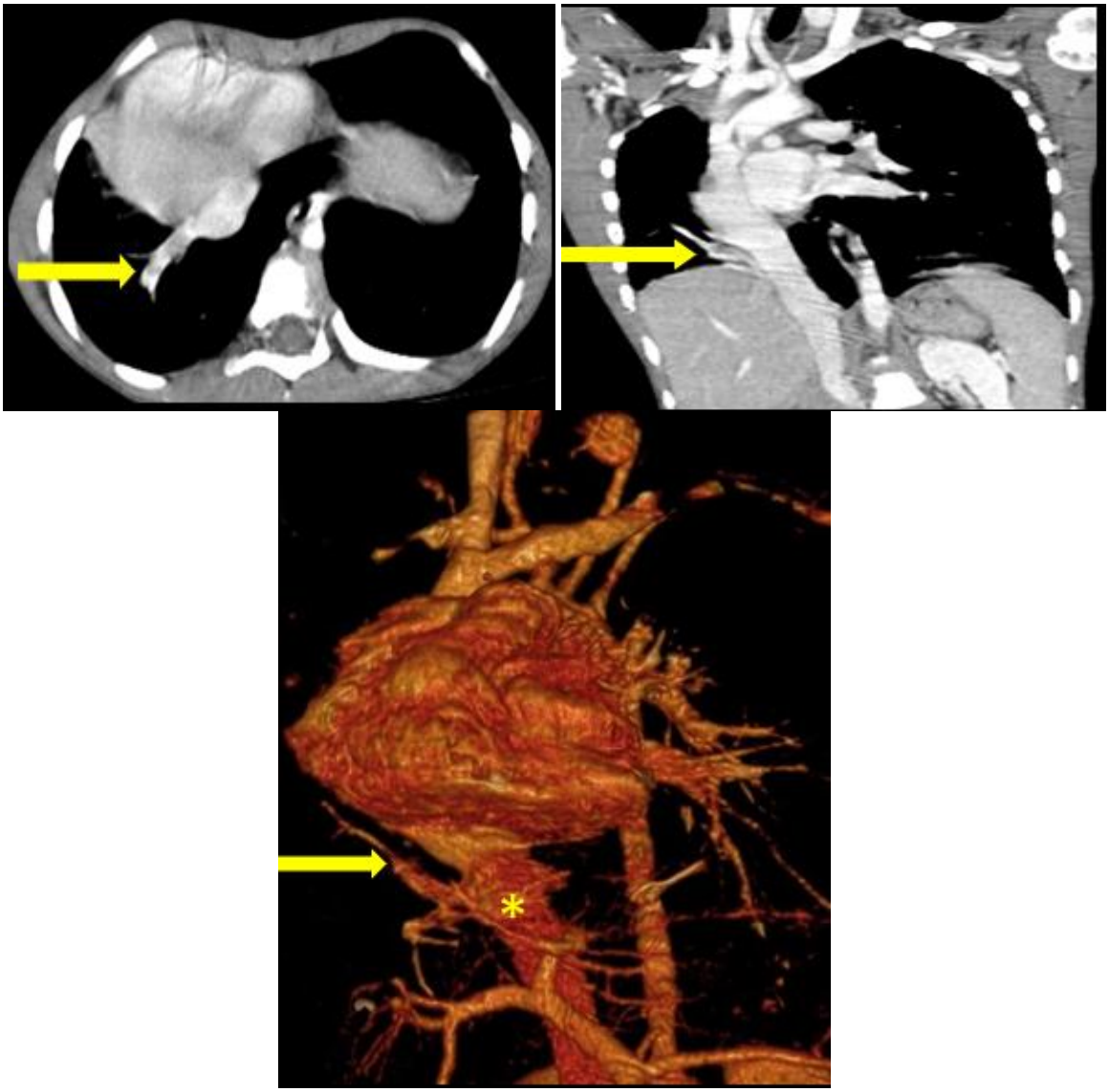

Figure 5(a) Axial, 5(b): Coronal angiography and 5(c) three-dimensional reconstruction images demonstrate anomalous pulmonary vein/scimitar vein (arrow) draining into IVC (*).

DISCUSSION

Scimitar syndrome (SS) also known as hypogenetic lung syndrome or congenital veno-lobar syndrome is primarily a complex developmental lung abnormality with anomalous venous return. The appearance on chest radiography resembles a curved Turkish sword, or scimitar. The estimated incidence of Scimitar syndrome ranges from 1 to 3 per 100,000 live births and a 2:1 female predominance.[1]

The disease aetiology is probably due to an embryological error of the fundamental development of the lung bud in early embryogenesis.[1]

Although various other congenital cardiopulmonary abnormalities such as hypoplasia of the right lung, pulmonary sequestration, persisting left superior vena cava, various congenital intracardiac defects and dextroposition of the heart have been described in association with scimitar syndrome, horseshoe lung anomaly with scimitar syndrome is a very rare association.[2]

The most common associated cardiac defects are atrial septal defect (80%), patent ductus arteriosus (75%), ventricular septal defect (30%) and pulmonary vein stenosis (20%); less commonly seen are tetralogy of Fallot, aortic arch hypoplasia or coarctation and hypoplastic left heart syndrome.

The term “horseshoe lung” denotes an anomalous isthmus of pulmonary parenchyma extending from the base of the right lung and fusing with the base of the left lung between the aorta and heart. Up to 80% of reported cases of horseshoe lung have been seen in association with scimitar syndrome.[3]

Clinical features seen during infancy maybe severe including failure to thrive, tachypnoea and heart failure.

Symptoms in the childhood/adult type are milder and most commonly include recurrent pneumonia.

The diagnosis can be confirmed by radiography, angiography, echocardiography, CT angiography (CTA), or magnetic resonance angiography (MRA).[4]

Radiography shows mediastinal shift and scimitar sign (a tubular structure paralleling the right heart border). On echocardiography visualization of a confluence behind the right atrium and a vertical vein are the most consistent findings.[5] On CTA or MRA the anomalous venous connection of the scimitar vein with the inferior vena cava is seen.[4]

Treatment depends on the associated clinical abnormalities. In case of incidental diagnosis there is no therapeutic intervention. In cases of associated abnormalities and severe clinical problems (pulmonary hypertension), operative repair is mandatory. The classic operation encompasses construction of a long intra-atrial baffle from the entry point of the scimitar vein into the inferior vena cava to the left atrium through an ASD.[6]

An early diagnosis with advance imaging facilitates the surgical strategy, contributing to low morbidity and mortality rates after corrective surgery. There is a need to define the best surgical option, but longer follow-up is necessary.[1]

CONCLUSION

Horseshoe lung is a rare congenital lung abnormality associated with Scimitar syndrome. Most aspects of this complex anomaly can be demonstrated via multidetector CT (MDCT). Chest radiograph, echocardiography and MR angiography are also useful in diagnosis. Surgical treatment depends upon presence of associated congenital cardiac anomalies and complications.

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