SCIMITAR SYNDROME WITH ABSENT LEFT KIDNEY- A CASE REPORT

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ABSTRACT

Scimitar syndrome is a rare congenital anomaly consisting a part of right pulmonary venous return to the inferior vena cava. There is a clear bimodal presentation of this syndrome with either infantile presentation or a paediatric/adult form. The infantile variant is severe form with significant mortality. The adult form is less severe and may be asymptomatic. The case we are reporting presented with respiratory distress , hypoplasia of the right lung, dextroposition of the heart and absent left kidney.

Key words: Scimitar Syndrome, Dextro Position of Heart, Hypoplasia of Lung.

INTRODUCTION

Scimitar syndrome is a rare association of congenital cardio-pulmonary anomalies that has been reported in 3%-6% of patients with partial anomalous venous connection. In these patients, some of the pulmonary veins drain into the upper part of the inferior venacava, [1] either above or below the diaphram[2] and usually drain the lower and sometimes the middle lobe of the right lung (79%) or the whole right lung (21%)[3]. The right lung is frequently hypoplastic and receives blood supply from the systemic arteries, mainly the thoracic or abdominal aorta, and this supply is usually to the lower lobes.[4] Neonates have severe symptoms and worse prognosis while older children come to light because of recurrent respiratory infections, heart murmur, or an abnormal chest radiograph.[5,6]

We report a case of 2 month old female infant who presented with pneumonia, failure to thrive, absent left kidney and Congenital Talipes Equino Varus (CTEV) of the right lower limb.

CASE REPORT

A two months old female infant presented with history of hurried respiration and chest in-drawal of 4 days duration. There was no history of cyanosis, lethargy or seizures. Baby was exclusively breast fed.

On examination, infant had tachycardia, tachypnoea and features of failure to thrive. Respiratory system revealed chest retractions, decreased breath sounds over right interscapular and right infrascapular region, crepitations over left infrascapular region. Cardiovascular examination revealed dextrocardia and patient also had CTEV of right lower limb. Laboratory investigations revealed normocytic normochromic anaemia and rest of the test results came back normal. Chest radiograph showed a small right hemi-thorax and an anomalous pulmonary vein on the right side (Fig.1).Scimitar syndrome was suspected on the basis of chest radiograph. To confirm the diagnosis, computer tomography (CT) of the thorax was done, which showed right sided volume loss of the lung parenchyma and dextroposition of the heart (Fig.2). Dilated vascularity was detected in the right lower lobe with homogenous enhancement. Abdominal ultrasonography revealed absence of left kidney. Echocardiography was normal except for the dextroposition of the heart. CT angiogram is confirmatory, as the facility is not available in our institute, with above findings itself, a diagnosis of congenital pulmonary veno-lobar syndrome also known as Scimitar Syndrome was made.

DISCUSSION

Scimitar syndrome is a rare constellation, estimated to occur in 2 out of 100,000 births with a 2:1 female preponderance.[7] This anomaly was first described by Cooper[8] in London in 1836 during an autopsy of an infant. Note was made of dextroposition of the heart and hypoplasia of the right lung in this specimen. The first diagnosis in a live (and asymptomatic) patient was made in 1949 by Dotter et al[9] on cardiac catheterization.

The scimitar (or Turkish sword; fig1) sign refers to the half crescent described by the descent of the anomalous

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pulmonary vein (the tip of the crescent points inferiorly and medially to the diaphragm and right heart border junction). The concavity of the crescent is adjacent to the right heart border. Tributary veins may be seen converging on Scimitar vein (SV) as the "hilt" of the dagger.

However, this sign is often absent. Reasons cited for the absence include the hypoplasia of the right lung and resultant abnormal positioning of the heart and prominent thymic shadow in infants.

It is known that in the course of normal lung development, pulmonary venous drainage to the left atrium is in place by 11th week of gestation and as the advancing lung bud develops, its primary blood supply changes from a plexus derived from the post-branchial descending aorta to the portion of the sixth aortic arch that becomes the pulmonary artery,[10] a transition that is complete after the seventh week. Some insult presumably culminates in the failure of this "hand-over", resulting in the observed persistence of systemic arterial supply to the right lung from the abdominal aorta, and the underdevelopment of the right pulmonary artery and right lung.

Scimitar syndrome has a variable presentation based on the age at which the diagnosis is made. Infants typically present with a clinical picture of severe respiratory insufficiency and cardiac failure.[11] Pulmonary hypertension is a commonly associated problem. In older children and adults the diagnosis of scimitar syndrome is often made incidentally in patients who undergo chest radiography for diverse reasons.[12] Recurrent respiratory infections and heart murmur may be the mode of presentation.[13]

The diagnosis is usually established on chest radiograph that shows dextroposition of the heart and hypoplasia of the right lung.[14]CT and magnetic resonance imaging (MRI) show anatomic findings of scimitar syndrome. For detecting cardiac and pulmonary anomalies, CT angiography is recommended.

Our patient had pneumonia and failure to thrive. Diagnosis of scimitar syndrome is usually made anytime between neonatal period till adulthood. Our patient also had other anomalies like absent left kidney which is never been reported before and CTEV. In general, infants presenting with heart failure have more associated anomalies and their prognosis is much worse.[14]The prognosis for older children is better, either with or without surgery. Treatment for symptomatic scimitar syndrome consists of surgical repair. Surgical repair seldom results in normal blood flow to the right lung but abolishes left to right shunt. Postoperative pulmonary venous obstruction is prevalent especially in infants.[15] Our patient showed significant improvement in clinical picture of pneumonia, hence she is on supportive care and regular cardiac follow up. In case of persistent pulmonary symptoms and failure to thrive, she would be an ideal candidate for surgical treatment.

Respiratory distress, right lung hypoplasia and dextroposition of heart should alert the clinicians towards the possibility of this syndrome.

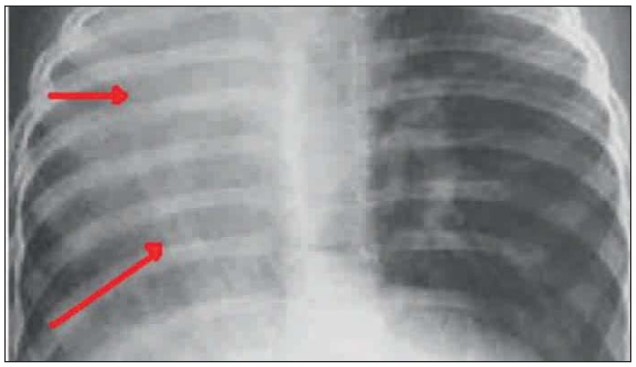

Fig 1. chest x ray showing small right hemithorax, anomalous pulmonary vein on the right side and dextropositon of the heart

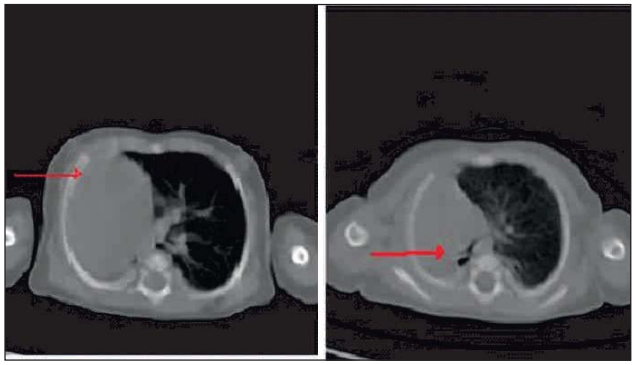

Fig 2. CT chest showing right sided volume loss, mediastinal shift to the right side and dextropositon of the heart.

CONCLUSION

Scimitar Syndrome is a rare condition which usually presents with severe respiratory insufficiency and cardiac failure. This article shows the diversity of the disease since our patient also had associated CTEV and absent left kidney.

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