Scimitar syndrome

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Sri Lanka Journal of Child Health, 2013; **42**(2): 103-104

(Key word: scimitar syndrome)

Introduction

Scimitar syndrome is a rare congenital anomaly, characterized by partial or complete anomalous pulmonary venous drainage of the right or left lung into the inferior vena cava[1]. Because the radiographic shadow of the anomalous vein resembles a curved Turkish sabre, this defect has been named the "scimitar deformity"[2].

Case report

An 8 year old boy presented to ward 2, Colombo North Teaching Hospital, with a history of lower respiratory tract infection and 3 episodes of haemoptysis over a 3 month period. Examination revealed, reduced air entry on the right side of the chest, dextroposition of the heart, grade 2 ejection systolic murmur at the left upper sternal border and absence of cyanosis. Chest x-ray showed the 'scimitar vein parallel to the right cardiac border, right lung hypoplasia, dextroposition of the heart and hyperinflation of the left lung (Figure 1).

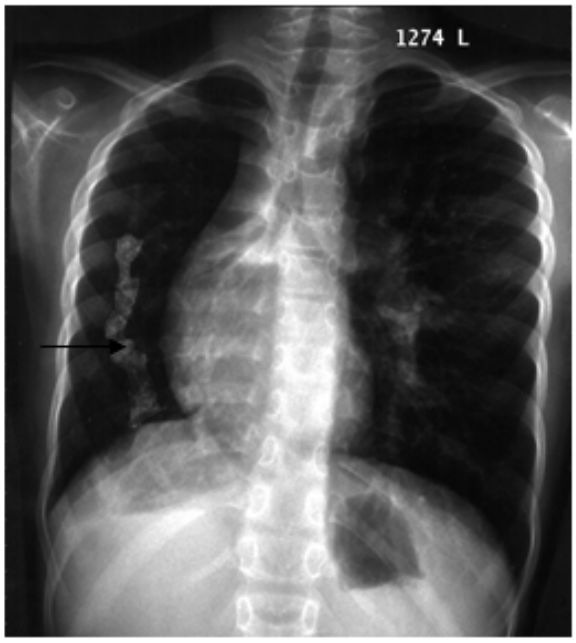

Figure 1: ***Chest X-ray (Scimitar vein shown by arrow)***

Computerised tomography (CT) contrast of chest revealed right lung hypoplasia, compensatory hyperinflation of left lung, dextroposition of the heart, hypoplastic right main pulmonary artery and the right lung drained by a scimitar vein into the infra-diaphragmatic inferior vena cava (IVC) (Figure 2).

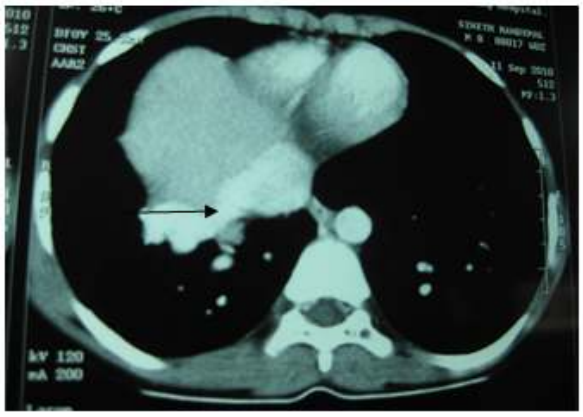

Figure 2: ***CT scan of chest. Scimitar vein opens into IVC as shown by arrow***.

Echocardiography revealed partial anomalous pulmonary venous connection of the right pulmonary vein, draining via scimitar vein into IVC close to IVC/right atrium junction, intact intra-atrial septum, with mild pulmonary hypertension. Cardiac catheterization was done to delineate pulmonary venous drainage, and to locate any systemic to pulmonary collaterals (Figures 3 and 4).

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(Received on 20 September 2012: Accepted after revision on 19 October 2012)

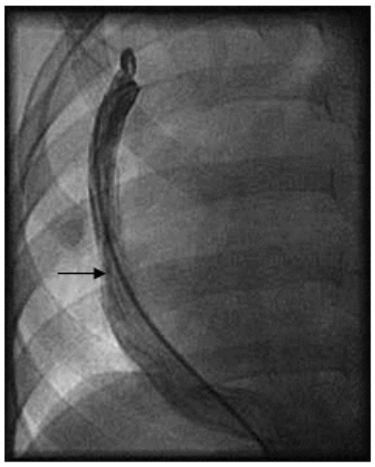

Figure 3: ***Angiogram of scimitar vein***.

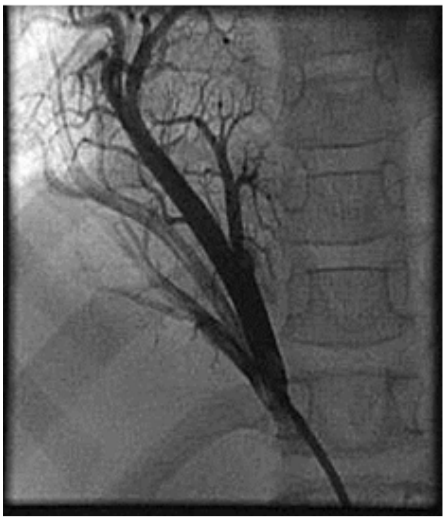

Figure 4: ***Angiogram of systemic pulmonary collaterals***

As CT showed mild hypoplastic right lung, it was planned for direction of scimitar vein to left atrium and not right pneumonectomy. Our patient successfully underwent trans-catheter occlusion of systemic to pulmonary collaterals, using multiple embolization coils and is awaiting re-routing of scimitar vein to left atrium.

Discussion

Scimitar syndrome is commonly associated with hypoplasia of the right lung, pulmonary sequestration, persisting left superior vena cava, and dextroposition of the heart[2]. The pathogenesis of the syndrome is unclear, but it seems to originate from a basic developmental disorder of the entire lung bud early in embryogenesis[2]. Two main forms of scimitar syndrome have been described. Signs and symptoms can start during infancy (infantile form) or beyond (childhood/adult form). The infantile form generally presents within the first 2 months of life with tachypnea, recurrent pneumonia, failure to thrive, and signs of heart failure[2]. The diagnosis of scimitar syndrome is usually made based on the characteristic chest X-ray films and can be confirmed by angiography[2]. In the chest x-ray, a diagnostic vertically directed crescent shadow (scimitar sign) is observed to the right of the mediastinal silhouette[3].Cardiac dextroposition was present in our patient and the chest x-ray and the scimitar sign was present in the chest x-ray (Figure 1).

Asymptomatic patients in the absence of associated abnormalities can be followed up conservatively. For symptomatic patients, pulmonary: systemic blood flow (Qp: Qs) ratio greater than 1.5 and pulmonary hypertension require rerouting of anomalous right pulmonary veins to left atrium in order to avoid progression of right ventricular failure and it may require occlusion of systemic to pulmonary collaterals to reduce left to right shunt and thereby prevent any complications until corrective surgery[1]. The presence of haemoptysis is likely to be secondary to an anomalous systemic arterial blood supply to lung and it may warrant trans-catheter occlusion of the anomalous feeding arterial vessels[4]. This was done in our patient.

The triad of respiratory distress, right lung hypoplasia and dextroposition of the heart should alert the clinician to think of scimitar syndrome.

References

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