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Turkish Sword can Track the Way of Pulmonary Vein: A Concealed Finding

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Abstract:

Scimitar syndrome is an unusual developmental anomaly causing partial anomalous pulmonary venous return (right sided pulmonary veins) with left-to-right shunt at inter atrial septal level. It occurs more commonly in females, with occasional familial occurrence. The clinical presentation is variable. Patient may remain asymptomatic or may present with heart failure. The objective of this clinical case report is to highlight this unusual anomaly to avoid incorrect diagnosis. A girl of 13 years presented with unusual symptom of nausea following taking meal and a systolic murmur. At first she was diagnosed as "ASD Secundum". But her typical "Turkish sword" feature of chest x-ray helped to identify anomalous drainage of right pulmonary veins into inferior vena cava and make the diagnosis as "Scimitar syndrome". It is a total corrective anomaly. She was operated through open heart surgery, recovered early and discharged with a stable condition without any complication. This case illustrates the importance of thorough medical history, careful evaluation of investigation for proper diagnosis, treatment procedure and complications of a rare anomaly.

Keywords: *Scimitar syndrome, partial anomalous pulmonary venous return, acyanotic, right pulmonary veins, inferior vena cava*

Introduction:

Scimitar syndrome is anomalous drainage of right sided pulmonary veins with hypoplasia of right lung and right pulmonary artery with dextroposition of heart. The partial anomalous venous connection of the right lung drains into the inferior vena cava or azygous system, and left sided pulmonary veins are draining normally at left atrium. Radiographs play a role in the incidental detection and initial imaging evaluation in patients with clinical suspicion of congenital lung anomalies as computed tomography and catheterization is frequently required as preoperative evaluation for confirmation of diagnosis in the case of surgical lesions. A proper evaluation can diagnose total anomalous condition of a patient.

Case report:

A 13 years old girl came to National Heart Foundation Hospital and Research Institute, Dhaka with complaints of nausea following taking meal. She had history of taking antiemetic and proton pump inhibitors. On auscultation she had a systolic murmur with grading of 3/6. She was acyanotic and on regular pulse with good volume. Initially she was diagnosed as "ASD Secundum". But her chest x-ray findings with typical "Turkish sword" help us to re-evaluate her.

Her transthoracic color doppler echocardiography demonstrated anomalous flow into the inferior vena cava. Right sided pulmonary veins forming a scimitar vein which draining to inferior vena cava and other left sided pulmonary vein draining normally to left atrium. There was left to right shunt at interatrial septal level with right sided chamber (atrium & ventricle) dilatation and her pulmonary arterial pressure was 38mmHg +Right atrial pressure.

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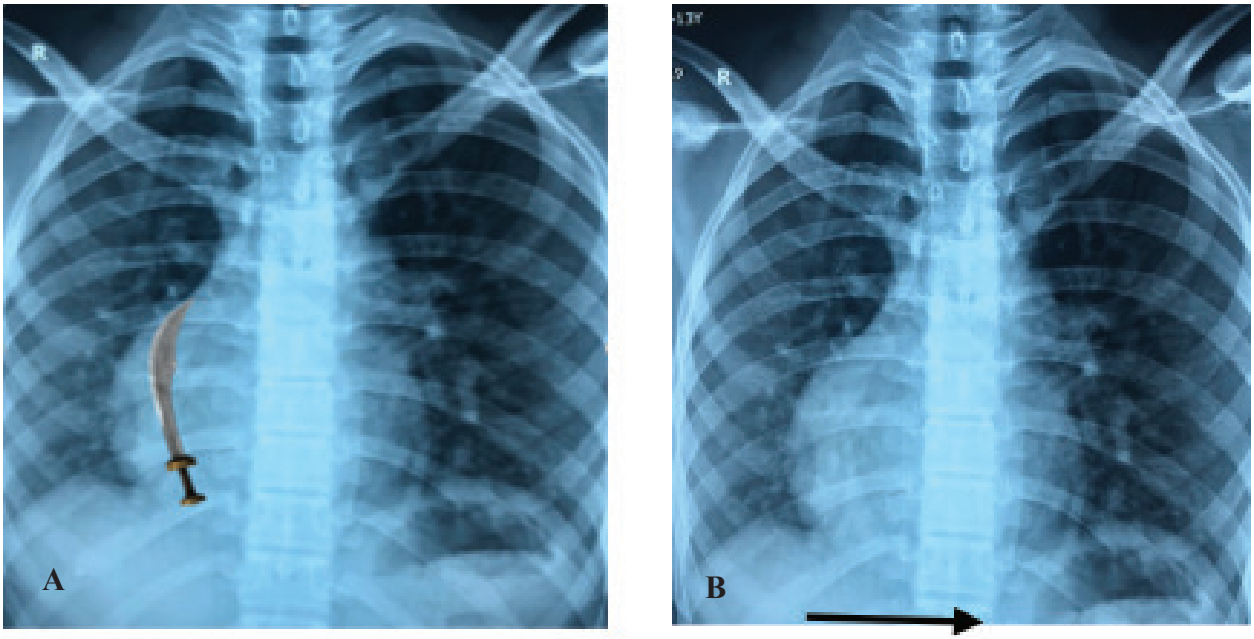

Fig.-1: *Chest x-ray P/A (A,B) view showing the dextroposition of heart with scimitar sign (Turkish sword)*

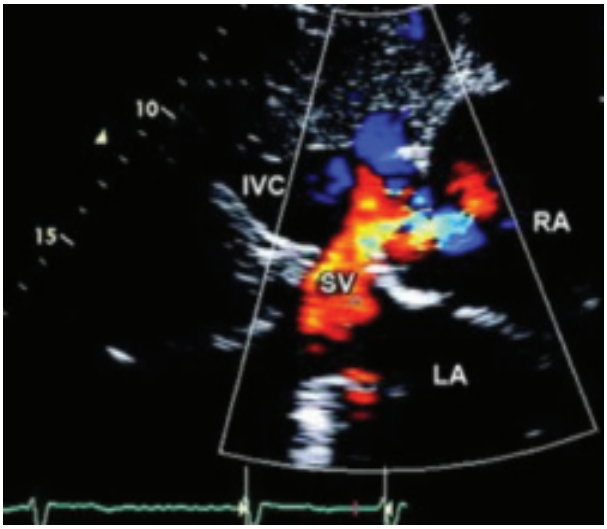

Fig.-2: *subcostal echocardiography showing modified bicaval view with draining of scimitar vein into IVC (SV= Scimitar vein, IVC=Inferior vena cava, RA= Right atrium, LA= Left atrium)*

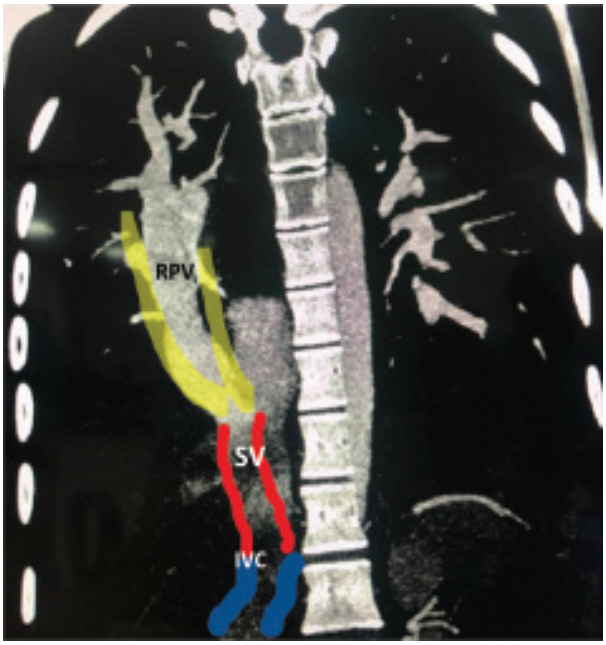

Fig.-3 : *CT image showing scimitar vein (SV) that is connected with inferior vena cava (IVC).*

We did CT angiogram of heart and great vessels for better understanding that confirm anomalous drainage of right sided pulmonary veins to scimitar vein to IVC and left sided pulmonary veins draining to left atrium. But she had no hypoplasia of right lung that could be an associated anomaly.

We attempted to do a cardiac catheter study for reversibility, that demonstrated a significant left-to-right shunt with a step-up in oxygen saturation from the inferior vena cava (IVC) to lower right atrium and a scimitar vein was confirmed. Pulmonary arterial pressure was 48/25/33 mmHg and systemic pressure 128/72/90 mmHg. Pulmonary flow: Systemic flow (Qp:Qs) =1.8:1 with pulmonary vascular resistance 3 WU.

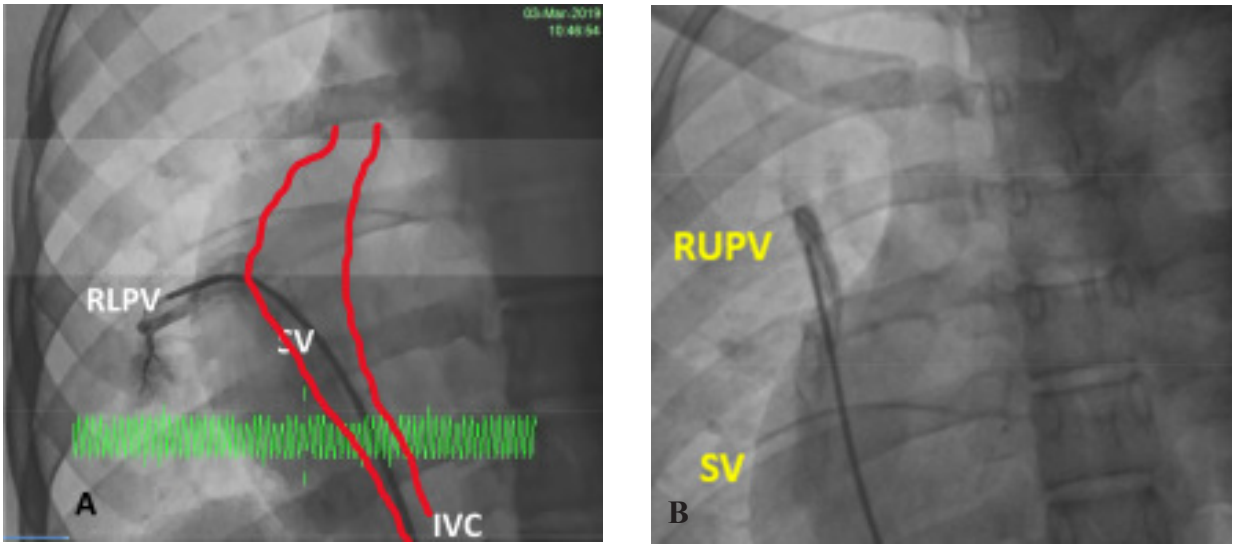

Fig.-4: *Cardiac catheterization showed Scimitar vein draining to the IVC. (scimitar vein =SV, Inferior venae cava=IVC, RPV= Right pulmonary vein.)*

We also did her spirometry and diffuse lung co-efficient for carbon monoxide (DLCO) test and took the opinion of a respiratory physician about outcome of her cardiac surgery. Her lung function test was perfectly alright.

Following proper evaluation- our patient was finally diagnosed as "Scimitar Syndrome" and had undergone open heart surgery. Per operative findings revealed right upper and lower pulmonary veins drain into the junction of RA and IVC after forming a common channel (scimitar). ASD enlarged inferiorly and Rerouting of the common venous channel to Left atrium done by a pericardial patch. Anterior wall of IVC was augmented by pericardial patch. Patient was shifted to ICU with stable haemodynamics. She was extubated on 1st post-operative day and her post-operative course was uneventful. She was discharged from hospital on 7th post-operative day. She was on sound health and post-operative echocardiography was normal on her 3,6 & 12 month

Discussion:

Scimitar syndrome is a rare, but well characterized disease accounting for 0.5–1% of all congenital heart disease[1]. One-quarter of the cases are symptomatic in the newborn, presenting as respiratory or cardiac failure needing corrective surgery. This syndrome occurs as the result of abnormal development of the right lung bud in the early embryogenesis. Most frequently it tends to involve the right lung. But involvement of left lung is rare[2]. It occurs more commonly in females, with occasional familial occurrence. The clinical presentation is variable. Patient may remain asymptomatic or may present with heart failure. This is a pulmonary venolobar syndrome and associated partial anomalous pulmonary venous connection of the right pulmonary veins, via the confluence to the infra-diaphragmatic inferior vena cava, above the drainage site of the hepatic veins. This congenital cardiovascular defect includes a hypoplastic right pulmonary artery and right lung, which leads to displacement of cardiac structures into the right hemithorax, anomalous systemic arterial supply to the right lung[3,4,5]. There is usually no obstruction to pulmonary venous flow. Rare cases of connection to the hepatic veins or to the portal vein have been documented[1]. Two varieties have been described. a) The infantile form presents in infancy or early childhood due to other associated cardiac anomalies like Fallot's tetralogy, co-arctation of the aorta, patent ductus arteriosus and ventricular septal defect. b) The adult form, where the syndrome is detected after the first year of life, presents late or may remain totally asymptomatic and is not associated with other cardiac anomalies[1]. Though our case is infantile form, late presentation make the diagnosis delayed. Chest radiography demonstrated mediastinal shift to the right, a small right hilum, volume loss affecting the right hemi-thorax and an abnormal vascular structure arcing supero-laterally

from the inferior right heart border. The anomalous vein is said to resemble a curved Turkish sword ('scimitar'). Computerized tomography of the chest revealed dextro-position of the heart, hypoplasia of the right lung and pulmonary artery and a large anomalous right pulmonary vein draining into the inferior vena cava.[6] This syndrome is frequently associated with a variety of congenital heart defects. Atrial septal defect, ventricular septal defect, patent ductus arteriosus and coarctation of the aorta are the most frequent associated defects. Tetralogy of Fallot, endocardial cushion defect, double-outlet right ventricle and hypoplastic left heart syndrome have also been reported in association with scimitar syndrome[7]. This syndrome also associated with congenital hydrocephalus but this is a negative prognostic factor[8]. This syndrome exhibits autosomal dominant inheritance with variable penetrance. The median age for presentation of this syndrome is 7 months, although the age at presentation varies[9]. Medical treatment may be of temporary benefit if there is increased pulmonary blood flow that may cause hyperkinetic pulmonary arterial hypertension or consequence of pulmonary vascular disease. But definite therapy is surgical correction. Post-operative impact depends on pulmonary vascular bed at the time of surgery. Though mortality following surgery is less than 1%, postoperative arrhythmia, pulmonary hypertension and pulmonary venous obstruction are reported in few studies[10].

Conclusion:

A rare disease can be diagnosed by proper evaluation of history, though patient may remain asymptomatic or may present with unusual symptoms - a small clue from initial investigation can help to identify rare anomaly. Patient with scimitar syndrome can remain asymptomatic or can be present with congestive failure depending on severity of disease. Correction of associated cardiac defects with rerouting of scimitar vein into left atrium lead to favorable outcome and postoperative follow up can identify related complications.

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