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Case Report

Fortuitious Diagnosis of Scimitar Syndrome in an Asymptomatic Adult

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ABSTRACT

Scimitar syndrome is a rare congenital anomaly seen as one of the components of congenital pulmonary venolobar syndrome. Simple and traditional investigation tools like CXR if interpreted with vigil can aid in prompt diagnosis and management of patients even in absence of clinical symptoms.

Key words: asymptomatic; adult; scimitar syndrome

Introduction

Scimitar syndrome is a rare congenital anomaly seen as one of the components of congenital pulmonary venolobar syndrome. It is a combination of pulmonary hypoplasia and partial anomalous pulmonary venous return (PAPVR) and occurs exclusively on the right side. The name 'scimitar' is derived from the curvilinear vascular pattern seen on chest x-ray due to the anomalous pulmonary vein coursing along the right side of the heart towards the diaphragm, resembling a 'Scimitar' or a Turkish sword. We report scimitar syndrome diagnosed after identifying this scimitar appearance on a routine chest x-ray done (CXR) in an asymptomatic embassy case.

CASE REPORT:

A 27 year old healthy male presented to our outpatient department for health check-up as an embassy case. He was asymptomatic with no significant past or family history. General physical examination, cardiovascular and respiratory examinations were unremarkable. Routine CXR revealed a curvilinear pattern on right side of the heart (Figure 1).

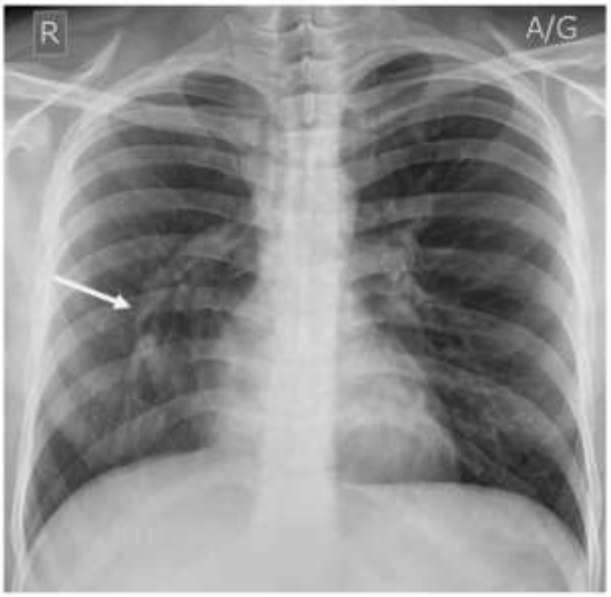

Figure1: CXR showing scimitar sign (arrow)

EKG done revealed normal sinus rhythm with right axis deviation. Transthoracic echocardiography (TTE) demonstrated dilated right atrium and right ventricle, mild tricuspid regurgitation and pulmonary artery systolic pressure of 30mm Hg. Transesophageal echocardiography (TEE) showed drainage of right upper and lower pulmonary veins into the inferior vena cava (IVC).

CT pulmonary angiogram was performed which revealed right upper and lower pulmonary veins draining into suprahepatic IVC, thus confirming scimitar syndrome (Figure 2).

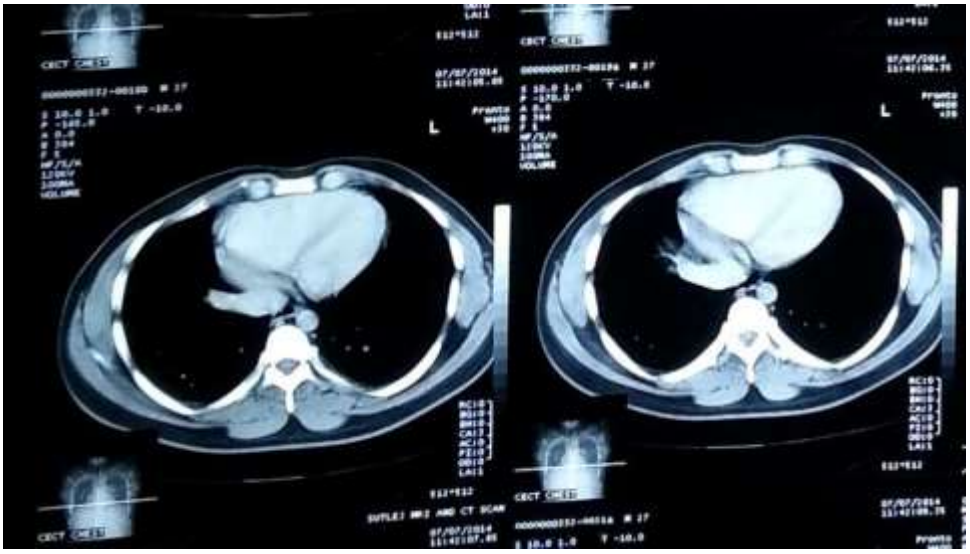

Figure 2: CT pulmonary angiogram showing right upper and lower pulmonary veins draining into suprahepatic IVC

Cardiac catheterization was done which demonstrated scimitar vein and a significant step up in oxygen saturation in IVC above and below renal veins (Figure 3).

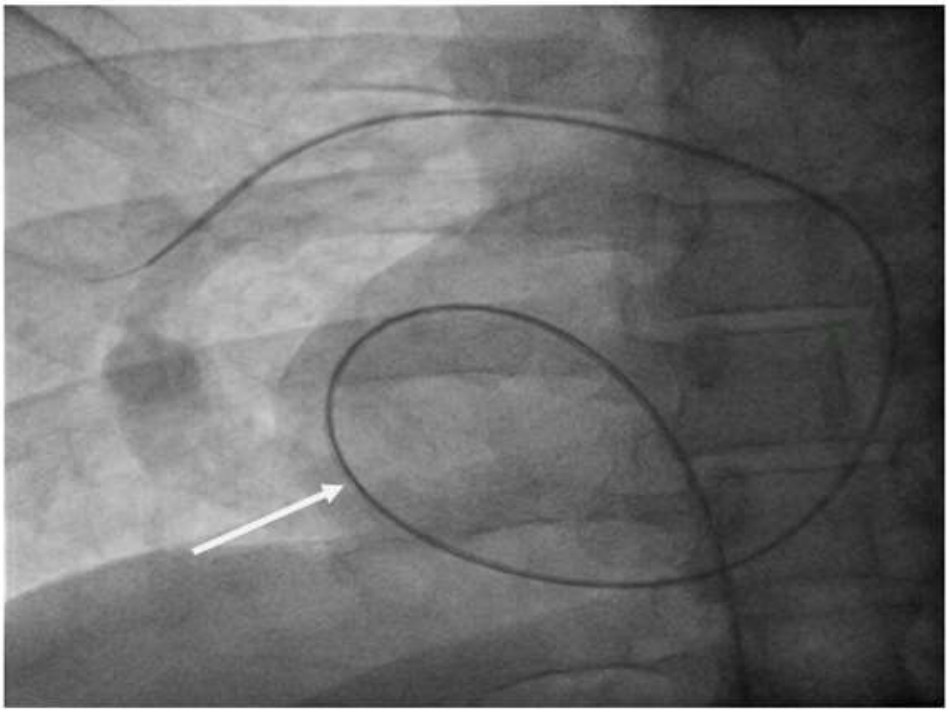

Figure 3: Cardiac catheterization showing scimitar vein (arrow)

Mean pulmonary pressures were normal and Qp/Qs was 1.5. No associated anomalies were noticed. Since the patient was completely asymptomatic, he was managed conservatively.

DISCUSSION:

Scimitar syndrome is a constellation of cardiopulmonary anomalies and accounts for about 0.5-1% of congenital heart diseases. It has an incidence of 1-3/1, 000, 00 live births [1, 2]. The exact cause is still not known. It is characterized by an anomalous pulmonary vein which usually drains into IVC (most common), right atrium or portal vein. Hemodynamically there

is an acyanotic left to right shunt due to connection of pulmonary venous circulation and systemic venous circulation. The presenting features depend on the age of the patient with infantile form being worse than the adult form. In infancy it usually presents with cyanosis, poor growth, tachypnea, pulmonary hypertension and congestive heart failure. However adults have a more benign course [3] and may experience recurrent pulmonary infections, hemoptysis or exertional dyspnea. Common associations of this syndrome include atrial septal defect, ventricular septal defect, dextrocardia, bochdeleck hernia, horseshoe lung, hemivertebrae and pulmonary sequestration [4]. CXR is diagnostic and shows a small lung with ipsilateral mediastinal shift and an anomalous draining vein seen as a curvilinear shadow paralleling the right heart border in the shape of a Turkish sword or 'scimitar'. It can be further confirmed by echocardiography and CT or MR angiography. Cardiac catheterization is done to identify the course of anomalous vein, measure the degree of left to right shunt, assess for pulmonary hypertension and to look for associated cardiac anomalies. Important differential diagnosis includes pulmonary sequestration, right middle lobe atelectasis and unilateral absence of pulmonary artery. Treatment primarily consists of surgical correction which includes direct anastomosis of the scimitar vein to the left atrium or reimplantation of the anomalous vein into the right atrium with baffle insertion to redirect the flow into left atrium. Indications for surgical correction include presence of left to right shunt exceeding 50% and lung sequestration with recurrent right sided chest infections [5]. We have reported the case of an asymptomatic adult with scimitar syndrome diagnosed incidentally on routine CXR. Simple and traditional investigation tools like CXR if interpreted with vigil can aid in prompt diagnosis and management of patients even in absence of clinical symptoms.

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