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A RARE CASE REPORT ON PULMONARY VENOLOBAR SYNDROME (SCIMITAR); UNUSUAL PRESENTATION WITH DEXTROCARDIA OF HEART

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Abstract:

Scimitar syndrome is a rare congenital heart disorder. This syndrome is referred to as scimitar syndrome or pulmonary venolobar syndrome because of a curvilinear pattern visible on the chest radiography, resembling a sword (scimitar) due to the abnormal drainage of pulmonary veins to the inferior vena cava instead of draining in to the heart. We intend to report the case of a female child, incidentally diagnosed as scimitar syndrome planned for cardiac catheterization.

Keywords: Scimitar, Congenital, Pulmonary venolobar syndrome, Catheterization, Dextrocardia, Collateral

INTRODUCTION

Scimitar syndrome is a rare congenital anomaly, demonstrating the partial of right pulmonary venous return to the inferior vena cava (IVC).[1] It consisting of an anomalous pulmonary venous drainage from the right lung to the inferior vena cava. It is also associated with other kind of abnormalities such as hypoplastic right lung, anomalous systemic arterial supply to the right lung with or without pulmonary sequestration, pulmonary hypertension, dextroposition of the heart and atrial septal defect (ASD), with ostium secundum being the most common.[2] In india, pulmonary hypertension is more prevalent.[3] The term "scimitar syndrome" was derived from the shadow extends from the superior lateral position of the right lung to the medial location; increases in caliber as it descends towards the cardiophrenic angle. This appearance is look likes that of a "Scimitar" or it closely resembles a curved "Turkish Sword"[4] (Figure 1). There are very few case reports of scimitar syndrome. Thus, we intend to report a case of a female child planned for cardiac catheterization, incidentally diagnosed as Scimitar Syndrome during evaluation.

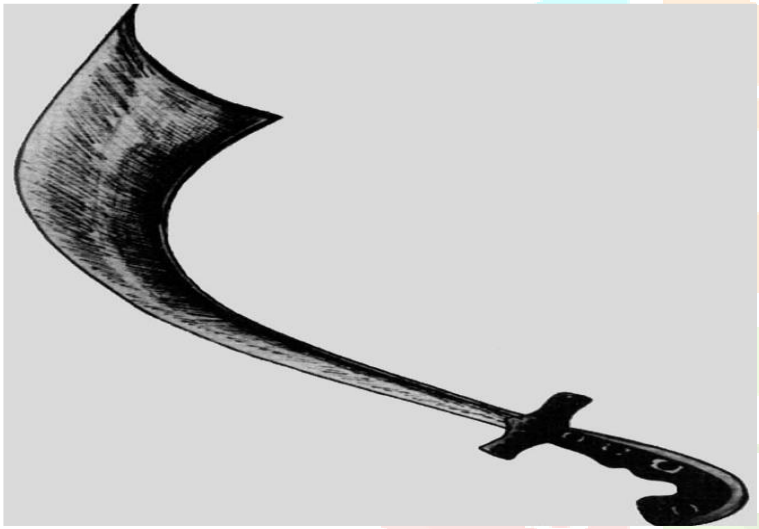

Fig. 1 sketch of scimitar

CASE REPORT

A 5yr old female was admitted in the Department of Pediatric Cardiology and was diagnosed as Scimitar Syndrome while being evaluated for lower respiratory tract infection(LRTI). She was treated as PTB at 1½ years of age for 6 months with ATT. She had no history of syncope, lightheadness or diaphoresis. Her family had a history of asthma in paternal grandmother and had a history of seizure disorder in siblings. Of note, her parents reported a history of recurrent respiratory infections and was treated on outside hospital.

Her physical examination shows no apparent distress. Chest examination demonstrated that she had normal chest wall excursion with clear bilateral breath sounds. Her heart rate was regular, S_1 was normal and S_2 was widely split and did not vary with respiration. No significant murmurs. Her chest radiography (Figure 2)showed dextrocardia with an anomalous drainage of only RLPV to IVC-RA function(Figure 3a, 3b); doubtful of scimitar syndrome. Oxygen saturation was 98%. Systolic pulmonary artery pressure was 23 mmHg. Laboratory findings were normal. She has been evaluated in detail and was planned for elective cardiac cath and was admitted for the same.

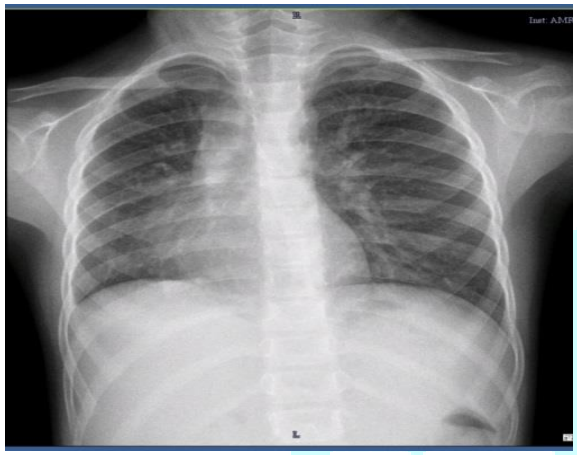

Fig. 2 chest x-ray shows dextroposed heart, reduced right lung volume

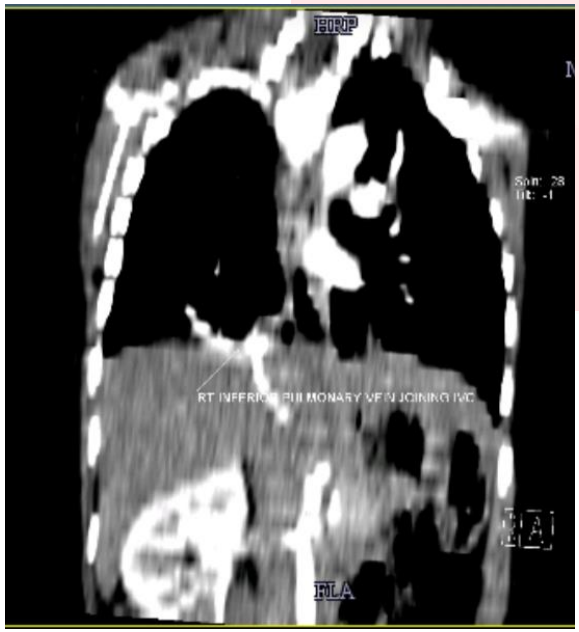

Fig. 3a

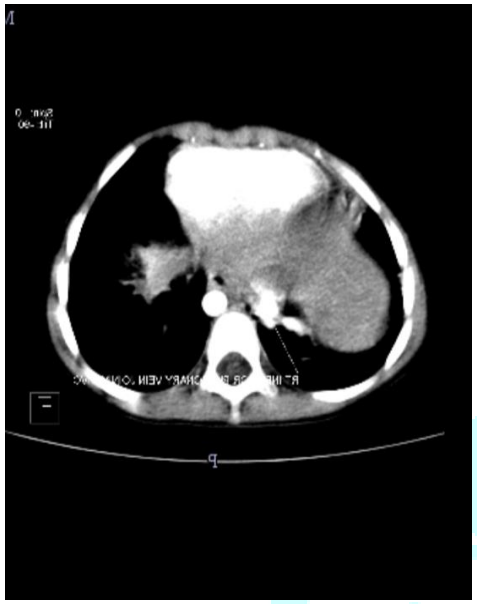

Fig. 3b

(Figure 3a, 3b: Right inferior pulmonary vein joining IVC)

DISCUSSION

Scimitar syndrome is a rare constellation of anomalies; resembles an ASD, estimated to occur in 2/100,000 births. It is more commonly seen in females and is occasionally familial.[1] It is defined as the malformation of the right lung characterized by an abnormal right sided pulmonary venous drainage in the inferior venacava. It was first described in 1836 by Cooper in London during an autopsy of an infant. Note was made of "dextropostion of the heart and hypoplasia of right lung.[2] In the same year, Chassinat documented similar finding on a necropsy in Paris.[5] The first diagnosis was made by Dotter et al in 1949 in a live and asymptomatic patient on cardiac catheterization.[6] The first surgery was performed in 1950 by Drank & Lynch.[7] The first corrective surgery was performed by Kirklin et al in 1956 using cardiopulmonary bypass on a patient with SV and an ASD.[8] Neil et al in 1960 reported the term Scimitar Syndrome in the Bulletin of John Hopkins Hospital entitled "The familial occurrence of hypoplastic right lung with systemic arterial supply and venous drainage-Scimitar Syndrome".[9]

Considering the rareness of the syndrome and it`s atypical form of presentation, this case is reported. In this case, the patient was admitted for elective cardiac catheterization for hemodynamic assessment and demonstration of aortopulmonary collateral. The procedure was done under IV sedation. No significant R-L shunt was noticed. There was anomalous drainage of only RLPV to IVC-RA junction. A collateral was demonstrated to right lower lobe of lung arising from celiac plexus and was coiled with 0.038"*5cm*5mm MREYE embolization coil. The child was tolerated the procedure and was stable at the time of discharge.

Cardiac catheterization is not considered a surgical procedure because there is no large incision used to open the chest and the recovery time is much shorter than that of surgery. In some cases, surgery may be recommended afterward, depending on the results of the procedure. During the course of the heart catheterization the anomalous pulmonary vein was entered from the vena cava. Outside of demonstrating a left to right shunt at the same level, the catheterization findings were normal. In this case, no significant shunt was noticed.

CONCLUSIONS

Scimitar syndrome is a very rare disease. It has varied presentation.[10] In adults, the syndrome is of mild variety, generally without pulmonary hypertension, treatment is not required in this case. When the syndrome appears in neonatal period, it shows a heart or respiratory failure secondary to pulmonary hypertension and generally requires surgery.[11] A plain chest radiography may help to reveal the diagnosis. Therefore, a careful evaluation of radiography scans is very important. Possible associated disorders should be screened for using echocardiography, as well as other advanced imaging methods. Treatment depends on age and symptomatology. Congenital heart diseases comprehend an unusual range of morphological defect. Proper and keen observation over its anatomy and morphology is helpful for the diagnosis and management.[12] Patients with CHD's in relation with an increased pulmonic blood flow, have a strange growth and remaking of vascular bed.[13] Children with uncorrected CHD have significant reduction in their overall health related quality of life.[14] Patients with scimitar syndrome coming out during infancy or significantly associated with congenital heart diseases usually have imperfect outcomes with either medical or surgical management.

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