



INTERNATIONAL JOURNAL OF RESEARCH IN PHARMACEUTICAL SCIENCES

Published by JK Welfare & Pharmascope Foundation

Journal Home Page: <https://ijrps.com>

Scimitar Syndrome in an Adult: Case Report and Review

Nithyananthan P^{*1}, Kannan R¹, Mahendrakumar K¹, Rajendran K¹, Haji M² and Meenakshi K³¹Department of General Medicine, Saveetha Medical College, Saveetha Nagar, Thanadalam, Chennai - 602105 India²Department of Radiology, Saveetha Medical College, Saveetha Nagar, Thanadalam, Chennai - 602105 India³Department of Cardiology, Saveetha Medical College, Saveetha Nagar, Thanadalam, Chennai - 602105 India

Article History:

Received on: 09.03.2018

Revised on: 14.07.2018

Accepted on: 19.07.2018

Keywords:

Scimitar syndrome,
Anomalous pulmonary
Venous drainage,
Right lung hypoplasia,
Dextrocardia

ABSTRACT

Scimitar syndrome is an uncommon entity characterised by a combination of cardio-pulmonary anomalies that is consistently associated with partial or total anomalous venous drainage of one lung into the inferior vena cava and right lung hypoplasia. In infants and young children, it usually presents with cyanosis and failure to thrive, whereas, the presentation in adults is rare. Here, we report a 45 year old female who presented with lower respiratory infection and the chest radiograph showed 'Scimitar sign'. Contrast CT chest was done which confirmed the diagnosis of scimitar syndrome. Due to the absence of severe symptoms, she was managed conservatively without cardiac catheterization and surgery. This case is reported due to the rarity of the condition in adults.



* Corresponding Author

Name: Nithyananthan P
Email: drnithykmc@gmail.com

ISSN: 0975-7538

DOI: <https://doi.org/10.26452/ijrps.v9i4.1669>

Production and Hosted by

IJRPS | <https://ijrps.com>

© 2018 | All rights reserved.

INTRODUCTION

Scimitar syndrome derives its name from the curved sword of Turkish origin, the 'Scimitar'. It is characterised by a combination of cardio-pulmonary anomalies that is consistently associated with partial or total anomalous pulmonary venous drainage into the systemic circulation (Cooper G 1836). The anomalous right pulmonary vein draining into the inferior vena cava (IVC) appears as a curvilinear structure producing the characteristic 'Scimitar sign' on chest X-ray. Infants with scimitar syndrome may present with cyanosis (Gudjonsson U, Brown JW 2006; Dupuis C *et al.*, 1993) and require immediate

surgical intervention for survival. Scimitar syndrome in adults is rare and may be asymptomatic (Huang SF *et al.*, 2011) or may sometimes present with recurrent chest infections and dyspnoea. Surgical repair in adults is dependent on the severity of left to right shunt (Schramel FM *et al.*, 1995).

CASE REPORT

A 45 year old female presented with a cough and expectoration with right sided pleuritic type of chest pain for one week. She denied any history of fever, breathlessness or other illness. Her pulse was 80 beats/min regular, blood pressure 110/80 mm Hg, and respiratory rate 16 breaths/minute. On examination, tracheal shift to the right, right infra-clavicular hollowing, few right basal inspiratory crackles and bilateral wheeze were present. Rest of the systemic examination was normal. Her oxygen saturation, routine blood investigations and electrocardiogram were normal. Sputum for culture and acid fast bacilli were negative. Chest X-Ray showed a volume loss on the right, and shift of mediastinum and heart to the right. On careful examination, there was a curvilinear 'scimitar' like shadow extending from the right middle zone to the right cardiophrenic

recess – Scimitar sign (Figure 1).

A contrast CT chest scan was done which showed hypoplastic right lung (Figure 2A) and a curvilinear enlarged anomalous right inferior pulmonary vein draining into the suprahepatic IVC – the Scimitar vein, and another vein branching from the Scimitar vein and draining into the left atrium (Figure 2B). A double superior vena cava (SVC) was also observed, with the right SVC draining into the right atrium and left SVC draining directly into the left atrium (Figure 2C). Both right superior and left pulmonary veins normally drained into the left atrium. Based on these findings, a diagnosis of Scimitar Syndrome was made.

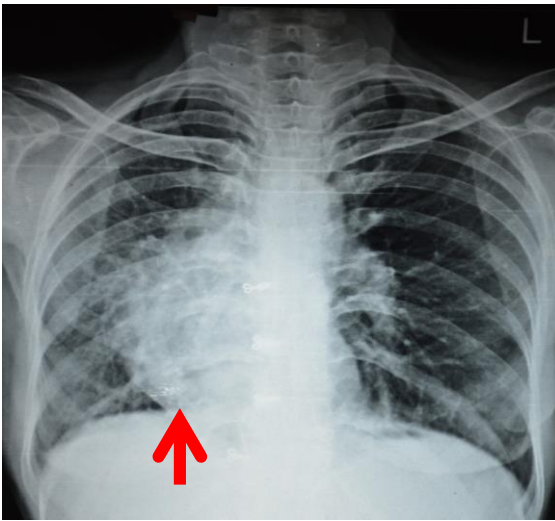


Figure 1: Chest X-Ray is showing small right hemithorax, mediastinal shift to the right and the Scimitar sign (arrow). Compare with the image of scimitar on the left.

Trans-thoracic echocardiography showed mild pulmonary hypertension (32 mmHg). Trans-oesophageal echo did not reveal any left to right shunt. The patient was treated with antibiotics for a lower respiratory infection after which her condition improved. She was discharged and has been on regular follow-up for 8 months without any symptoms.

DISCUSSION

Scimitar syndrome comprises about 3–5% of all cases of partial anomalous pulmonary venous connection (PAPVC). The other associated anomalies include dextrocardia, right lung hypoplasia, pulmonary artery hypoplasia and systemic arterial supply to the lung. The case presented here is a middle-aged female with no similar illnesses in the family. Studies show that women are more commonly affected and that most of the cases are sporadic, but a few familial cases have also been reported (Gudjonsson U, Brown JW 2006). The diagnosis of scimitar syndrome is usually made in infants; however, it may rarely be diagnosed after

1 year of age. Infantile form is most often associated with congenital heart diseases and usually presents with more serious problems like cyanosis, failure to thrive, respiratory insufficiency, pulmonary hypertension (PAH) and cardiac failure (Gudjonsson U, Brown JW 2006; Dupuis C *et al*, 1993). Mortality is high in these cases and immediate surgery is necessary for survival.

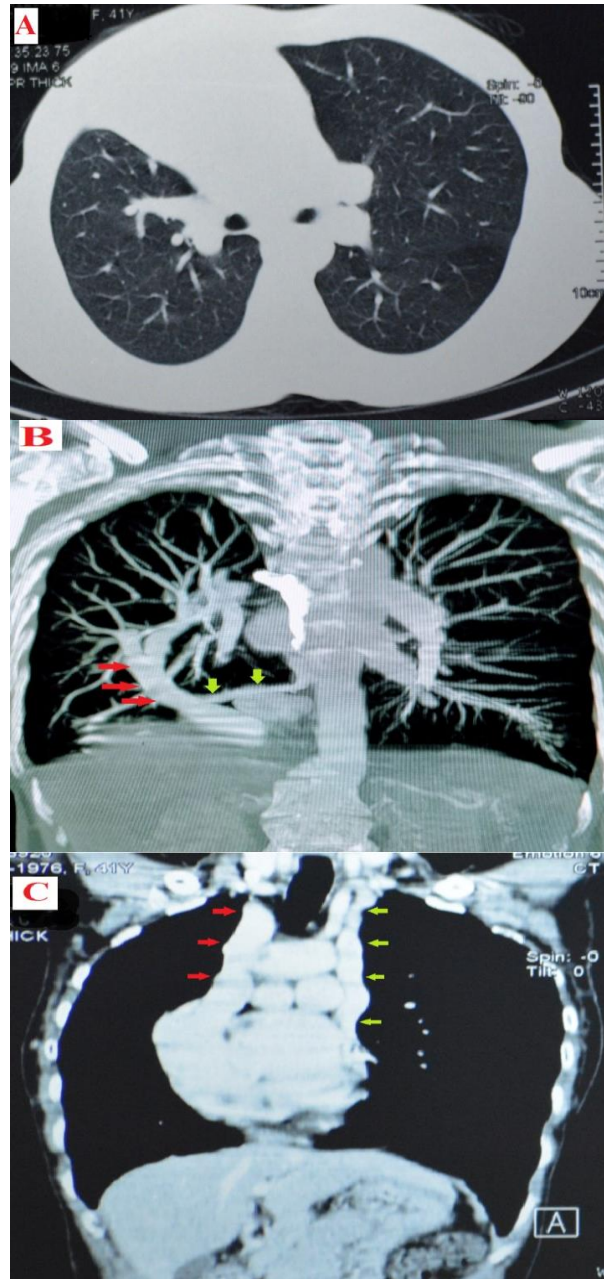


Figure 2: Contrast CT chest showing A) Hypoplastic right lung B) Scimitar vein (red arrows) and the vein branching from the Scimitar vein and draining into the left atrium (green arrows) C) Double SVC (Right SVC shown by red arrows and Left SVC shown by green arrows)

As in the present case, the adult form of scimitar syndrome is usually asymptomatic. Dupuis *et al*, 1992 showed that adult patients tend to develop

fatigue, exertional dyspnoea, recurrent chest infections and pulmonary hypertension only if the left-to-right shunt is more than 50%. Another striking feature in the study was that PAH was absent or only mild in almost all the patients. Zagol *et al.*, 2006 reported the oldest adult with scimitar syndrome in an 85 year old lady who was previously fit and had presented with infantile form complications such as PAH and right heart failure. Haemoptysis as a presenting feature in adult patients with scimitar syndrome is also exceptionally rare (Reddy R *et al.*, 2002; Gupta ML *et al.*, 2009; Matzinger FR *et al.*, 1994).

In a review of 9 cases, Honey (Honey M 1977) observed that patients who present beyond infancy usually do not have a pulmonary arterial pressure greater than 50 mm Hg. He also noted that PAH in this population might also be associated with cardiac abnormalities. In our patient, there was only mild PAH in the absence of other associated congenital cardiac defects. Canter CE *et al.*, 1986 showed that the occurrence of PAH without significant left-to-right shunt might be explained by the larger degree of anomalous pulmonary venous drainage, a greater extent of hypoplasia of right lung and pulmonary artery and increased systemic blood flow to the hypoplastic lung.

Chest X-ray shows the characteristic "Scimitar sign" in almost 70% cases and the diagnosis may be confirmed by non-invasive techniques like contrast CT chest, CT angiography or MRI angiography. However, the gold standard for diagnosis confirmation is cardiac catheterization, as it provides a quantitative assessment of the ratio of pulmonary to systemic flow (Qp/Qs) and also helps in planning therapeutic intervention (Huang SF *et al.*, 2011).

Schramel FM *et al.*, 1995 showed that the indications for surgical correction in adults include severe left to right shunting and lung sequestration with recurrent right-sided chest infections. However, in a study of 122 adult patients who were not overtly symptomatic and did not have severe PAH, Dupuis C *et al.*, 1992 showed that these asymptomatic adults might lead a normal life without surgical intervention.

In our patient, invasive cardiac catheterization and surgery were not necessary due to the absence of severe symptoms and severe left-to-right shunt. However, there has been a case report of a previously fit elderly lady presenting with infantile form complications such as PAH and right heart failure later in life (Zagol B *et al.*, 2006). Hence, regular monitoring of the patient and follow-up is warranted.

CONCLUSION

Scimitar syndrome in adults is rare and may be incidentally diagnosed. Asymptomatic adult patients usually do not require any surgical intervention. An awareness of this syndrome may avoid unnecessary invasive diagnostic procedures and surgical treatment in asymptomatic adult patients.

REFERENCES

- Canter CE, Martin TC, Spray TL, Weldon CS, Strauss AW. Scimitar syndrome in childhood. *Am J Cardiol* 1986;58: 652-4.
- Cooper G. Case malformation of the thoracic viscera: consisting of imperfect development of right lung and transposition of heart. *London Med Gaz* 1836; 18: 600-602
- Dupuis C, Charaf LA, Breviere GM, Abou P, Remy-Jardin M, Helmius G. The "adult" form of the scimitar syndrome. *Am J Cardiol* 1992;70:502-7.
- Dupuis C, Charaf LA, Breviere GM, Abou P. Infantile" form of the scimitar syndrome with pulmonary hypertension. *Am J Cardiol* 1993; 71: 1326-30.
- Gudjonsson U, Brown JW: Scimitar Syndrome. *SeminThoracCardiovascSurgPediatr Card Surg Annu* 2006; 2: 56-62.
- Gupta ML, Bagarhatta R, Sinha J. Scimitar syndrome: A rare disease with unusual presentation. *Lung India : Official Organ of Indian Chest Society.* 2009; 26(1): 26-9.
- 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: 463-83.
- Huang SF, Yu WC, Chern JH, Lee YC. Scimitar Syndrome in an Older Adult. *J Chin Med Assoc* 2011; 74(11): 516-9.
- Matzinger FR, Bhargava R, Peterson RA, Shamji FM, Perkins DG. Systemic arterial supply to the lung without sequestration: An unusual cause of haemoptysis. *Can Assoc Radiol J* 1994; 45: 44-7.
- Reddy R, Shah R, Thorpe JA, John G. Scimitar syndrome: A rare cause of haemoptysis. *Eur J Cardiothorac Surg* 2002;22:821
- Schramel FM, Westermann CJ, Knaepen PJ, van den Bosch JM. The scimitar syndrome: clinical spectrum and surgical treatment. *Eur Respir J* 1995; 8: 196-201.
- Zagol B, Book S, Krasuski RA. Late "adult form" scimitar syndrome is presenting with "infant form" complications. *J Invasive Cardiol* 2006; 18: E82-5.