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RESEARCH ARTICLE

SCIMITAR SYNDROME IN A TYPE 1 DIABETES MELLITUS: A CASE REPORT

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HRCT: High Resolution Computed Tomography

ABSTRACT

Scimitar syndrome or pulmonary venolobar syndrome is a rare, complex and variable cardio-pulmonary malformation, characterized by partial anomalous pulmonary venous connection. It can present in neonatal period as well as later in life. We present a case of 13 year old female who presented with DKA in type 1 diabetes mellitus and was diagnosed with pulmonary venolobar syndrome. Considering the rareness of the syndrome and its unusual form of presentation, this case is reported. Scimitar syndrome is a congenital deformity which consists of abnormal right side pulmonary venous drainage in the inferior vena cava, right lung hypoplasia, dextraposition of heart, and anomalous systemic arterial supply from aorta or one of its branches to the right lung. This syndrome has varied presentations, from an asymptomatic state to severe pulmonary hypertension and/or heart failure. A 13 year old female child presented with vomiting, abdominal pain & breathlessness and was diagnosed with DKA who was a known case of type 1 diabetes mellitus on insulin therapy. Detailed examination and routine work up was done. Chest skiagram (PA) showed scimitar sign formed by anomalous vein with right sided lung hypoplasia and shift of trachea and heart to the right side. Electrocardiography was normal except R/S >1 in V1. Transthoracic echocardiography showed no evidence of any septal defect or other abnormality. HRCT thorax was performed which showed, hypoplastic right lung with complete non visualization of the right upper lobe and anomalous drainage of the right middle & lower lobe veins into the sub-diaphragmatic, supra hepatic portion of the inferior vena cava. No medical management or surgical interventions was required as patient was asymptomatic.

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INTRODUCTION

"Scimitar syndrome" was first described by Chassinat in 1836 (Dupuis *et al*., 1992; Canter *et al*., 1986 and Oakley, 1984), is a congenital deformity which consists of abnormal right sided pulmonary venous drainage in the inferior vena cava, right lung hypoplasia, dextraposition of heart, and anomalous systemic arterial supply from aorta or one of its branches to the right lung (Zagol *et al*., 2006). This syndrome has varied presentations, from an asymptomatic state to severe pulmonary hypertension and/or heart failure. Neonatal symptomatic scimitar syndrome can be associated with other congenital deformities like atrial septal defect (most common), ALCAPA (Anomalous origin of left coronary artery from pulmonary artery), coarctation of aorta, tetralogy of fallot, ventricular septal defect (Roehm *et al*., 1986 and Godwin *et al*., 1986).

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About half the patients with Scimitar syndrome are asymptomatic or midly symptomatic when the diagnosis is made, despite varying degrees of pulmonary hypoplasia and pulmonary artery hypertension. Neonates have severe symptoms and worse prognosis while older children come to light because of recurrent respiratory infections, a murmur or an abnormal chest radiograph (CXR). Although the diagnosis can frequently be made on a CXR, further imaging is needed to confirm the diagnosis and demonstrate other associated abnormalities (Canter *et al*., 1986 and Oakley *et al*., 1984). 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 (Clements *et al*., 1987; Clements *et al*., 1987 and Fraser *et al*., 1989).

Case Report

A 13 year old female child presented with vomiting, abdominal pain & breathlessness and was diagnosed with diabetic ketoacidosis who was a known case of type 1 diabetes mellitus on insulin therapy. On examination patient was comfortable

and her vitals stable. Routine work up was done. Chest skiagram (PA) showed right hemithorax white-out & scimitar sign formed by anomalous vein with right sided lung hypoplasia and shift of trachea and heart to the right side. Electrocardiography was normal except R/S >1 in V1.

Figure 1. The name scimitar comes from the anomalous pulmonary vein that courses along the right cardiac margin as a curvilinear shadow and is said to resemble a "scimitar," or Turkish sword[8]

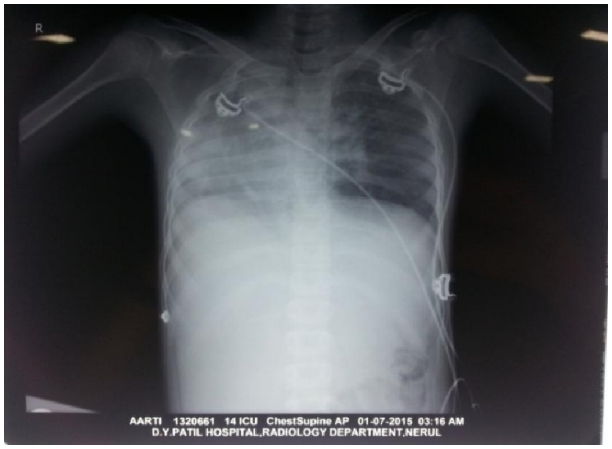

Figure 2. Chest X-ray was clearly suggestive of right lung hypoplasia with shift of the mediastenum to the right side & scimitar sign (arrow)

Transthoracic echocardiography showed no evidence of any septal defect or other cardiac abnormality. HRCT thorax was performed which showed, hypoplastic right lung with complete non visualization of the right upper lobe and anomalous drainage of the right middle & lower lobe veins into the sub-diaphragmatic, supra hepatic portion of the inferior vena cava.

Figure 3. HRCT confirmed, hypoplastic right lung with complete non visualization of the right upper lobe and anomalous drainage of the right middle & lower lobe veins into the sub-diaphragmatic, supra hepatic portion of the inferior vena cava

Patient's DKA part was managed with I/V fluids, insulin, potassium chloride and sodium bicarbonate in the ICU setting and patient was then shifted to general wards and then discharged with an uneventful course. As scimitar syndrome was an incidental finding and patient didn't have any complaints regarding it, treatment was not required.

DISCUSSION

Scimitar syndrome is described in three forms.

1) In the Infantile form patients develop severe pulmonary hypertension, cardiac failure and cyanosis during infancy.
2) The adult form patients are generally asymptomatic, but recurrent pneumonia, mild exertional dyspnoea and haemoptysis may be reported.
3) The third form is associated with other complex cardiac malformations and is more significant clinically (Dupuis *et al*., 1992).

The characteristic abnormality is anomalous pulmonary venous return of part (PAPVR) or the entire right lung to the IVC, though drainage into the hepatic vein or the portal vein can occur. Isolated partially anomalous pulmonary venous return to the inferior caval vein is also called "incomplete Scimitar syndrome". In the infantile form, the anomaly is usually associated with a variety of thoracic abnormalities and a high proportion of other vascular malformation. PAPVR of the right lung is usually associated with an ASD and an intact atrial septum is exceptional. Approximately 10-15% of patients with ostium secundum ASD have PAPVR (Gotsman *et al*., 1965).

Other cardiac malformations include coarctation of aorta, Tetrology of Fallot's, patent ductus arteriosus and ventricular septal defect (Roehm *et al*., 1986 and Godwin *et al*., 1986). The adult form is characterised by a small shunt with minor symptoms and lack of associated anomalies.

Classic appearance on chest radiograph-the scimitar sign appears because the anomalous venous connection causes a curvilinear shadow adjacent to the right heart border that resembles a curved Turkish sword. However, in some, when the scimitar vein is masked by the overlying cardiac shadow, other modalities are required. In present case, the classical scimitar sign is visible of chest radiogram (Fig 2). Doppler US conventional X-ray angiography has been traditionally considered to be the most accurate method for pulmonary venous abnormalities though current MR technology enables excellent visualization of vascular anatomy. Cine MRI10 and 3-D contrast enhanced MR angiography (Ferrari *et al*., 1998) provides a non-invasive diagnostic technique in the evaluation of anomalous pulmonary venous return. Surgical treatment of Scimitar syndrome may be required in the symptomatic patient, with other associated cardiac abnormalities or when left to right shunt is greater than 2:1 (Lucas *et al*., 2003).

Owing to its rarity, requirement of high end diagnostic testing and mostly asymptomatic state in adult patients, Scimitar syndrome might go unnoticed in many patients.

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