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Scimitar syndrome in an elderly female without any cardiac anomaly: A case report

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Abstract
Scimitar syndrome or congenital pulmonary venolobar syndrome, is a rare congenital cardiovascular defect charactarized by anomalous venous return from the right lung (to the systemic venous drainage, rather than directly to the left atrium). This anomalous pulmonary venous return can be either partial (PAPVR) or total (TAPVR). 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. Considering its rareness we are reporting scimitar syndrome in middle aged female patient with long standing dyspnea and recurrent chest infections without any cardiac anomaly.

Keywords: PAPVR, TAPVR, Venolobar

Introduction

Scimitar syndrome or congenital pulmonary venolobar syndrome, is a rare congenital cardiovascular defect charactarized by anomalous venous return from the right lung (to the systemic venous drainage, rather than directly to the left atrium). This anomalous pulmonary venous return can be either partial (PAPVR) or total (TAPVR). 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. This radiographic density often has the shape of a scimitar, a type of Turkish curved sword. The syndrome was first described by Catherine Neill in 1960.

A classification has been proposed according to the age of presentation and the associated congenital cardiac malformations. The 'infantile' form, diagnosed in children younger than 1 year of age, is characterised by cardiorespiratory symptoms, pulmonary hypertension and severe adverse outcome. Conversely, the 'adult' form remains asymptomatic for many years and has a much more favourable outcome.

Considering its rareness we are reporting scimitar syndrome in middle aged female patient with long standing dyspnea and recurrent chest infections.

Case Report

53 year old female patient presented to us with complaints of long standing dyspnea with recurrent chest infections, cough and sputum. Her chest x ray showed reduced lung volume and abnormal right cadiac-mediastinal silhuotte. CT scan showed bronchiectatic changes in the posterior segment of right upper lobe and posterior aspect of apical and posterior basal segments of left lower lobe. On CT pulmonary angiography there was tortuous contrast filled dilated venous channels seen involving right lower lobe pulmonary parenchyma with stenotic ipsilateral single pulmonary vein and anomalous drainage of tortuous channel into the in fradiaphragmatic inferior vena cava which is suggestive of partial anomalous pulmonary veinous drainage (PAPVR). On 2D echocardiography there was trivial mitral regurgitation (MR), no other significant abnormality. Left ventricular ejection fraction was 60%. PFT showed moderately restrictive changes. ECG showed no abnormality. Other blood tests were within normal limit.

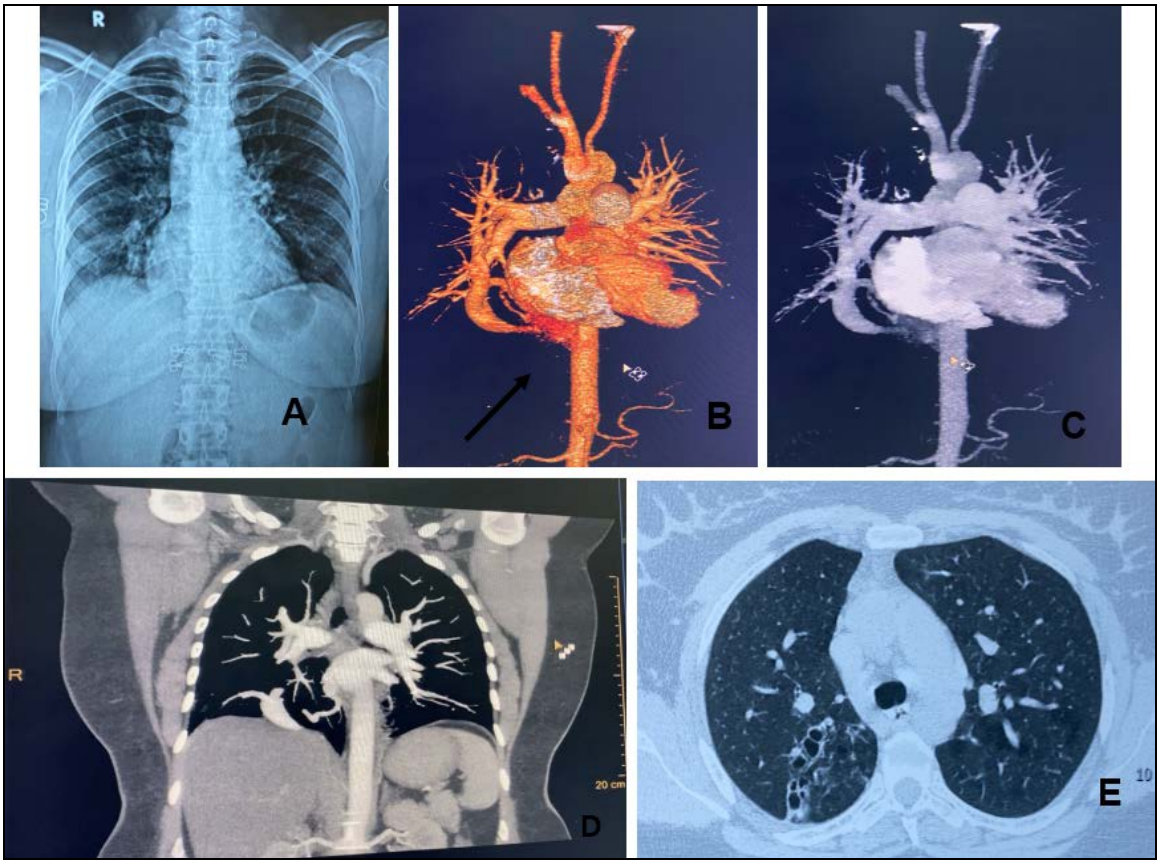

Fig 1: Figure A. Chest radiograph PA view showing reduced right lung volume in lower zone, bronchiectasis and increased bronchovascular markings. Figure B&C. CT pulmonary angiography 3D reconstructed picture of heart and major vessels showing stenotic ipsilateral single pulmonary vein and anomalous drainage of tortuous channel into the in fradiaphragmatic inferior vena cava. Figure D. CT pulmonary angiography showing ipsilateral single pulmonary vein and anomalous drainage of tortuous channel into the in fradiaphragmatic inferior vena cava. Figure E. CT thorax axial section showing bronchiectasis in right lower lobe.

Discussion

Scimitar syndrome, or congenital pulmonary venolobar syndrome, is a rare congenital heart defect characterized by anomalous venous return from the right lung to the systemic venous drainage, rather than directly to the left atrium [1]. Anomalous drainage of right pulmonary vein into inferior vena cava was first described in 1836 by George Cooper during the autopsy of a 10-month-old infant. The syndrome was first described by Catherine Neill in 1960 [2]. The incidence of scimitar syndrome is estimated at 1 to 3/100,000 live births [3, 4].

This anomalous pulmonary venous return can be either partial (PAPVR) or total (TAPVR). 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 [5]. This radiographic density often has the shape of a scimitar, a type of curved Turkish sword [5] known as scimitar sign.

A classification has been proposed according to the age of presentation and the associated congenital cardiac malformations [6]. The 'infantile' form, diagnosed in children younger than 1 year of age, is characterised by cardiorespiratory symptoms, pulmonary hypertension and severe adverse outcome. Pulmonary hypertension is a problem seen in infants and older children with scimitar syndrome [7] conversely, the 'adult' form remains asymptomatic for many years and has a much more favourable outcome [8].

Dupius *et al* studied 122 adult Scimitar syndrome from different cardiology centres to find about clinical, radiographic and hemodynamic findings in these patients. Their study showed that adult patients with scimitar syndrome have common association with hypoplasia of lung, dextrocardia, and abnormal bronchial segmentation but association with bronchiectasis is rare. The left to right shunt was < 50% in 100 of the 122 patients. The pulmonary arterial pressures were normal in 94 patients and slightly elevated in 28. A follow-up study of these patients showed that, without surgical correction, they lead a normal life

Traditionally, the initial diagnosis in most patients with the scimitar syndrome is established by chest X-radiography and is completed by echocardiography [9]. Cardiac catheterization should always be performed to confirm the diagnosis, identify the course of the anomalous venous drainage, measure the degree of left-to-right shunt, determinate the presence of scimitar vein stenosis and pulmonary hypertension and detect any associated cardiac abnormalities [10]. The increasing use of imaging techniques (computed tomography and magnetic resonance imaging) leads to increasing diagnosis of asymptomatic forms [11, 12]. Computed Tomography and Magnetic Resonance Imaging are currently available noninvasive imaging technique for complete mapping of anomaly.

In present case report chest x ray did not showed typical Scimitar Sign but there was reduced right lung volume in lower zone, presentation was typical indicating less severe form of disease. As per CT angiography and echocardiography reports there was no need of any surgical intervention. At present patient is on regular follow up with

good symptomatic relief. Awareness of this syndrome will avoid unnecessary invasive diagnostic workup in these types of patients with minimal symptoms due to Partial Anomalous Pulmonary Venous Drainage usually as they lead to normal life without any surgical intervention.

References

1. Sehgal A, Loughran-Fowlds A. Scimitar syndrome. Indian J Pediatr, 2005:72:249–251. https://doi.org/10.1007/BF02859268
2. Oransky Ivan. "Catherine Neill". The Lancet, 2006:367(9519):1312. DOI:https://doi.org/10.1016/S0140-6736(06)68565-6
3. Midyat L, Demir E, Aşkin M *et al*. Eponym. Scimitar syndrome. Eur J Pediatr,2010:169(10):1171-1177. DOI: 10.1007/s00431-010-1152-4
4. Juraszek AL, Cohn H, Van Praagh R, Van Praagh S. Isolated left-sided scimitar vein connecting all left pulmonary veins to the right inferior vena cava. Pediatr Cardiol,2005:26(6):846-847. doi: 10.1007/s00246-005-0920-9.
5. Scimitar Syndrome. Children's Hospital Boston, retrieved, 2008-02-28.
6. Dupuis C, Charaf LA, Brevière GM, Abou P. "Infantile" form of the scimitar syndrome with pulmonary hypertension. Am J Cardiol, 1993:71(15):1326-1330.doi:10.1016/0002-9149(93)90549-r.
7. Gupta ML, Bagarhatta R, Sinha J. Scimitar syndrome: a rare disease with unusual presentation. Lung India, 2009:26(01):26–29. doi: 10.4103/0970-2113.45202
8. Gudjonsson U, Brown JW. Scimitar syndrome. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu, 2006, 56-62. doi: 10.1053/j.pcsu.2006.02.011
9. Miguel Angel Ramirez-Marrero. Manuel de Mora-Martin, "Scimitar Syndrome in an Asymptomatic Adult: Fortuitous Diagnosis by Imaging Technique", Case Reports in Vascular Medicine, vol. 2012, Article ID 138541, 2012, 3. doi: 10.1053/j.pcsu.2006.02.011.
10. Brown JW, Ruzmetov M, Minnich DJ *et al*. Surgical management of scimitar syndrome: an alternative approach. J Thorac Cardiovasc Surg,2003:125(2):238-245. doi: 10.1067/mtc.2003.113
11. Gavazzi E, Ravanelli M, Farina D, Chiari ME, Maroldi R. “Scimitar syndrome comprehensive, noninvasive assessment with cardiovascular magnetic resonance imaging,” Circulation,2008:118(3):e63–e64. doi: 10.1161/CIRCULATIONAHA.107.748707
12. Inoue T, Ichihara M, Uchida T, Sakai Y, Hayashi T, Morooka S. “Three-dimensional computed tomography showing partial anomalous pulmonary venous connection complicated by the scimitar syndrome,” Circulation,2002:105(5)663. https://doi.org/10.1161/hc0502.101512