Multiple Detector Computed Tomography Evaluation of Congenital Pulmonary Venolobar Syndrome: Scimitar Syndrome with Pulmonary Arterio-Venous Fistula

Anand Hatgaonkar¹, Kajal Salpekar²
¹Associate Professor, Department of Radiodiagnosis, B. J. Government Medical College and Sasoon Hospital, Pune, Maharshtra, India, ⁲Senior Resident, Department of Pathology, NKP Salve Institute of Medical Sciences and Lata Mageshkar Hospital, Nagpur, Maharshtra, India

The term dysmorphic lung has been introduced to describe any complex congenital malformation that involves both abnormal pulmonary vascular morphology and disordered growth of the entire lung. The major group within this definition of the dysmorphic lung is the scimitar syndrome. Scimitar syndrome is a rare anomaly consisting of partial anomalous pulmonary venous return into the supra or infradiaphragmatic portion of the inferior vena cava, right lung hypoplasia, dextroposition of the heart, and anomalous systemic arterial supply from the aorta or one of its branches to the right lung. It can be associated with congenital cardiovascular defects namely dextrocardia, coarctation of the aorta, Tetralogy of Fallot, patent ductus arteriosus, atrial or ventricular septal defect, and right pulmonary artery hypoplasia. Associated pulmonary anomalies are lung hypoplasia, sequestration, and tracheobronchial anomalies. Thus, it can manifest with heart failure and recurrent pneumonia. Other reported associated anomalies include bronchogenic cysts, horseshoe lung, accessory diaphragm, and hernias. We report an 8-year-old boy with complaint of chest pain, who was diagnosed as having scimitar syndrome with rare association of pulmonary arteriovenous fistula within the involved lung and role of newer faster multiple detector computed tomography scanner in its evaluation.

Keywords: Anomalous pulmonary venous return, Pulmonary arteriovenous fistulas, Scimitar syndrome

INTRODUCTION

Scimitar syndrome or congenital pulmonary venolobar syndrome is a rare anomaly consisting of partial pulmonary venous drainage into the supra or infradiaphragmatic portion of the inferior vena cava (IVC), dextroposition of the heart, right lung hypoplasia, and anomalous systemic arterial supply from the aorta or one of its branches to the right lung. Because the radiographic shadow of the anomalous vein resembles a curved or Turkish sword, this defect has been named the “scimitar deformity.”

We report an 8-year-old boy with complaint of chest pain, who was diagnosed as having scimitar syndrome with rare association of pulmonary arteriovenous (A-V) fistula within the involved lung and role of newer faster multiple detector computed tomography (MDCT) scanner in its evaluation.

CASE REPORT

The 8-year-old male child brought by parents with complaints of chest pain since 2 months. CT pulmonary angiography was advised to rule out pulmonary hypertension.

Radiograph chest posterioranterior (PA) revealed cardiomegaly with dextrocardia and right lung oligemia with left hilar prominence. There was vessel running parallel to the right heart border and draining in infradiaphragmatic region, suggestive of partial anomalous pulmonary venous drainage, scimitar syndrome (Figure 1). However, as it appears in our patient, this radiologic sign can be hidden behind the dextrocardia on chest PA radiograph (Figure 2).

CT pulmonary angiography was done using Philips Brilliance i 256 slice scanner. It revealed dextroposition of the heart with predominantly right-sided cardiomegaly and large atrial septal defect (ASD) (Figures 3 and 4). The right main pulmonary artery was stenosed with resultant right pulmonary oligemia. Right posterior and medial basal...
segments had systemic supply from infradiaphragmatic portion of abdominal aorta as branch of celiac trunk (Figure 5).

Right-sided pulmonary veins are seen draining the entire right lung into the IVC at the level of opening of the right hepatic vein suggests partial anomalous right pulmonary venous return (Figures 5-7). Right lung pulmonary A-V fistula is seen draining into anomalous right pulmonary vein (Figure 8).

The right lung was hypoplastic with hypoplastic right upper lobe and middle lobe bronchus. Left oblique fissure visualized normal, however, right-sided fissures not visualized (Figures 7 and 9).

**DISCUSSION**

The term dysmorphic lung implies any complex congenital malformation that involves both abnormal vascular morphology and disordered growth of the entire lung. The major group within this definition of the dysmorphic lung is the scimitar syndrome.

Scimitar syndrome also called congenital pulmonary venolobar syndrome is a rare anomaly consisting of partial anomalous pulmonary venous return into the hepatic portion of the IVC, anomalous systemic arterial supply from the aorta or one of its branches to the right lung, right lung hypoplasia and dextroposition of the heart. As the radiographic shadow of the anomalous vein resembles a curved or Turkish sable, this defect has been named the “scimitar deformity.”

Scimitar syndrome is a very rare, estimated to occur in 2 out of 100,000 births with a 2:1 female preponderance. It’s association with congenital cardiopulmonary anomalies has been reported in 3-6% of patients with the partial anomalous venous connection. In these patients, some of the pulmonary veins drain into the upper part...
of the IVC, either above or below the diaphragm, mostly drains the lower and sometimes the middle lobe of the right lung (79%) or the whole right lung (21%). The right lung is frequently hypoplastic and receives blood supply from the systemic arteries, mainly the thoracic or abdominal aorta, and this supply is usually to the lower lobes.4

The triad of respiratory distress, right lung hypoplasia, and dextroposition of the heart should alert the clinician to the possibility of this syndrome.2 Many patients are asymptomatic, and the condition is often discovered accidentally. Symptomatic patients are often infants with severe symptoms and worse prognosis and marked left-to-right shunt along with associated severe congenital heart disease. While older children present
with recurrent respiratory infections, heart murmur, or an abnormal chest radiograph.\(^5,6\)

This anomaly was first described by Cooper in London in 1836 during an autopsy of an infant. In which he made note of dextroposition of the heart and hypoplasia of the right lung in this specimen.\(^7\) The first diagnosis in live and asymptomatic patient was made in 1949 by Dotter et al. on cardiac catheterization.\(^8\)

The abnormal pulmonary vein drains the entire lung or only the lower lobe, the middle lobe, or both lobes. The upper portion of the lung drains normally into the left atrium, as does sometimes the middle lobe. As in our case, there is often a reduced size of the right pulmonary artery and some degree of bronchial anomalies, with the absence of fissures, abnormal bronchial connection, which may sometime give an aspect of bronchial isomerism and the right-side anatomy resembling the left one.

The existence of an aberrant artery arising from the aorta or one of its side branches may mimic sequestration. However, because bronchopulmonary sequestration is as a definition an area of nonfunctional lung parenchyma that has no connection with the bronchial system, this designation is not correct.\(^5\) As stressed by Panicek et al., pulmonary anomalies exist as a continuum, often frustrating our attempts to classify them.\(^1\)

Pulmonary A-V malformations are frequently associated with Rendu-Osler-Weber syndrome. They usually act as direct right-to-left shunts, resulting in fatigue, cyanosis and polycythemia, with a risk of neurologic manifestations.\(^5\) This association of pulmonary A-V fistulas with a scimitar syndrome is difficult to understand but suggests that all of these abnormalities occurred at a similar time early in pulmonary embryologic development.

A careful approach is warranted while treating these cases as almost all the cases of the syndrome the cause of death can be from the associated causes.\(^3,10\) Rarely, a scimitar syndrome of the left lung has been reported.\(^10,11\) Pulmonary venous drainage into the right atrium, superior vena cava, the azygos system, the hepatic vein, or the left atrium has also been described.\(^2,11\)

Scimitar syndrome can be associated with congenital cardiovascular defects namely dextrocardia, coarctation of the aorta, Tetralogy of Fallot, patent ductus arteriosus, ASD, or ventricular septal defect and right pulmonary artery hypoplasia. Associated pulmonary anomalies are lung hypoplasia, sequestration and tracheobronchial anomalies. Thus, it can manifest with heart failure and recurrent pneumonia. Other reported associated anomalies include bronchogenic cysts, horseshoe lung, accessory diaphragm and hernias.\(^12\)

In these patients, results of reimplantation of the anomalous vein have been disappointing;\(^13\) resection of the abnormal lung when feasible, being the treatment of choice.\(^13\) Pre-operative MDCT evaluation is helpful by depicting the feeding vessels and other associated findings.\(^6\)

**CONCLUSION**

Unusual association of congenital heart disease with bronchopulmonary abnormalities is not new. New multislice faster and efficient CT scanner had changed the face of the diagnostic evaluation of congenital heart disease. Evaluating abnormalities of heart and bronchopulmonary tree is not possible using single modality as two-dimensional ecocardiography (2D echo) examination. Despite of the fact that 2D echo is time tested modality and gold standard in diagnosing congenital heart disease, 2D echo cannot provide information about peripheral pulmonary circulation and bronchial anatomy. For studying pulmonary vasculature and bronchial anatomy other modalities like Digital Subtraction Angiography (DSA) or ventilation/perfusion lung scan (V/Q lung scan) are needed. Considering the Radiation exposure, newer CT machines are efficient enough to reduce the radiation significantly much less than a older scanner and negligible as considered to the DSA. Hence to conclude, MDCT scanner is a better modality to evaluate heart disease and bronchopulmonary abnormalities together as in our case MDCT turn out to be single modality adequate for diagnosing the abnormality and provided road map to treating surgeon for proper planning of treatment.

**REFERENCES**


