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Case Report

Paediatrics Section

Scimitar Syndrome Presenting as Recurrent Pneumonia: A Paediatric Case Report

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ABSTRACT

Recurrent pneumonia in children is frequently associated with congenital heart disease, developmental delay, anatomic abnormalities and cystic fibrosis. The challenge for physicians managing recurrent pneumonia lies in identifying underlying cause which if treated early will reduce further consequences. The identification of risk factors and adherence to immunisation schedules are essential to reduce disease burden and the mortality. Presenting a two year six-month-old male child, with history of recurrent admissions for pneumonia, the cause for which was not identified and now diagnosed as Scimitar syndrome, which is found to be as a rare congenital heart disease, described as a variant of Partial anomalous pulmonary venous return, in turn leading to left to right shunt. The condition ideally requires an very efficient interdisciplinary professional team approach which involves a Pediatrician, trained Pediatric cardiologist, experienced cardiothoracic surgeon and a radiologist. Early intervention can significantly reduce morbidity and improve outcomes.

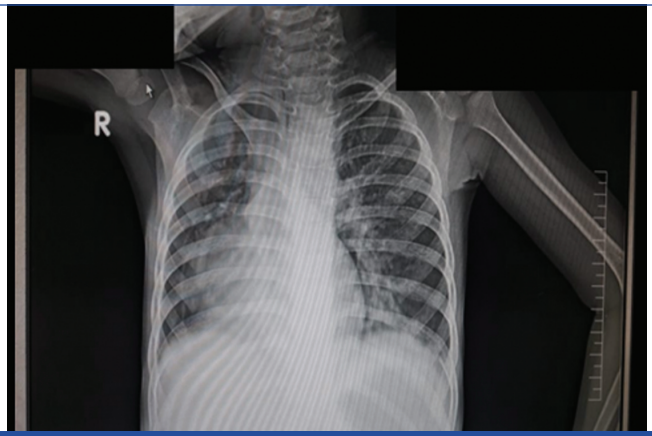
Keywords: Cardiac dextroposition, Congenital heart defect, Pulmonary vein, Scimitar sign, Venous return

CASE REPORT

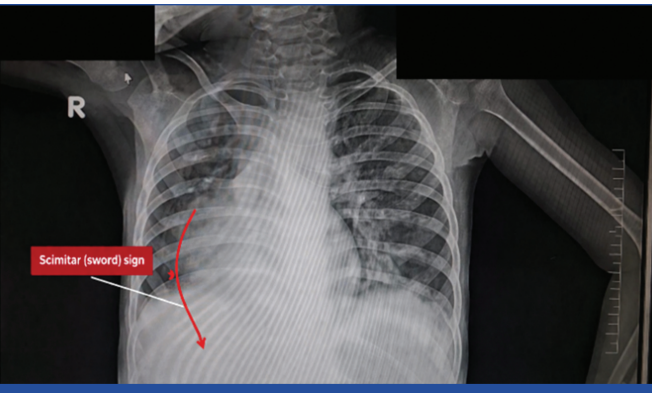
A two-year-six-month-old male child, presented to the Paediatric department with chief complaints of fever, cough and fast breathing since three days, had received nebulisation at a local hospital. Fever was continuous, high grade, not associated with chills or rigors and relieved on medications. There were no rashes, and wet cough was associated with post-tussive vomiting. Baby was first born, completely immunised and was developmentally normal. There was no significant family and birth history. However, there was a significant past history, in the form of recurrent admissions in the past for fever and cough. First episode being at the age of six months and four such episodes so far, each lasted for three to five days. Child was admitted and received oxygen, nebulisation and Intravenous medications every time and last episode occurred at the age of two year two months.

On examination, child was sick, lethargic, febrile Temp 100.0°F (37.8°C) fast breathing present with RR of 66/min and HR of 110/min, oxygen saturation in room air was 90%. On auscultation, wheeze and crepitations were heard and heart sounds were more on right-side. The provisional diagnosis was a case of recurrent pneumonia and congenital heart disease with Mesocardia/dextrocardia. The patient received symptomatic and supportive treatment such as piperacillin tazobactam (100 mg/kg/dose) 1.2 gm, three times daily intravenous, Ambroxol cough syrup 3.5 mL twice daily fluid rehydration and antipyretic Syp Paracetamol (250 mg/5 mL) 3.5 mL QID.

Investigations showed that except for C-Reactive Protein (CRP) elevated to 22 mg/L, which was above normal level, white blood cells, red blood cells, neutrophils, haemoglobin, and platelets were all in the normal range. Blood culture was sent and revealed no growth. Chest x-ray revealed dextroposition of the heart and a curved shadow on the right mid lung zone extending to the diaphragm, the scimitar sword sign as shown in [Table/Fig-1,2]. Differential diagnosis considered was Scimitar syndrome, pulmonary sequestration and bronchopulmonary mass. Echocardiography showed findings of dextroposition of the heart and partial anamolous vein draining from the right lung into the Inferior Vena Cava (IVC), dextraposition of the heart. No congenital malformation such as atrial septal defect, ventricular septal defect, and patent ductus arteriosus was found as shown in [Table/Fig-3]. Hence, we made a final diagnosis as Scimitar Syndrome.



[Table/Fig-1]: Chest X-ray of the patient.

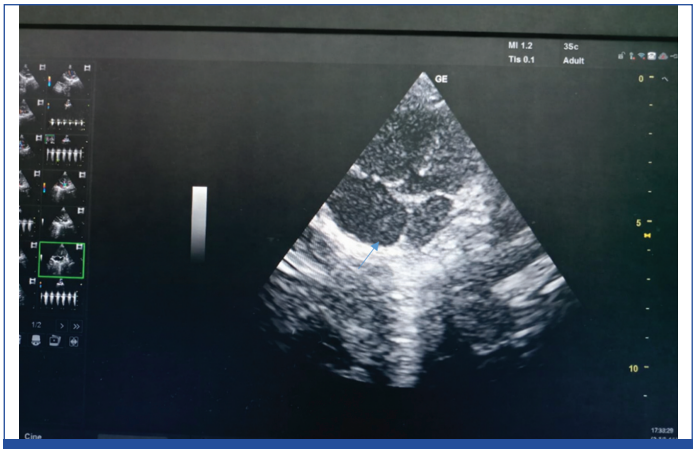


[Table/Fig-2]: Chest X-ray of the patient showing scimitar sign (sword).

The patient was suggested, to go for Computed Tomography (CT) lung and further detailed cardiac evaluation and management, but patient attenders were not willing to continue treatment due to personal reasons.

DISCUSSION

Scimitar syndrome,otherwise known with different terminologies like Halasz syndrome, congenital venolobar syndrome, sometime



[Table/Fig-3]: Echocardiography showing anomalous pulmonary venous drainage to the IVC with dextroposition.

mirror image lung syndrome, is a rare congenital heart disease and its incidence is found to be 1-3 per 1 lakh live births [1]. It is often viewed as a variant of a partial anomalous venous return from the lungs, resulting in left to right shunt, with a very unique anatomic feature as it drains, that resembles a back sword or also known as saber with a blade which is curved, known by the name as Turkish sword, the Scimitar. The characteristics that are featured distinctly in Scimitar syndrome include partial or sometime total anomalous venous drainage, that is curved from the right lung to the IVC, and most often associated with hypoplasia of variable degree of right lung and pulmonary artery, the heart will be dextroposed and there will be anomalous blood supply to the ipsilateral lung from systemic circulation. The other coexisting findings sometimes may have Atrial septal defects and Aortopulmonary collaterals also.

George cooper, was the first who described Scimitar, as he was conducting an Autopsy in a 10 month old baby in 1836, following which in 1949, Dotter etal described the first imaging diagnosis by cardiac catheterization and later in 1950 Drake and Lynch performed the first surgical intervention, by resecting the right lower lung [2]. The condition is attributed, as a result of an embryological error during the early embryogenesis of lung bud development in its fundamental stage [3].

Scimitar syndrome can have variable presentation, which ranges from asymptomatic patient with just an isolated finding, often with benign outcome to the other end presenting as congestive cardiac failure in two different age groups as infantile form and childhood or adult form. A three-year-eight-month-old girl who presented with prolonged cough, fever, and recurrent respiratory infection. Investigations showed elevated CRP and sputum growth of *Haemophilus influenzae*, suggesting a superadded infection. Radiological imaging revealed a characteristic anomalous pulmonary vein draining into the IVC with right lung hypoplasia. These findings confirmed the diagnosed condition as scimitar syndrome and child was discharged after 10 days of treatment and advised follow-up with cardiology department [4].

A retrospective observational study of Paediatric group of patients who were diagnosed as Scimitar syndrome was conducted, where patients underwent systematic evaluation including medical and family history, chest X-ray, 12-lead electrocardiogram, echocardiogram, angiography and/or Computed Tomography (CT); or magnetic resonance angiography. Around ten Pediatric patients with scimitar syndrome were actually included. The median age at diagnosis was 10.4 (0.1-150.2) months and the median follow-up time was 7.7 (1.3-15.3) year [5].

The cardiac defects that are commonly associated with this syndrome include atrial septal defect accounting for (80%), patent ductus arteriosus about (75%), ventricular septal defect around (30%) and pulmonary vein stenosis is found in (20%); but sometimes one can also find association of aortic arch hypoplasia or coarctation, tetralogy of Fallot, and hypoplastic left heart syndrome with this condition. The infantile form is often found to correlate with higher co-morbidities like multiple congenital defects of diaphragm viz (accessory diaphragm, eventration or partial absence of the diaphragm; sometimes phrenic cyst; horseshoe lung; and also pericardial absence), and also is usually complicated with severe pulmonary hypertension which presents as congestive heart failure, contributing to significant mortality in the patients [1].

Patients usually may present with respiratory tract infections with probable involvement of right lower lobe, features of bronchiectasis and interstitial pulmonary disorders also seen in some cases The childhood/adult variant invariably tends to be milder, form of presentation, associated with lower mortality rates. Patients sometimes may present with just an incidental finding of an unexplained dilatation of the right heart, as well as occurrence of frequent pulmonary infections predominantly involving the right side of the lung as in the present case. On Physical examination, one can characteristically demonstrates a shift of the heart sounds and find the cardiac impulse to the right, along with a systolic murmur [6,7].

Scimitar syndrome is a rare clinical condition, the diagnosis is primarily by imaging, where the chest X-ray reveals a shadow of the descending pulmonary vein along the side of right cardiac border, a hypoplastic lung, and the dextroposition of the heart. There can be shifting of mediastinum along with features of atelectasis or pulmonary agenesis. To start the work-up for diagnosis of this syndrome, Echocardiography is always a preferred imaging study and foetal echocardiography also aids with the prenatal diagnosis by helping in visualisation of an obstructed pulmonary venous pathway, along with confluence behind the right atrium and a usual vertical vein is also seen. Three-dimensional CT and cardiac-gated Magnetic Resonance Imaging (MRI) are to be considered as the best diagnostic modalities, often providing an excellent anatomical structure delineation of the abnormal pulmonary vein, including its course, the connection, and final draining path. Cardiac catheterisation and angiography are one of the most useful diagnostic studies, for confirming the diagnosis of scimitar syndrome, but still not needed at all times [6-8]. Management includes treatment of infections, heart failure and growth. The approaches for surgical intervention for correction of scimitar syndrome varies case wise,its clinical presentation,anatomical and pathological findings, Cardiac surgeon preference. Surgical intervention involves either resecting the portion of the lung that is drained by anomalous vein or some times also corrective procedure which re routes the blood flow [1]. Lobectomy or sometimes pneumonectomy is often indicated in a specific group of patients who presents with recurrent pulmonary infections,persistent hemoptysis, diffuse bronchiectasis, if there is marked hypoplasia of the right lung or any associated thrombosed –intraarterial baffles [3,6,9].

CONCLUSION(S)

Scimitar syndrome a rare congenital heart lung syndrome with varied presentation in different age groups, that is to be approached by Interprofessional team of doctors including Paediatrician,paediatric cardiologist, trained cardiac surgeon and a experienced radiologist. It this condition is not diagnosed at early age, especially in infantile form, they can have very poor outcomes. An early suspicion and confirmation of diagnosis with the available advance cardiac imaging facilities and early surgical interventions contributes to reduced rate of morbidity and mortality after the corrective surgery and long term follow up and monitoring.

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