Case report

Scimitar syndrome

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Abstract:
A female child of age five attended our institution in Jan 2006 with repeated upper respiratory infection. Plain skiagram and CT examinations confirmed the case to be a Scimitar syndrome.

Key words: scimitar syndrome; hypogenetic lung syndrome; pulmonary venolobar syndrome

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Introduction

The scimitar syndrome is a rare congenital anomaly consisting of hypogenetic right lung with systemic venous drainage and arterial supply. The shape of the anomalous vein draining from right lung into IVC resembles a Turkish sword (scimitar) .Hence Halasz [1]used the name scimitar in 1936 and Neil [2] named it as scimitar syndrome in 1960.

Case report

A five-year-old female child presented at paediatric OPD of MIMS hospital in Jan 2006 with history of repeated respiratory tract infections since last 3 years and being treated previously for the same complaint at
several hospitals without any releive. Plain radiograph of the chest demonstrated shift of mediastinum to the right. A curvilinear dense opacity on the right side of the right cardiac border, is seen traversing below the diaphragm (Fig -1).

A CT angio in spiral mode with 3D reconstruction has been done

CT findings

CT angio showed the right descending pulmonary vein traversing below the diaphragm and draining into the IVC, which is dilated. The right atrium is prominent. Associated hypoplasia of
right lower lobe is noted. Pulmonary arteries appear normal. No associated pulmonary arterial or cardiac anomalies noted.(Fig 2,3)

A diagnosis of scimitar syndrome is made.

Fig 3 – CT angio showing right lower lobe vein (scimitar vein) draining to I.V.C.

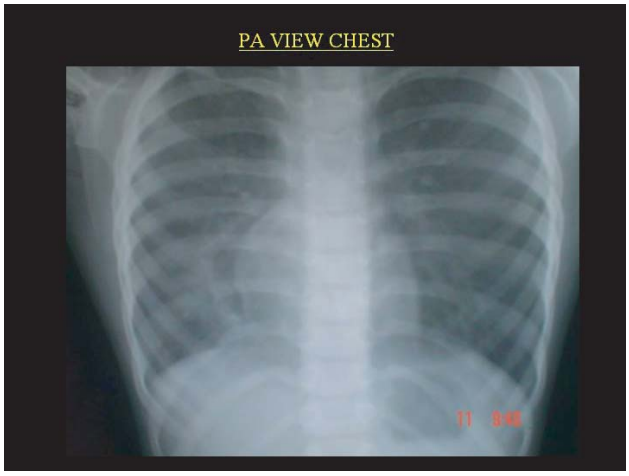

Fig 1 – Plain skiagram of chest showing shift of heart to right and the scimitar vein.

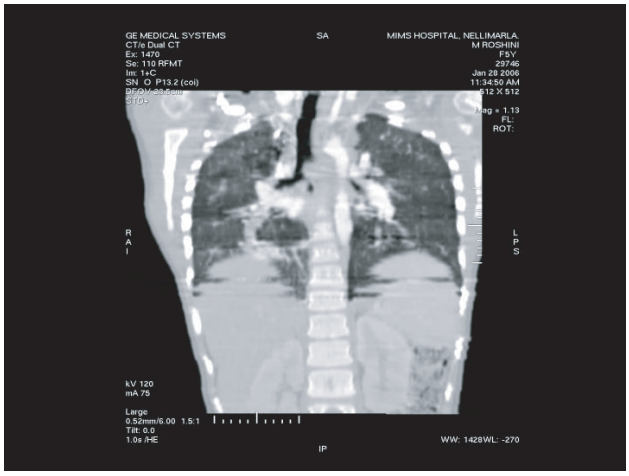

Fig 2 – CT angio showing hypoplastic right lower lobe.

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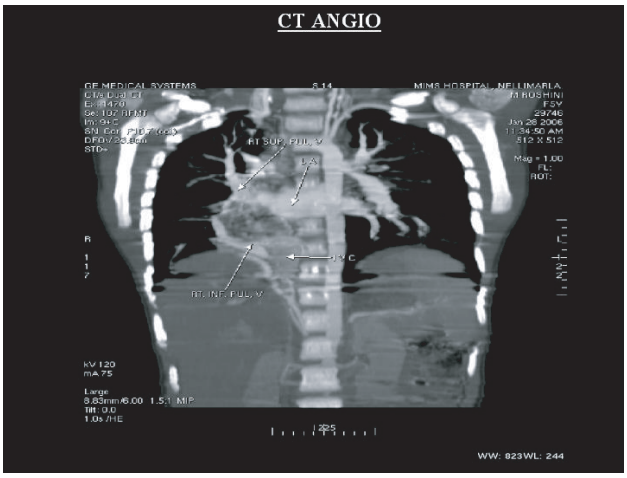

Fig 3 – CT angio showing right lower lobe vein (scimitar vein) draining to I.V.C.

Discussion:

This syndrome is also known as hypogenetic Lung syndrome, pulmonary venolobar syndrome, mirror venolobar syndrome, Mirror Image lung syndrome, Epibronchial right pulmonary artery syndrome, Venacava bronchovascular syndrome . As the anomalous vein in the right lower lobe resembles a Turkish sword which is known as a scimitar, hence the name of the syndrome. The incidence is about 3 in 1 lakh live births. It is common in Girls. Familial occurrence has been noted with an autosomal dominant inheritance with variable expression.. Persistance of early embroyanal connections between the post branchial pulmonary parenchyma & the primitive dorsal aorta results in systemic arterial supply.[3]

Scimitar syndrome has pulmonary , vascular involvement with associated abnormalities.[4]

Pulmonary component:

There may be hypoplasia, aplasia, agenesis of a lobe or a segment of a lobe.

There may be abnormal lobulation or error in segmentation.The reason why the right lung is affected is not known.

Vascular component:

The constant feature of this syndrome is the scimitar vein. The anomalous right pulmonary vein is seen draining below the diaphragm to the IVC , portal or hepatic vein. It lies lateral to the right cardiac border with a sickle shape (scimitar) . As the vein drains downward it becomes broader. In some cases it may not be visualized as it lies behind a dilated right atrium due to right sided shunt. The pulmonary arteries may be hypoplastic and there may be systemic supply to the right lung from an intercoastal artery or directly from descending aorta. In addition to classic findings other anomalies include absent pulmonary artery, broncho-pulmonary sequestration, absence of inferior vena cava and presence of accessory diaphragm [5]

Associated anomalies:

There may be :

Congenital anomalies of bony thorax

Hemi vertebrae

Accessory diaphragm

Ipsilateral hemidiaphragm.

Excessive extrapleural areolar fatty tissue - right side chest

Horse-shoe lung

Absent IVC

Left to right shunt-Mostly ASD

PDA , VSD

Manifestations of clinical syndromes are dependent on the size of the Scimitar vein draining to IVC resulting in right-sided hyper dynamic circulation. The infants with this syndrome may develop high blood flow to the hypoplastic right lung from anomalous systemic branches arising from aorta or intercostal arteries may result in large left to right shunt. Hence the clinical presentation is quite variable ranging from being asymptomatic to significant heart failure. Age of presentation may be anywhere from infancy to adulthood. When the patient presents in the infancy the symptoms are more severe and are associated with more anomalies [6]. In older children it may present as recurrent respiratory infection or a heart murmur.

Differential diagnosis:

The scimitar sign may not be apparent and is attributed to a very small right lung, and marked mediastinal shift, enlarged right atrium and small caliber of the scimitar vein. In such cases it has to be differentiated from Dextrocardia and Swyer James syndrome [7]

Right lower lobe atelectasis/ agenesis

Broncho pulmonary sequestration with arterial supply from abdominal aorta.

Congenital lobar agenesis.

Diagnosis:

In most cases the diagnosis is evident from plain x ray of chest which shows the classical scimitar vein. In cases the scimitar vein is not visible, multi sectional helical C.T angio can generate angiogram like images to demonstrate scimitar vein, pulmonary hypoplasia, and can easily differentiate it from other

conditions like pulmonary sequestration [8]

Other Radiological Methods Of Investigations

Aortogram may document aberrant artery from descending Aorta supplying the right lung

Pulmonary arteriography may demonstrate absent or hypoplastic right pulmonary artery. Venous phase will show the scimitar vein draining to the IVC, hepatic or portal vein.

Radionuclide perfusion scan can suggest abnormal arterial supply.

Radionuclide ventilation scans can give information regarding pulmonary hypoplasia or agenesis.

Doppler ultra sound may document the site of entry of the scimitar vein into the IVC, portal vein or hepatic vein and can verify venous flow.

Treatment and prognosis:

Symptomic cases may require surgical repair or mobilization of anomalous vein or excision of involved lung.

Embolisation of the scimitar vein appears to offer a satisfactory result. Where the interventional radiologist plays an important role in treatment.

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