Scimitar syndrome with Renal Hypoplasia

S. M. Gour[a], A. T. Nikhare[b], R. Munje[c]

Abstract

We report a 10 year young child with gradually progressing breathlessness who was diagnosed to have "Scimitar syndrome with Right Renal Hypoplasia" on the basis of history, clinical features, Computed tomography and CT Angiography.

KEYWORDS : Scimitar syndrome, Renal Hypoplasia, Anomalous Pulmonary Venous Drainage, Pulmonary Hypoplasia,

Introduction

Scimitar syndrome is a form of anomalous pulmonary venous drainage (APVD). APVD implies partial or total failure of the pulmonary veins to reach the left atrium. Instead, pulmonary venous drainage is anomalously connected to systemic vein/s, typically to the superior or inferior vena cava (SVC or IVC) or directly to the right atrium (RA).The syndrome associated with PAPVR is more commonly known as *Scimitar syndrome* after the curvilinear pattern created on a chest radiograph by the pulmonary veins that drain to the inferior vena cava.

Case Report

A 10 year young male presented with complaints of gradually progressive dyspnoea of Grade 2 since 3 months and Cough on and off since 3 months. Patient attended a Cardiologist for his complaints and was diagnosed to have Severe Pulmonary Hypertension and was then referred to a Chest Physician. There was no history of fever, hemoptysis. No history of Pulmonary Tuberculosis in past. There was no history of previous hospitalization or any major illnesses in the past. Patient was a full term baby, immunized for age with all developmental milestones normal.

On examination, his vitals were stable& had no cyanosis or clubbing. Except for occasional crepitations respiratory system examination revealed normal findings.

On cardiovascular system examination, heart sounds were heard over the sternum instead of the precordial area. Abdomen examination did not reveal any abnormality.

Laboratory evaluation revealed nothing significant. Routine Hemogram was normal. Peripheral smear showed normocytic, normochromic red cells. Sputum was negative for Acid fast bacilli (AFB).

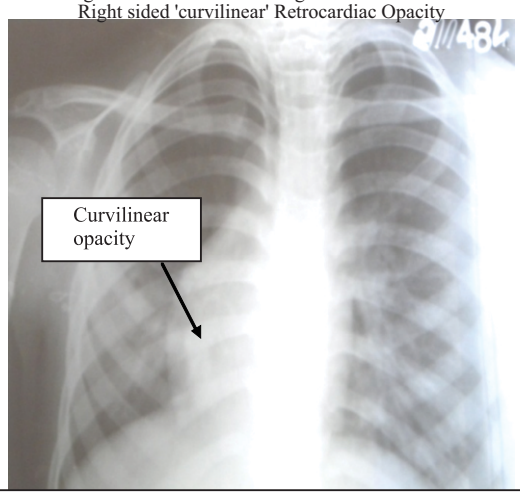
Chest X-ray showed Dextrocardia with a right sided retrocardiac opacity **(Fig 1.)**

a Assistant Professor, b senior Resident , c Professor
Dept. of Chest & TB, IGGMCH, Mayo Hospital, C.A. Road, Nagpur 440018.

Address for correspondence

S. M. Gour
Email dr_smg2004@yahoo.com

Fig 1. CXR PA view showing Dextrocardia with Right sided 'curvilinear' Retrocardiac Opacity



X-ray PNS was normal. 2D-ECHO showed Meso-Dextrocardia with Severe Pulmonary Hypertension. Pulmonary Function test(Spirometry) showed a Forced Vital Capacity (FVC) of 75 % predicted with no obvious Restrictive pattern.

A Contrast Enhanced CT scan of Thorax & CT Pulmonary angiogram showed Dextrocardia with Absent Right sided Pulmonary Veins, **(Fig 2 & 3)**

Right lung draining through anomalous dilated veins into Inferior vena cava **(Fig 4,& 5),**

Bilobed right lung, Dilated Pulmonary Trunk suggestive of Pulmonary Hypertension, Enlarged right and left ventricles, Nonunion of both the brachio-ceplalic veins with independent drainage into the right atrium, Right Kidney small and compressed with compensatory hypertrophy of the left kidney (**Fig 6**).

DISCUSSION

The Scimitar syndrome, first described by Chassinat in 1836 is a rare but well described constellation of cardio-pulmonary anomalies.[1]

The incidence is estimated to be 1 to 3 per 1,00,000 births.[2] It occurs more commonly in females and is occasionally familial.[5]

Scimitar syndrome is a form of anomalous pulmonary venous drainage (APVD). APVD implies partial or total

Fig 2. CT Pulmonary angiography showing Dextrocardia

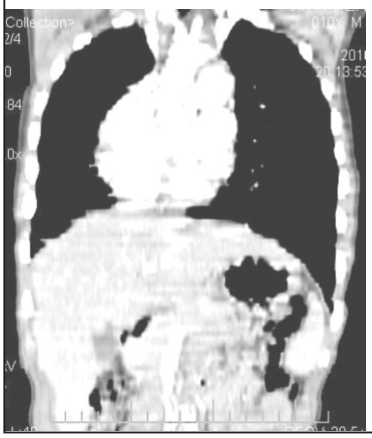


Fig 3. CT Angiography (lateral view) showing 'curvilinear opacity extending from above downwards into the diaphragm

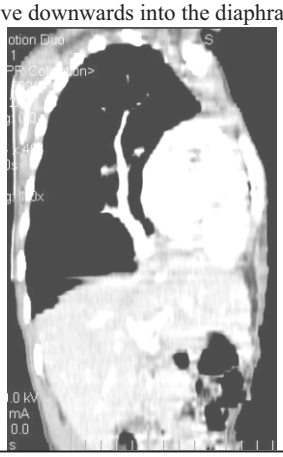


Fig 4. CT Pulmonary Angiogram showing the anomalous curvilinear opacity uniting with the I.V.C .

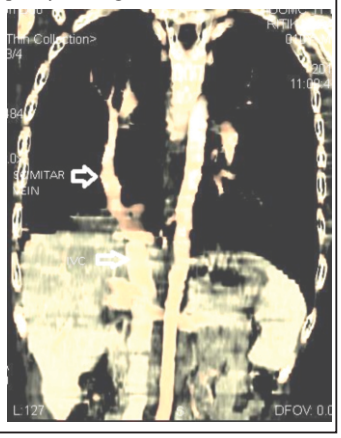
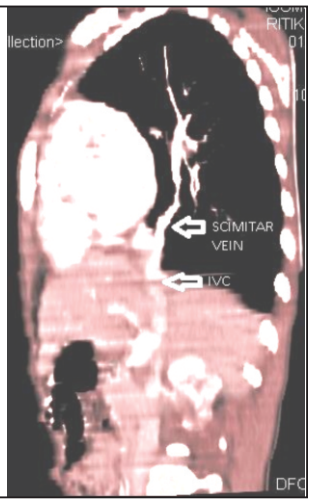
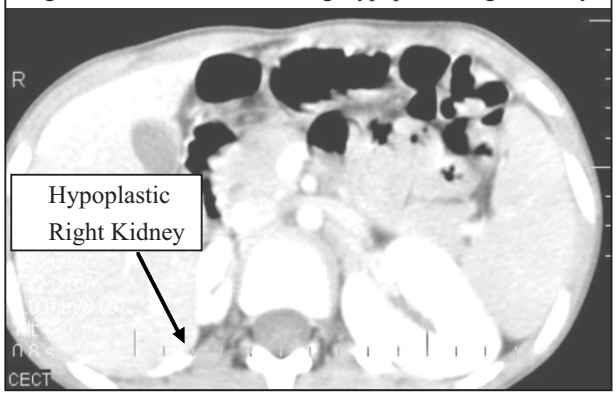


Fig 5. CT Pulmonary Angiogram (lateral view) demonstrating Scimitar vein draining into the



failure of the pulmonary veins to reach the left atrium. Instead, pulmonary venous drainage is anomalously connected to systemic vein/s, typically to the superior or inferior vena cava (SVC or IVC) or directly to the right atrium (RA).

Fig 6. CECT Abdomen showing Hypoplastic Right Kidney



Thus it is Hypoplasia of Right lung & anomalous pulmonary venous drainage from it to the Inferior vena cava.[3] The anomalous pulmonary vein most commonly drains into the Inferior vena cava below the level of Right Hemidiaphragm.[5]

Additional anomalies that occur with this syndrome include : [5,6]

1. Partial agenesis or hypoplasia of the right lung with bronchial isomerism
2. Diverticulum or hypoplasia of the right bronchial system.
3. Hypoplastic or absent pulmonary artery,
4. Anomalous systemic arterial supply to right lung from aorta or one of its branches,
5. Dextrocardia due to right lung hypoplasia with mediastinal shift
6. Bronchopulmonary sequestration,
7. Absence of inferior vena cava, and accessory diaphragm
8. Phrenic cyst
9. Horse-shoe lung
10. Oesophageal and gastric lung
11. Absence of pericardium
12. Other congenital cardiac malformations (25% of cases) including ASD, ventricular septal defect, coarctation of the aorta, tetralogy of Fallot, pulmonary stenosis, absent inferior vena cava with azygos continuation to superior vena cava and persistent left superior vena cava. Moreover, the APVD may also be partially obstructed, further contributing to pulmonary hypertension.

Some of the above anomalies were present in the present case, however some additional findings were

a. Right Kidney small and compressed with compensatory hypertrophy of the left kidney.

b. Dilated Pulmonary Trunk suggestive of Pulmonary Hypertension, Enlarged right and left ventricles,

c. Nonunion of both the brachioceplalic veins with independent drainage into the right atrium, Associated Renal Hypoplasia has not been reported in literature till now.

Though this finding of renal hypoplasia is unique it appears to be a cause rather than effect of APVD because Fetal urine is an important component of amniotic fluid during late gestation and contributes to lung growth. Also kidney is also a major source of proline. Proline aids in the formation of collagen and mesenchyme in the lung, thus explaining the severe pulmonary hypoplasia in renal agenesis and dysplasias.

There are unequivocally two forms of Scimitar syndrome in terms of clinical presentation: [1]

(1) an infantile syndrome associated with significant mortality and

(2) a child/adult presentation that is a milder form of the syndrome and in fact is frequently asymptomatic, with diagnosis being made incidentally because of radiographic abnormalities.

In this case ,it was an adult form as the child was of 10 years and presented with progressive dyspnoea secondary to severe pulmonary hypertension.

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