Pictorial CME

Partial Anomalous Pulmonary Venous Return (PAPVR) - Scimitar Syndrome

Reena Mathur*, Avinash Gupta, Prateek Sihag***, Kalpana Manral******

*(*Professor and Head, **Professor, ***Assistant Professor, ****Resident,*
Department of Radio-diagnosis, JLN Medical College, Ajmer)

ABSTRACT:

Scimitar syndrome is a rare congenital anomaly. This syndrome is characterized by partial or complete anomalous pulmonary venous drainage of the right lung to the inferior vena cava. There is a characteristic abnormal radiographic shadow which descends along the right cardiac border (scimitar sign).

Keywords: *Scimitar Syndrome, Congenital Heart Disease.*

INTRODUCTION:

The term scimitar syndrome derives from the shadow created by the anomalous vein on the chest radiograph. This shadow extends from the lateral superior position of the right lung to a more medial location and increases in calibre as it descends toward the cardio-phrenic angle. The appearance closely resembles that of a curved Turkish sword or scimitar. This rare anomaly has an incidence of approximately 1 to 3 per 100 000 live births;3 the true incidence may be higher because many patients are asymptomatic. Functionally, scimitar syndrome resembles an ASD. The left lung is very rarely involved for unknown reasons.

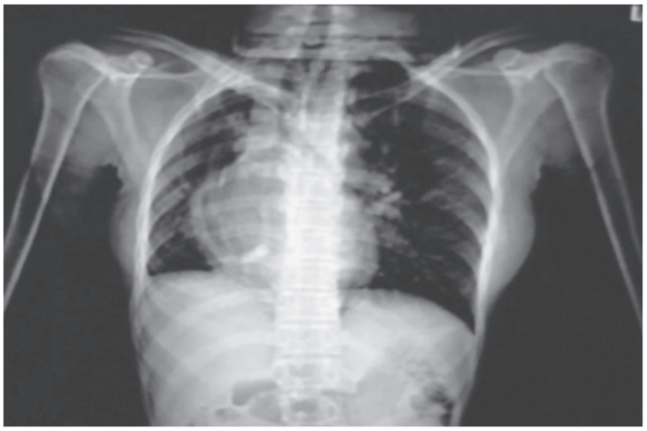
CASE PRESENTATION:

A 35 year old male patient presented with gradual onset shortness of breath on exertion for 3 months. The patient denied any history of chest pain, wheeze , fatigue. Also no history of tuberculosis, smoking , exposure to chemicals ,organic dust was present.

On clinical examination , patient was afebrile and comfortable in resting state. Blood pressure was within normal limit -120/70.

Plain radiograph

Upon taking a PA view of chest, right sided lung appeared small compared to left with ipsilateral mediastinal shift.A radiopaque tubular structure paralleling along the right heart border was seen (scimitar vein). A diagnosis of scimitar syndrome was made and futher imaging with contrast enhanced computed tomography of chest done.



Contrast enhanced computed tomography of chest

The contrast-enhanced CT of chest confirmed the anomalous venous return from the hypo-plastic right lung by a main scimitar vein into inferior vena cava just above the diaphragm.

Other associated abnormalities demonstrated on CECT examination of our patient were:

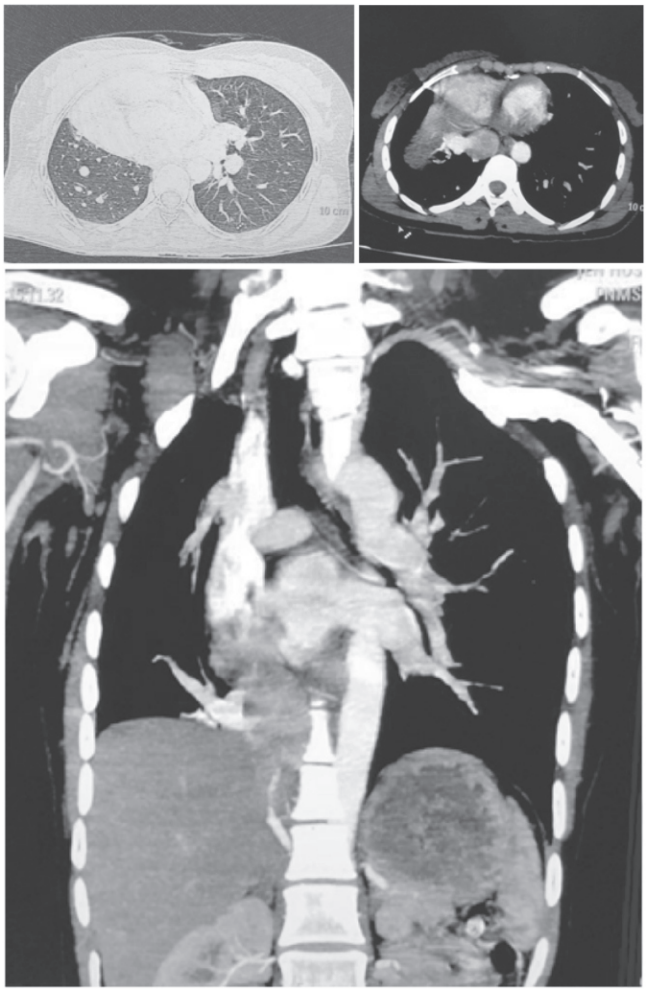
a) Hypoplastic right main pulmonary artery measured 16mm

Corresponding Author : Dr. Reena Mathur Professor & HOD, Department of Radio-diagnosis JLN Medical College Ajmer.

b) Systemic arterial supplyn of right lung base by a small branch from abdominal aorta.

c) Cardio-thoracic ratio was less than 50% , left main pulmonary artery and IVC were normal. Both lungs were normal in attenuation.

An echo-cardiograhic examination was normal.



DISCUSSION:

Hypogenetic lung syndrome, also known as congenital venolobar syndrome or scimitar syndrome (SS), is primarily a complex developmental lung abnormality with anomalous venous return. The most common features are lung hypoplasia, anomalous pulmonary venous return to IVC, pulmonary artery hypoplasia, bronchial anomalies and systemic arterial supply to hypoplastic lung. It almost always occurs on the right side, slightly more common in women (1.4:1); but there are rare cases reported on the left side and very rarely bilateral. The pulmonary artery hypoplasia determines the degree of systemic arterial supply to hypoplastic lung. In most patients, pulmonary artery hypoplasia is mild. However, it may be very significant or even absent.In our reported case, the right pulmonary artery was hypoplastic and there was systemic supply to right lung base by a small branche from abdominal aorta .

The other most constant component of this syndrome is an anomalous pulmonary vein or veins draining at least a part or the entire affected lung most commonly to inferior vena cava just above or below the diaphragm. Uncommonly, the anomalous vein may drain into hepatic, portal, azygos veins; coronary sinus; or right atrium. This results in extra-cardiac left-to-right shunt, which is usually mild in most patients and determines the clinical presentation and symptomatology.

Scimitar sign and scimitar syndrome are not synonymous. An isolated partial anomalous pulmonary venous return may be present with scimitar sign. Rarely, a scimitar vein courses down towards the medial costo-phrenic angle to drain into IVC and left atrium and is called scimitar variant. A wandering or meandering pulmonary vein resembling a scimitar on frontal chest X rays is reported to drain into left atrium without any connection to IVC. This is called by some authors as pseudo-scimitar syndrome and may be associated with lung hypoplasia and systemic arterial supply

Three forms of scimitar syndrome have been described in the literature: (a) infantile, (b) adolescent and (c) adult

a) Patients presenting in infancy have more severe symptoms, higher incidence of pulmonary hypertension and heart failure. Majority of them have cardiac anomalies, most commonly ASD and VSD; and the left-to-right shunt resulting from scimitar vein is considerably large. An obstructed scimitar vein and systemic supply to hypo-plastic lung further contribute to severity of pulmonary hypertension. The pulmonary over-circulation can prevent the normal postnatal regression of pulmonary artery muscularity, resulting in persistent pulmonary hypertension of newborn.

b) In the adolescent type, the clinical symptomatology mainly depends on the degree of lung hypoplasia and presents commonly with wheeze, shortness of breath, fatigue and failure to thrive, like in our case.

c) In adults, the scimitar syndrome is diagnosed incidentally on chest radiographs obtained for other reasons, as most are asymptomatic. They typically have no associated cardiac anomalies, and left-to-right shunt is insignificant.

CONCLUSION:

Scimitar syndrome is a rare complex congenital abnormality of the chest, frequently diagnosed on chest radiographs by presence of the scimitar vein.

Rarely, a meandering pulmonary vein, scimitar variants and pseudo-scimitar syndrome are easily mistaken for scimitar syndrome on chest X rays. Therefore, these individuals must be further evaluated with MSCT or MR angiography. Multislice CT with multiplanar and three-dimensional image reconstruction not only elegantly demonstrates the anatomy of various vascular anomalies but also gives a detailed account of common and rare known associations.

REFERENCES:

Cooper G. Case of malformation of the thoracic viscera consisting of imperfect development of the right lung and transposition of the heart. Londdon Med Gas. 1836;18:600–601.

Neill CA, Ferenca C, Sabiston DC. The familial occurrence of hypoplastic right lung with systemic arterial supply and venous return, "Scimitar Synddrome" Bull Johns Hopkins Hosp. 1960;107:1–21.

Dupuis C, Charaf LA, Breviere GM, Abou P, Remy-Jardin M, Helmius G. The "adult" form of the scimitar syndrome. Am J Cardiol. 1992;70:502–07

Drake EH, Lynch JP. Bronchiectasis associated with anomaly of the right pulmonary vein and right diaphragm: Report of a case. J Thorac Surg. 1950;19:433–437.

Kirklin JW, Ellis FH, Wood EH. Treatment of anomalous pulmonary venous drainage of the pulmonary venous connection in association with interatrial communications. Surg. 1956;39:389–98.

Honey M. Anomalous pulmonary venous drainage of right lung to inferior vena cava ("Scimitar syndrome"): clinical spectrum in older patients and role of surgery. Q J Med. 1977;46:463–83.

Shumacker HB, Judd D. Partial anomalous pulmonary venous return with reference to drainage into the inferior vena cava and to an intact atrial septum. J Cardiovasc Surg (Torino) 1964;5:271–278.

Zubiate P, Kay JH. Surgical correction of anomalous pulmonary venous connection. Ann Surg. 1962;156:234–50.

Puig-Massana M, Revuelta JM. A new technique in the correction of partial anomalous pulmonary venous drainage. J Thoracic Surg. 1972;64:108–13.

Tsuchida K, Anzai N, Hashimoto A, Fukushima Y, Yamada M. Intraatrial conduit repair in Scimitar syndrome. Chest. 1987;92:1120–2.

Casha AR, Sulaiman M, Cale AJR. Repair of adult Scimitar syndrome with an intra-atrial condduit. Interactive Cardiovasc Thorac Surg. 2003;2:128–30.