

*Case Report***Scimitar Syndrome: A Rare Reality**Ashfaq ul Hassan^{1*}, Ghulam Hassan², Shabin ul³, Zahida Rasool⁴¹Lecturer, Clinical Anatomy, Sheri Kashmir Institute of Medical Sciences, College Bemina, Srinagar, Kashmir²Ex Prof and Head, Department of Anatomy, Govt. Medical College, Srinagar, Kashmir, India³Resident, Sheri Kashmir Institute of Medical Sciences College Soura Srinagar Kashmir India⁴Medical Consultant, IUST Awantipora, Srinagar, Kashmir, India

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*Received: 27/06/2013**Revised: 16/09/2013**Accepted: 17/09/2013***ABSTRACT**

The scimitar syndrome is also termed as Hypogenetic lung syndrome or Pulmonary Venolobular syndrome or Halsalz lung. We represent the case of a 26 year old unmarried female who presented to the Medical OPD of SKIMS Soura Srinagar Kashmir India with repeated Chest infections. The female had made many OPD visits and despite the fact that antibiotics were administered on regular basis, still there was failure to respond to different antibiotics and a Chest Examination with investigation of Chest in the form of CXR and CT Scan Revealed the Syndrome. The syndrome can have many manifestations and Associations and a detailed investigation is warranted after diagnosis.

Key Words: Scimitar, Syndrome, Pulmonary, Radiograph, Anomalous.

INTRODUCTION

The scimitar syndrome is also termed as hypogenetic lung syndrome or pulmonary venolobular syndrome or Halsalz lung. It is named after the Turkish sword for its curvilinear opacity adjacent to the right heart border on a chest radiograph. Basically 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 caliber as it descends toward the cardiophrenic angle. The appearance closely resembles that of a curved Turkish sword or

scimitar. Physiologically, Scimitar syndrome represents a left to right shunt.

CASE REPORT

In our case the patient was a 26 year old unmarried female who presented to the Medical OPD of SKIMS Soura Srinagar Kashmir India with repeated Chest infections. The base line investigations as far as Hemogram, LFT, KFT, electrolytes were all within normal range. She was non hypertensive, euglycemic and Euthyroid with normal Menstrual History. The female had sought advice from doctors and made many OPD visits and despite the fact that antibiotics were administered on regular

basis, still there was failure to respond to different antibiotics and a Chest Examination with investigation of Chest in



Fig:1 CXR Scan Showing Scimitar Syndrome.

the form of CXR and CT Scan Revealed the features of the Syndrome.



Fig: 2. CT Scan Showing Scimitar Syndrome.

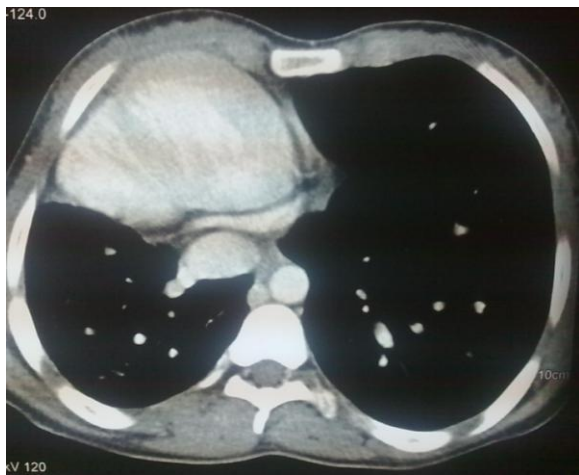


Fig: 3. CT Scan Showing Scimitar Syndrome.

The diagnosis is usually based on clinical presentation and transthoracic or transesophageal echocardiography, angiography, computed tomography and magnetic resonance angiography. The characteristic feature on chest radiographs, giving the condition its name, is a lesion in the shape of a scimitar (a type of curved Turkish sword). Prenatal diagnosis is feasible by fetal echocardiography. Management depends on the hemodynamic state. No therapy is required if the amount of blood flowing to the inferior caval vein is

small. In case of significant left-to-right shunt and pulmonary hypertension, surgical correction is warranted, and can include repair of the anomalous venous return, ligation of collateral arteries, and right pneumonectomy. When diagnosed in infancy, the syndrome is associated with significant mortality due to severe respiratory insufficiency, cardiac failure, and pulmonary infections.

DISCUSSION

Most patients are either minimally symptomatic or asymptomatic, and the treatment is usually conservative. The scimitar syndrome was first described by Neillet *al.* in 1960, who first while described this as a syndrome of partial anomalous pulmonary venous drainage of the right lung into the inferior vena cava, partial systemic arterial blood supply, and hypo-plasia of the affected lung, with bronchial abnormalities and abnormal lobation. It can present in late life or in Pediatric age group as well.

Mostly it is characterized by a combination of cardiopulmonary anomalies including partial anomalous pulmonary

venous return connection of the right lung to the inferior cava vein leading to the creation of a left-to-right shunt. Females are more frequently affected than males. In the majority of cases, the disease manifests in the first months of life. In the neonatal period, the disease presents with congestive cardiac failure, most commonly due to pulmonary hypertension and respiratory distress. The right lung is most frequently involved. Not a single spectrum of disease is seen but Variable and different degrees of hypoplasia and malformations of the pulmonary arteries are found in the affected lung, as well as arterial supply from the aorta, which can also arise above or below the diaphragm.

An association with Congenital heart disease (Coarctation of aorta, Tetralogy of Fallot, Patent arterial duct or Ventricular septal defect is seen). Other reported associated anomalies include bronchogenic cysts, horseshoe lung, accessory diaphragm and hernias. A look out is warranted in almost all the cases of the syndrome as the cause of death can be from the associated causes as well. It can be present in early life as well as in adults.

^[1] Pulmonary venous drainage into the right atrium, superior vena cava, the azygos system, the hepatic vein, or the left atrium has also been described. ^[2-8] Rarely, a scimitar syndrome of the left lung has been reported. ^[9,10] In most of the cases there is a combination of hypoplastic lung and partial anomalous pulmonary venous return. The anomalous vein drains into right atrium, IVC or even portal vein. It can be associated with Congenital heart diseases like ASD, VSD, TOF, Hemivertebrae^[11] genitourinary anomalies and a strong association can be resulting in an unfavourable prognosis.

CONCLUSION

The disease in itself is a rarity and few clinicians come across such cases and that too when they do not suspect the disease and is usually an incidental finding. But a detailed investigation can reveal the disease which otherwise can lead to fatal outcome. Fortunately in our case the female had no associated defects which can occur as a part of the syndrome.

REFERENCES

1. Neill CA, Ferenca C, Sabiston DC. The familial occurrence of hypoplastic right lung with systemic arterial supply and venous return, "scimitar syndrome".
2. 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 (new series XLVI)* 1977; 184: 463–483.
3. Canter CE, Martin TC, Spray TL, Weldon CS, Stauss AW. Scimitar syndrome in childhood. *Am J Cardiol* 1986; 58: 652–654.
4. Kuiper-Oosterwal CH, Moulart A. The scimitar syndrome in infancy and childhood. *Eur J Cardiol* 1973; 1:55–61.
5. Sanger PW, Taylor FH, Charlotte FR. The scimitar syndrome: diagnosis and treatment. *Arch Surg* 1963; 86:84–91
6. Gazzaniga AB, Matloff JM, Harken DE. Anomalous right pulmonary venous drainage into the inferior vena cava and left atrium. *J Thorac Cardiovasc Surg* 1969; 57:251–254.
7. Woody JN, Graham TP, Bradford WD, Sabiston DC, Canent RV, Durham NC. Hypoplastic right lung with systemic blood supply and anomalous pulmonary

- venous drainage: reversal of pulmonary hypertension with surgical management in infancy. *Am Heart J* 1972; 83: 82–88.
8. Huebsch P, Neuhold A, Mayr H, Gloger D. Anomalous pulmonary venous drainage shown by duplex sonography, computed tomography and plain radiography. *Thorax* 1989; 44: 63–65. *Surg* 1972; 64: 203–210.
 9. D'Cruz IA, Arcilla RA. Anomalous venous drainage of the left lung into the inferior vena cava: a case report. *Am Heart J* 1964; 67: 539–544
 10. Mardini MK, Sakati NA, Nyhan WL. Anomalous left pulmonary venous drainage to the inferior vena cava and through the pericardiophrenic vein to the innominate vein: left-sided Scimitar syndrome. *Am Heart J* 1981; 101:860–862.
 11. Berrocal T, Madrid C, Novo S et-al. Congenital anomalies of the tracheobronchial tree, lung, and mediastinum: embryology, radiology, and pathology. *Radiographics*. 24 (1): e17. doi:10.1148/rg.e17.

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