

Scimitar syndrome

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Abstract

A seven month old female child admitted to intensive care unit with increasing respiratory distress. Clinical examination revealed absence of cyanosis, wide and fixed normal intensity second heart sound. Chest radiograph revealed crescent like radio opacity in right lower lung and subsequent contrast enhanced computed tomography (CT) chest demonstrated abnormal vessel in right lung draining into the inferior vena cava.

Key words: Scimitar syndrome, partial anomalous pulmonary venous return, computed tomography

Scimitar syndrome, also known as congenital pulmonary venolobar syndrome, is a rare congenital anomaly most commonly consists of partial anomalous pulmonary venous connection of right lung to the inferior vena cava, right lung hypoplasia, dextroposition of the heart¹. We report a case of scimitar syndrome with absence of right upper lobe bronchus in a seven month old child who presented with respiratory distress, where computed tomography (CT) chest showed abnormal vessel draining into the inferior vena cava, absence of right upper lobe bronchus and minimal dextroposition of the heart.

Case Report

A seven month old female child admitted to intensive care unit with increasing respiratory distress. She was



Fig 1: Chest radiograph shows crescent like radio opacity in the right lower lung field and hyperinflation of left lung.

delivered by normal vaginal delivery at home at full term. Her immediate postnatal period was uneventful. Her past medical history includes three times admissions in the hospital for respiratory tract infections. Clinical examination revealed absence of cyanosis, wide and fixed normal intensity second heart sound. Chest radiograph revealed volume loss of right lung, crescent like radio opacity in the right lower lung field and hyperinflation of left lung (Fig 1). Echocardiography shows ostium secundum atrial septal defect of 5 mm size. Contrast enhanced computed tomography (CT) of chest revealed abnormal vessel draining into the inferior vena cava, absence of right upper lobe bronchus and minimal dextroposition of the heart (Fig 2).



Fig 2: Three dimensional reformatted image demonstrates the anomalous vein drains into the IVC below the level of the right hemidiaphragm.

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Discussion

Partial anomalous pulmonary venous return (PAPVR) is a relatively uncommon congenital anomaly accounting for 0.5- 1 % of congenital heart disease². Scimitar syndrome is associated with a hypoplastic lung and partial anomalous venous return with systemic venous drainage³. Additional anomalies that occur with this syndrome include: hypoplastic or absent pulmonary artery, anomalous systemic arterial supply to the right lung from aorta or one of its branches, pulmonary sequestration, absence of the inferior vena cava and accessory diaphragm⁴. The associated congenital cardiovascular abnormalities include atrial septal defect, ventricular septal defect, coarctation of the aorta, abnormalities of the aortic arch and abnormal relationship of the pulmonary arteries and bronchi⁵. The rare combination of an association of PAPVR, hypoplasia of right lung and dextroposition of the heart is designed scimitar syndrome.

The characteristic abnormality of scimitar syndrome is partial or complete anomalous pulmonary venous drainage of the right lung to the inferior vena cava. The name comes from this anomalous pulmonary vein, which may be visible on chest radiograph as a curvilinear shadow just above to the right diaphragm said to resemble a "Scimitar" or Turkish sword⁶. The diagnosis can frequently be made on a chest radiograph and echocardiography. There are various presenting symptoms of this disease. The triad of respiratory distress, right lung hypoplasia and dextroposition of the heart should alert the clinician to the possibility of this syndrome⁷.

Contrast enhanced cardio computed tomography (CT) or multi detector CT (MDCT) is useful to confirm the diagnosis, identifying the anomalous pulmonary vein and demonstrate other associated abnormalities. Three-dimensional (3D) reconstructions give the surgeon a clear picture of the malformation for the appropriate management. In our patient, 3D reconstruction imaging provided an excellent demonstration of the abnormal vessel. Standard diagnostic measures for suspected PAPVR consists of transthoracic and transesophageal echocardiography as well as invasive heart catheterization including quantitative determination of the pulmonary to systemic blood flow by oxymetry².

Most frequently, patients are asymptomatic in the absence of associated abnormalities². The extent of

symptoms are dependent on the degree of shunting, the number of anomalous veins, any associated valvular anomalies and the presence of concomitant cardiac or pulmonary disease. Surgical correction is recommended for symptomatic patients with lung sequestration and recurrent right sided chest infections 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 failure¹. Our patient was treated conservatively and advised for the regular follow up. Surgical treatment is best managed in two steps: 1) the anomalous systemic arteries are ligated and the scimitar vein is re-implanted into the left atrium; and 2) resection of sequestered or chronically infected lung parenchyma is performed⁸.

References

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