Scimitar Syndrome
A Rare but Important Condition to Diagnose

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Abstract

Scimitar syndrome is a rare but well defined disorder, involving anomalies of lung and pulmonary vein1. Children may present with tachypnea early in life or with repeated chest infection at any age. It is important to diagnose or at least to rule the syndrome as management depends upon the severity of different components. Treatment may be medical, surgical or combined in majority of cases. A case is described here which was fully investigated and family was counseled regarding disease, its prognosis and long term management.

Keywords: Scimitar syndrome, bronchiectasis, diagnosis, prognosis, management

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Introduction

Scimitar syndrome is a rare constellation of cardio-pulmonary anomalies[1]. There is hypoplastic right lung which is drained by anomalous pulmonary vein into inferior vena cava instead of left atrium. There may be other associated cardiopulmonary anomalies as well e.g. aberrant right pulmonary arterial supply directly from Aorta. Reported incidence is 1-5/100000 from different studies[2]. Most of the time there are case reports or case series however a few review articles and large studies are also available[3]. Few studies reported associated co-morbidities like Down syndrome or VATER association etc. VATER association consists of Vertebral anomalies, Anal atresia, Tracheosophageal atresia, Renal and Radial anomalies[4,5].

It is important to consider Scimitar syndrome in differential diagnosis, if a neonate presents with tachypnea or a child with bronchiectasis later in life. Right lung haziness and dextroposition of heart on x-ray should warrant full battery of investigations. Many cases need surgical intervention by cardiac or cardio thoracic surgeon[6,7]. There should be multidisciplinary; a well-coordinated team consists of pediatric pulmonologist or a pediatrician with special interest in pulmonology, cardiologist, cardiothoracic surgeon and physiotherapist should take care of child. Prognosis depends on the type and severity of anomalies. High pulmonary pressure, associated intra-cardiac anomalies and abnormal arterial connections carry poor prognosis[2,6].

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Case Report

Eight year old boy was referred from Khuzdar, Balochistan for the workup of repeated chest infections and 'dextrocardia' found on chest x-ray.

His parents are first cousins, he has three brothers and one sister, and all are healthy. He was born normally at home but was admitted in hospital on second day of life. Parents were told he had chest infection. His second admission was at one year of age, again with chest infection. After that he never admitted but had repeated chest infections

especially during cold weather. His admissions were very short, less than a weak long and response to antibiotics was always good. There is no other significant past medical or surgical history. He is partially vaccinated. Recently when he got fever and cough, local doctor asked for chest x-ray. There were signs of chest infection and heart shadow was found on right side. He was then referred to Karachi for further workup.

On examination he was an active boy, comfortable and cooperative Fig.1. There was no dysmorphology and his height and weight were on 25th centile. There was no anemia, cyanosis, clubbing, lymphadenopathy or any sign of distress. All the vitals were normal and oxygen saturation in room air was 99%. Chest auscultation revealed crepts over right lower lobe of lung, while normal heart sounds were found on the right side. Liver was palpable two cm below the right costal margin while spleen was not palpable. No other abnormality was found on physical examination.

Laboratory investigations showed normal blood counts. Hb 14g/dl, WBC 12.2 x 10^9/L, platelets 413 x 10^9/L, ESR 4 mm/1st hour. Chest x-ray showed heart and mediastinum shifted to the right side. Right lung field was hazy and right costo-pherenic angle blunt. Scimitar sign was seen i.e. abnormal right pulmonary vein draining into inferior vena cava, looking like curved Turkish sword (Fig. 2,3).

CT chest was suggested. High Resolution Computed Tomography (HRCT) showed bronchiectatic changes in right lower lung. Right lung and pulmonary artery appeared hypoplastic. An anomalous pulmonary vein was seen in right lower lobe draining into suprahepatic inferior vena cava. Mediastinal shift was present towards right side with hyperinflation of left lung. Horse shoe lung - an isthmus of pulmonary parenchyma arising from right lower lobe extending behind the heart, crossing midline and joined basal segment of left lower lobe. No lymphadenopathy was seen. (Fig 4,5). The ECHO of heart revealed dextroposition no abnormal connections and normal cardiac pressures. Our patient had five out of six usual components of Scimitar syndrome, mentioned below, except for lung sequestration. Aortography was not done to find out any anomalous arterial supply because he did not need any pulmonary surgery.

Family was counseled in detail. Child was thriving well, symptom free most of the time of year and response to oral antibiotics was good. However they were advised to bring the child, six monthly, for follow up review.

Discussion

Scimitar Syndrome is a veno-lobar syndrome i.e. a combination of abnormal pulmonary vein and lobe of lung. The disease can present from neonatal period to old age depending upon the severity of anomalies; or patient may remain asymptomatic and diagnosis is incidental only[8] Cooper[9] and Chassinat[10] first described the Scimitar syndrome in 1836 on post mortem examination. In 1949, Dotter[11] et al demonstrated the syndrome on live patient by catheter study and angiography. In 1956, Halasz[12] used term Scimitar syndrome. Scimitar is curved Turkish sword Fig. 2. Anomalous right pulmonary vein appears like a right sided curved structure on chest x-ray - Scimitar sign, which makes the basis of nomenclature Fig. 3.

Scimitar syndrome consists of multiple anomalies like right lung hypoplasia, dextroposition of heart, right lobe sequestration, horseshoe lung, pulmonary artery hypoplasia, anomalous pulmonary venous drainage from right lung to inferior vena cava (IVC) rather left atrium - Partial Anomalous Pulmonary Venous Return (PAPVR) and anomalous systemic arterial supply to right lung from Aorta or its branches. All these components may be documented by High Resolution Computed Tomography (HRCT) of chest with contrast Fig. 4,5.

These components lead to bronchiectasis and pulmonary hypertension. Child may present with repeated chest infection, like our patient, or with cyanosis and distress when there is pulmonary hypertension. Bronchiectasis is irreversible abnormal dilatation and distortion of bronchial tree. There is difficulty in clearing secretions leading to recur-

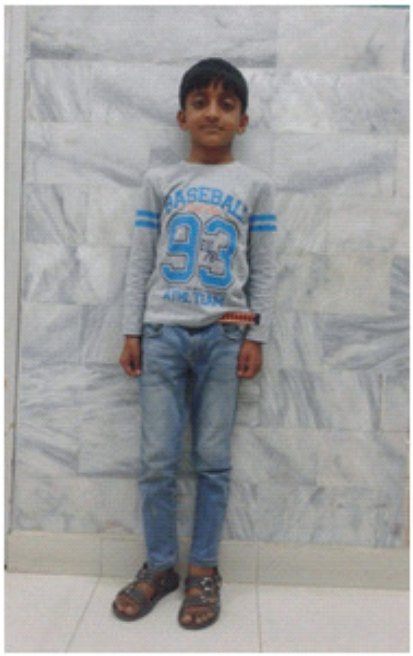

Fig. 1. Patient with Scimitar syndrome

Fig. 2. Scimitar is a Curved old Turkish sword

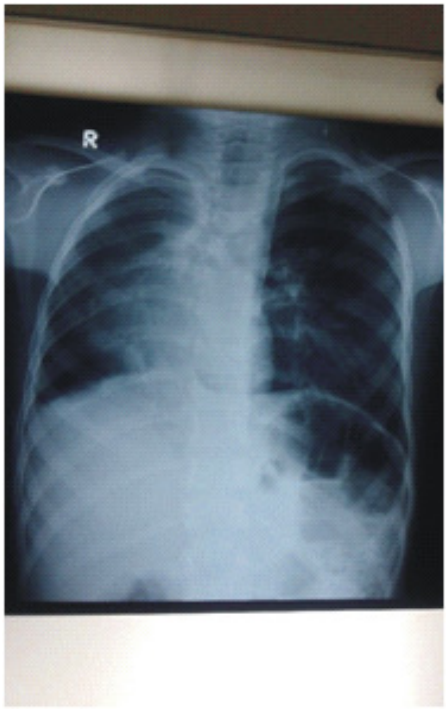

Fig. 3. Chest x-ray Dextropostion of heart, right lung haziness and Scimitar sign

Fig. 4. High Resolution Computerized Tomograph (HRCT), longitudinal sections

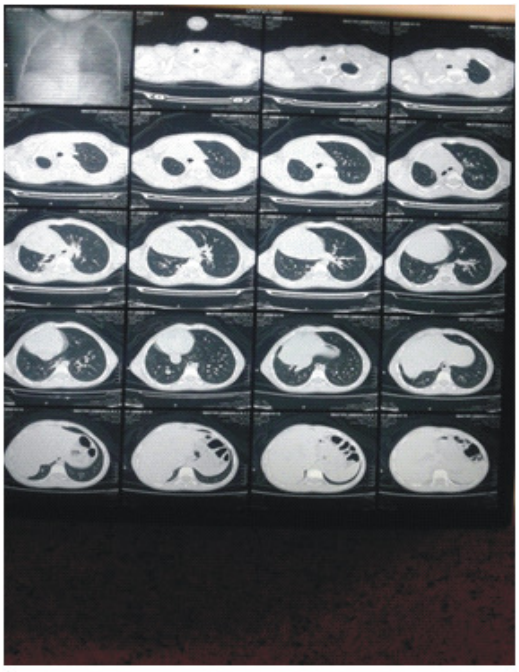

Fig.5. High Resolution Computerized Tomograph (HRCT), transverse sections

rent infections. Common organisms colonizing the respiratory tree are - Streptococcus pneumoniae, Haemophilus influenzae, Moraxella catarrhalis and Mycoplasma pneumoniae.

Differential diagnosis which must be kept in mind include causes of bronchiectasis such ascystic fibrosis, Kartagener syndrome, foreign body aspiration, immunodeficiency, Tuberculosis and complicated pneumonia.

Patient was investigated according to presentation to find out underlying anomalies or any other association. Complete blood count, Erythrocyte Sedimentation Rate (ESR), CReactive Protein(CRP) and chest x-ray are basic investigations. HRCT is gold standard investigation for bronchiectasis, while cardiovascular anomalies can be delineated by Magnetic Resonance Angiography (MRA)[13]. ECHO was done to rule out abnormal connections, blood flow and to measure pressures. Expert anomaly ultrasound scan can also detect the syndrome antenatally[14].

Management depends upon presentation and underlying pathology. Repeated chest infections can be treated with antibiotics like Amoxicillin and Clavulanic acid. In severe persistent infections, prophylaxis can be given either as oral marcolides or nebulized tobramycin, colistin or aztreonam etc. In case of thick secretions, normal or hypertonic saline nebulization can help to thin the mucus. Bronchodilators may be needed in case of wheeze. Chest physiotherapy and postural drainage are helpful to physically drain the secretions. These children need yearly influenza vaccine at the beginning of cold weather in October and five yearly pneumococcal vaccine. Surgery is needed in case of sequestered lobe, either ligation / embolization or pneumonectomy. Cardiac surgery to re-implant the anomalous vessel into left atrium is indicated in case of pulmonary hypertension or if left to right shunt is > 2:1[3,15].

Scimitar syndrome is a rare condition, but recognition is important because child needs multi disciplinary input from pulmonologist, cardiologist, cardiothoracic surgeon and physiotherapist etc.

Ethical concerns

Father permitted thepublication of the data and pictures.

Conflict of interest

Author has no interest other than contribution in academic and clinical pediatrics.

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