

Adult Type Scimitar Syndrome With Recurrent Pneumonia And Absence Of Right Upper Lobe Bronchus

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Citation

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Abstract

Scimitar syndrome consists of anomalous right lung vessel, draining often into IVC. We present a case of 34 years aged lady with recurrent pneumonia and volume loss of right lung, where CT demonstrated abnormal vessel, absence of right upper lobe bronchus, bronchiectasis in right middle lobe of the lung, small sized right pulmonary artery and minimal dextro-rotation of the heart.

INTRODUCTION

The scimitar syndrome (pulmonary venolobar syndrome, hypogenetic right lung syndrome) has anomalous vein carrying all or part of right lung venous return to IVC, portal or hepatic vein or atria; hypoplasia of right lung; dextrorotation of heart and hypoplastic right pulmonary artery (_{1,2,3}). We present a case which also showed absence of right upper lobe bronchus.

CASE REPORT

A 34 years aged non-diabetic, non-hypertensive lady presented with the chief complaints of cough with expectoration and fever, since one week. History of recurrent attacks of lower respiratory tract infection was elicited. On examination, the patient was febrile and auscultation revealed crepitations in the right middle and lower zones. No other positive finding was seen on the physical examination.

Chest radiograph revealed ill-defined haziness in the right middle and lower zones. Reduced volume of the right hemi thorax, with minimal dextro-rotation of the heart was also seen (fig. 1).

Figure 1

Figure 1: Chest scout view shows haziness in the right middle and lower zones, with volume loss on the right.

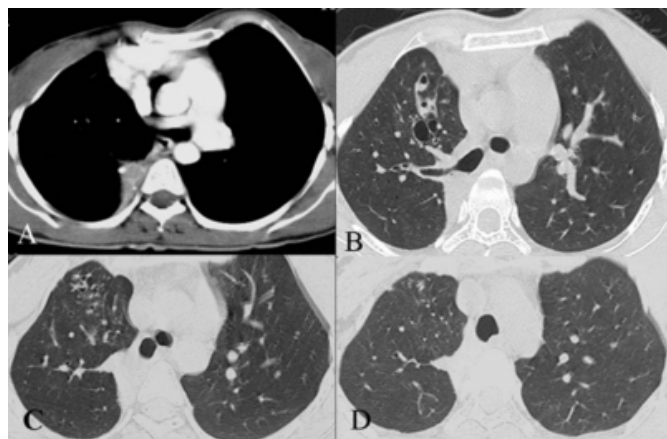


A contrast enhanced CT scan of the chest was then performed. Volume loss in the right hemi thorax was seen, with reduced antero-posterior dimensions of the right thoracic cage. Dextro-rotation of the heart was also observed. A small right main pulmonary artery, which measured 1.0 cm in diameter was seen (left measured 1.5 cm). The right upper lobe bronchus was not visualized. The bronchus intermedius was instead rising from the trachea in place of the right main bronchus. The right middle lobe was collapsed, cystic bronchiectatic changes containing fluid levels were seen in right middle lobe with adjacent reticulations and fibrosis. A small consolidatory patch was

also noted in right lower lobe (fig. 2).

Figure 2

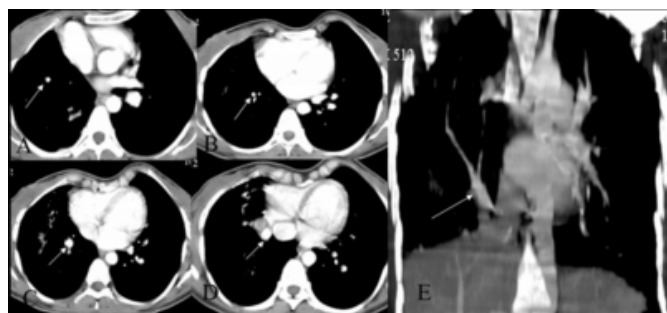
Figure 2: Axial contrast enhanced CT sections at the level of main pulmonary artery bifurcation reveals a small right pulmonary artery (A). Volume loss on the right side with collapse of middle lobe and bronchiectatic changes are also noted on lung window settings (B). Right lower lobe infiltrates are also noted in right paracardiac region (white arrow). There is absence of right upper lobe bronchus (B,C,D) with presence of bronchus intermedius (black arrow).



In addition, an anomalous vessel was seen in the right lung inferiorly, increasing in size, as it approached the diaphragm, opening into the supradiaphragmatic inferior vena cava. This aberrant vessel was laterally convex, and was well seen on the coronal reconstructed images (fig. 3).

Figure 3

Figure 3: Series of cranio-caudad axial CT scan images (A-D) show an anomalous vein in the right lung, leading into the terminal inferior vena cava (arrow). This vessel is well seen on the coronal reconstructed Maximum Intensity Projection (MIP) image (E).



On the basis of these imaging findings, the diagnosis of an adult type of Scimitar syndrome with infected bronchiectasis was made. Patient was managed conservatively with antibiotics and physiotherapy. The CT diagnosis is confirmatory except for any aberrant arterial supply from the

aorta, which was not found on CT angiography. However, conventional DSA was not done, for 100% ruling out of aortic supply.

DISCUSSION

The scimitar syndrome also called as pulmonary venolobar syndrome and hypogenetic right lung syndrome, is an uncommon congenital cardio-vascular anomaly, involving the right lung (^{1,2}). In its complete form, the syndrome consists of partial or total anomalous pulmonary venous drainage of the right lung into the inferior vena cava, hypoplasia of the right lung, dextrorotation of the heart and hypoplasia or other malformation of the right pulmonary artery. Anomalous systemic arterial supply to the lower lobe of the right lung from the sub-diaphragmatic aorta or its main branches may be associated. Scimitar syndrome is usually right-sided; however, rare cases have been reported that involved the left side (³). Other abnormalities which may be seen with this condition are abnormal bronchial anatomy, diaphragmatic abnormalities, hemi vertebrae, and anomalies of the genitourinary tract. Cardiac abnormalities like interatrial septal defect and patent ductus arteriosus are also associated (⁴). Our case showed abnormal bronchial branching in the right lung with absence of right upper lobe bronchus. 2D echo examination of heart was however normal.

This condition is named after the anomalous vein carrying all or part of the right lungs venous return to the inferior vena cava. The collector usually drains to IVC; but portal vein, hepatic vein, right or event the left atrium may also receive the vein. This is in contrast to the anomalous vein in total anomalous pulmonary venous drainage, which virtually never opens into the inferior vena cava. Three forms of scimitar syndrome have been described (¹), on the basis of age at presentation. In the infantile form, there is a large shunt between the abnormal artery that supplies the lower lobe of the right lung and the sub-diaphragmatic aorta. Patients in whom the diagnosis was made during the first year of life had more severe symptoms and had a higher incidence of heart failure and pulmonary hypertension than did the patients in whom the diagnosis was made after age 1 year (⁵). In the adult form, there is a small shunt between the right pulmonary veins and inferior vena cava. A left-to-right shunt is established because the anomalous pulmonary vein drains blood from the right lung into the IVC, resulting in an increased risk of developing right ventricular failure due to long-standing right ventricular volume overload. The clinical symptoms usually manifest in the 2nd to 3rd decade of life,

with shortness of breath or asthenia most commonly and recurrent pulmonary infection being less common. It may also be discovered incidentally when patients undergo routine chest radiography by detecting the anomalous vein as a “scimitar”-like shadow on a chest radiograph. A rare third form is characterized by additional cardiac and extra cardiac malformations.

The chest radiograph is characteristic, and usually diagnostic. It shows the abnormal vein draining into the inferior vena cava as a curved vascular shadow with a lateral convexity in the right lower zone, called the scimitar sign (6). However, in some cases, when the scimitar vein is masked by the overlying cardiac shadow or superadded lung pathology, Colour Doppler, computed tomography, and magnetic resonance imaging can be helpful by showing the abnormal vein and its insertion into the inferior vena cava. They also aid in detecting other abnormalities usually associated with this condition. Radiographs also show reduced volume of the right lung with dextro-position of heart.

Scimitar syndrome seldom necessitates surgical intervention in the adult type (2). Surgical correction is recommended for symptomatic patients or asymptomatic patients with a pulmonary-to-systemic blood flow ratio exceeding 1.5–2 because of their higher likelihood of progression to pulmonary hypertension and right ventricular failure. However, repeated lung infections can sometimes also

require lobectomy or pneumonectomy. Surgical repair seldom results in normal blood flow to the right lung but abolishes left-to-right shunt. Post-operative pulmonary venous obstruction is prevalent, especially in the infants (5).

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