SCIMITAR SYNDROME WITH UNILOBAR RIGHT LUNG – A CASE REPORT

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ABSTRACT Scimitar syndrome is characterised by hypoplasia and anomalous pulmonary venous drainage of the right lung. Because of its rarity, we present a case of a 21 years old male who came with respiratory tract symptoms and pulmonary arterial hypertension and was diagnosed with Scimitar syndrome with unilobar right lung based on imaging methods and FOB findings.

KEYWORDS Scimitars syndrome, pulmonary arterial hypertension, lung hypoplasia

Introduction

Scimitar syndrome (SS) or pulmonary venolobar syndrome or hypogenetic lung syndrome is a rare congenital condition characterised by partial anomalous pulmonary venous connection of right lung to inferior vena cava, dextroposed heart and right lung hypoplasia. [1] The most common occurrence of this anomalous connection is below the diaphragm. It can occur as isolated or be associated with cardiac malformations. [2] Scimitar syndrome presentation can vary from asymptomatic to severe symptomatic (pulmonary hypertension, heart failure). [3] Infants and neonates present with severe symptoms due to the underlying congenital cardiac defects like atrial septal defects (ASD), whereas adults present with recurrent respiratory tract infections or haemoptysis. [4] The diagnosis of scimitar syndrome can be diagnosed by imaging techniques such as chest x-ray, 2D echo, contrast CT of the lung, CT angiography and bronchoscopy. We present a case of the unilobar right lung, who presented at OPD with respiratory tract symptoms and pulmonary arterial hypertension, subsequently diagnosed as Scimitar syndrome.

Case report

A 21 years old male patient was admitted to our hospital with cough, expectoration, dyspnoea and chest pain for five months. Shortness of breath (SOB) was of grade 1, and retrosternal chest pain was dull, aching in character without any radiation. He was neither a smoker nor an alcoholic with no other relevant history. Vitals were within the normal range. Respiratory system examination revealed decreased breath sound intensity on the right side and small right hemithorax. Cardiac examination showed prominent pulsations in the right parasternal area, but no murmur was observed. His blood counts, liver and renal function tests were within normal limits. Chest x-ray (fig 1) showed a small right hemithorax with a shift of heart towards the right side with a retrocardiac curved band like opacity extending towards the diaphragm (SCIMITAR’S SIGN). 2D echo revealed that the heart is dextroposed with mildly dilated RA/RV with pulmonary arterial hypertension (RVSP = 55 mmHg). Fibre optic bronchoscopy (FOB) revealed a lengthy right main bronchus with a missing upper lobe bronchus. The main bronchus continued to divide into a middle and lower lobe like pattern (fig.2). At the same time, branching was normal on the left side. CECT THORAX lung window was suggestive of a smaller right lung with no evidence of minor and significant fissures, a dextroposed heart and engorged pulmonary vasculature (fig 3). Axial section showed scimitar vessel draining into IVC just above the diaphragm and dilated RA and RV (fig 4). Left pulmonary draining into the left atrium was seen, but similar venous drainage from the right side was absent (fig 5). Sagittal section showing a retrocardiac vessel extending towards the diaphragm (fig 6). CT angiography with 3D reconstruction conformed with the same findings. Anomalous systemic arterial supply was not evident.

The diagnosis of scimitar syndrome with unilobar right lung
was made upon the findings from imaging tests and FOB mentioned above. The patient’s respiratory infection was treated with antibiotics and discharged in stable condition with no further complications.

**Discussion**

Scimitar syndrome was initially described by George Cooper in 1836 during the autopsy of a 10-month-old infant. The estimated incidence of scimitar syndrome is 1-3/100,000 live births. Scimitar Syndrome (SS) is also called hypogenetic lung syndrome, pulmonary venolobar syndrome, epibranchial right pulmonary artery syndrome, mirror image lung syndrome, Halasz’s syndrome and vena cava bronchovascular syndrome. Its main elements are 1. Hypoplasia/lobar aplasia / agenesis involving the right lung. Abnormal lobation, including bronchial isomerism with hyparterial bronchus and unilobar lung, may be present. Our patient presented with unilobar right lung associated with lobar agenesis (absence of parenchyma and corresponding bronchus). Errors of segmentation may occur, and sometimes it may present as horseshoe lung. 2. An anomalous right pulmonary vein draining into the systemic venous circulation either below or above the diaphragm, most commonly to the inferior vena cava, occasionally into the hepatic vein, portal vein, azygos vein, coronary sinus or the right atrium. The vein may drain the whole lung or part of it. 3. Partial or complete anomalous systemic arterial supply from the aorta or one of its branches to the right lung with the absent or hypoplastic right pulmonary artery. Other associated anomalies can be ASD, VSD, PDA, accessory diaphragm and absent inferior vena cava. Embryologically, SS represents a basic developmental derangement of the entire lung bud early in embryogenesis with no clear understanding for consistent occurrence on the right side. [5, 6]

Scimitar syndrome symptoms can be variable depending on age at presentation and the degree of the left to right shunt. In
infancy, the symptoms are severe due to congestive heart failure secondary to right heart volume overload. As in the present case, in older children and adults, it may present as recurrent respiratory infections. A plain radiograph may show a small right lung, small hilum with diminished vascularity and dextroposed heart. The characteristic scimitar sign consists of a broad curved, vertically oriented band like opacity along the right heart border, disappearing at the right hemidiaphragm, the anomalous vein. It is like the shape of a Turkish sword (Scimitar).

[7] CECT helps define the degree of lung hypoplasia and the anomalous vein with other associated anomalies, thus differentiating it from atelectasis, bronchopulmonary sequestration. True dextrocardia. [8, 9] MDCT with 3D reconstruction will detect the entire course of the scimitar vein and its eventual drainage site [10] and reveal anomalous arteries, which would help prevent torrential bleeding during surgery. Occasionally, a wandering right pulmonary vein may confuse normally entering into the left atrium. The diagnosis can be made with MR imaging also. Echocardiography to rule out associated cardiac anomalies like ASD should be done. Treatment of SS encompasses treating respiratory tract infections, haemodynamic correction and bleeding control in case of haemoptysis. Surgical repair consists of direct anastomosis of scimitar vein to the left atrium or creation of an ASD, transposition of anomalous vein followed by occlusion of ASD with a Dacron patch. Surgery is recommended when pulmonary systemic blood flow exceeds 1.5-2 to mitigate the risk of progression to PAH and right ventricular failure. [11, 12]

**Conclusion**

SS is a rare congenital bronchopulmonary vascular malformation, which can be diagnosed at the earliest by radiological imag-
ing methods like CT Angiography with 3D reconstruction or MRI. Scimitar sign is evident on the chest radiograph of many patients. However, confirmation of scimitar syndrome diagnosis often necessitates additional diagnostic procedures, which also help evaluate associated abnormalities. Outcome prediction depends on the age of presentation and the presence of associated anomalies. In general, presentation in infancy and the presence of heart failure are poor prognostic factors.

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**Conflict of interest**

There are no conflicts of interest to declare by any of the authors of this study.

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