Journal of Advanced Medical and Dental Sciences Research

@Society of Scientific Research and Studies *NLM ID: 101716117*

Journal home page: www.jamdsr.com doi: 10.21276/jamdsr Indian Citation Index (ICI) Index Copernicus value = 100

(e) ISSN Online: 2321-9599; (p) ISSN Print: 2348-6805

Case Report

Classical Case Of Congenital Venolobar Syndrome

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ABSTRACT:
Congenital venolobar syndrome is a cardio-pulmonary anomaly and is a form of anomalous pulmonary venous drainage in which there is pulmonary venous system connected to the systemic vein/s, typically to the inferior vena cava, superior vena cava, portal vein, or directly to the right atrium. Here we present a case of 14-day-old newborn presented with refractory respiratory distress since birth, poor feeding, and vomiting & and was admitted with signs of cardiac failure. Associated with cleft palate and Micrognathia. Both Chest X-ray and Multi-detector Computed tomography following contrast were done. Chest radiographic showed a small, hypoplastic right lung with ipsilateral cardio-mediastinal shift. Contrast CT & multiplanar reconstruction images showed the course of this anomalous right lower lobe pulmonary venous drainage to supradiaphragmatic IVC The case was diagnosed as congenital venolobar syndrome. Key words- Congenital venolobar syndrome, Refractory respiratory distress, Anomalous right lower lobe pulmonary venous drainage

Received Date: 22 July, 2024 Accepted Date: 24 August, 2024

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This article may be cited as: Bhagat S, Ohri P, Swami J, Bansal N, Kumar H. Classical Case Of Congenital Venolobar Syndrome. J Adv Med Dent Scie Res 2024; 12(10):124-127.

INTRODUCTION

Congenital venolobar syndrome is a cardio-pulmonary anomaly and is a form of anomalous pulmonary venous drainage in which there is pulmonary venous system connected to the systemic vein/s, typically to the inferior vena cava, superior vena cava, portal vein, or directly to the right atrium. The vein is said to resemble a curved Turkish sword called a "Scimitar". The appearance on chest x-ray is referred to as the “Scimitar Sign”. The other components of the syndrome include hypoplasia of the lower lobe of the right lung, hypoplasia of the right pulmonary artery or its lower branch, abnormal lobar architecture, abnormalities of tracheobronchial architecture and sequestration of the right lower lobe. Here we present a case of 14-day-old newborn presented with refractory respiratory distress since birth, poor feeding, and vomiting & and was admitted with signs of cardiac failure. Associated with cleft palate and Micrognathia.

CLINICAL HISTORY

A 14-day-old newborn presented with refractory respiratory distress since birth, poor feeding, and vomiting & and was admitted with signs of cardiac failure. Associated with cleft palate and Micrognathia.

IMAGING FINDINGS

Both Chest X-ray and Multi-detector Computed tomography following contrast were done. Chest radiographic showed a small, hypoplastic right lung with ipsilateral cardio-mediastinal shift. Faintly visualized curved tubular structure behind the right cardiac border in the shape of a turkish sword was noted. The right cardiac border is blurred. Later computed tomography was done following contrast administration demonstrated partial volume loss of the right middle and lower lobes with ipsilateral cardio-mediastinal shift suggesting hypoplastic right middle and lower lobe. Multiple areas of consolidation were also seen in the right upper, mid, and lower lobes. Curved tubular opacity showed contrast enhancement suggesting vascular structure. Multiplanar reconstruction images showed the course of this anomalous right lower lobe pulmonary venous drainage to supradiaphragmatic IVC just before its confluence with the right atrium. Right pulmonary artery appears smaller as compared to the left pulmonary artery.

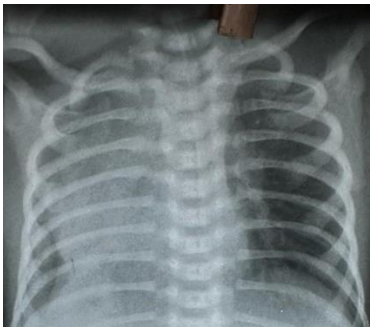

Figure 1: Chest radiographic showing a) small, hypoplastic right lung with ipsilateral cardio-mediastinal shift. b) Faintly visualized curved tubular structure behind the right cardiac border in the shape of a turkish sword was noted. c) The right cardiac border is blurred

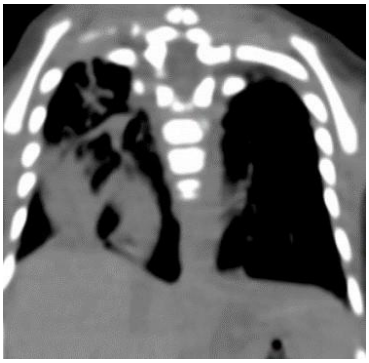

Figure 2: NCCT coronal image showing curve linear soft tissue density with convexity toward left side and reaching upto medial aspect of right hemidiaphragm. Reduced right lung volume with ipsilateral cardio-mediastina shift.

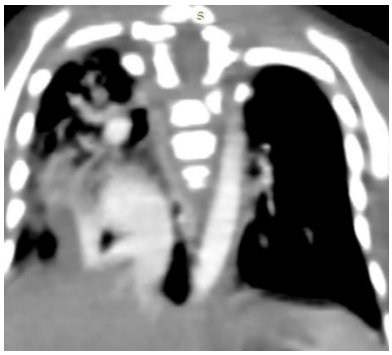

Figure 3: CECT chest axial and coronal images showing contrast opacification of curvilinear soft tissue density. Anomalous right lower lobe pulmonary venous drainage to supradiaphragmatic IVC just before its confluence with the right atrium is noted.

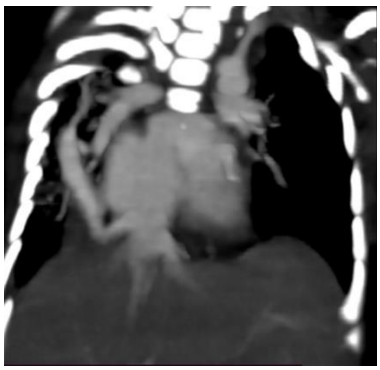

Figure 4: MPR reconstruction showing Anomalous right lower lobe pulmonary venous drainage to supradiaphragmatic IVC just before its confluence with the right atrium is noted.

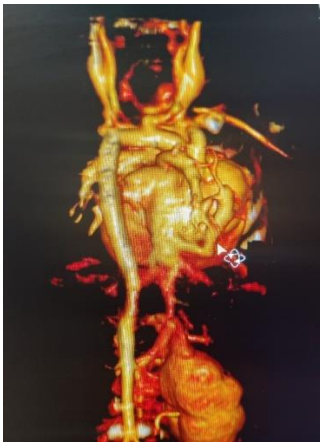

Figure 5: MPR 3D reconstruction showing Anomalous right lower lobe pulmonary venous drainage to supradiaphragmatic IVC.

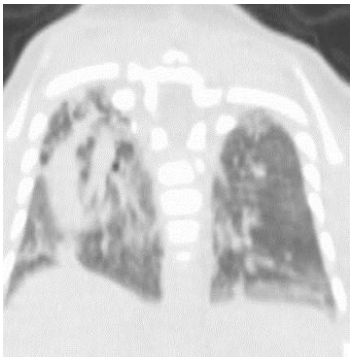

Figure 6: CT with lung window coronal image showing multiple areas of consolidation in the right upper, mid, and lower lobes.

DISCUSSION

Scimitar syndrome is a congenital cardio-pulmonary anomaly and was first described in 1836 (1). The syndrome is a form of anomalous pulmonary venous drainage in which there is partial or total failure of the pulmonary veins to reach the left atrium. Instead, pulmonary venous drainage is anomalously connected to the systemic vein/s, typically to the inferior vena cava, superior vena cava, portal vein, or directly to the right atrium (2). The vein is said to resemble a curved Turkish sword called a "Scimitar". The appearance on chest x-ray is referred to as the “Scimitar Sign”. The other components of the syndrome include hypoplasia of the lower lobe of the right lung, hypoplasia of the right pulmonary artery or its lower branch, abnormal lobar architecture, abnormalities of tracheobronchial architecture and sequestration of the right lower lobe (abnormal vascular supply from a branch of the aorta). Patients presenting in infancy have more severe symptoms and a higher incidence of pulmonary hypertension and cardiac failure (3). Majority of them have cardiac anomalies, most commonly atrial septal defect and ventricular septal defect; and the left-to-right shunt resulting from anomalous pulmonary venous drainage is considerably large. An obstructed anomalous pulmonary drainage and systemic supply to the hypoplastic lung further contribute to the severity of pulmonary hypertension. In the adolescent type, the clinical symptomatology mainly depends on the degree of pulmonary hypoplasia and presents commonly with shortness of breath, fatigue, and failure to thrive, like in our case. In adults, the venolobar syndrome is diagnosed incidentally on chest radiographs obtained for other reasons, as most are asymptomatic (4). They typically have no associated heart anomalies, and left-to-right shunt is insignificant. Various imaging modalities contribute diagnostic information in patients with congenital heart disease. Computed Tomography and Magnetic Resonance imaging are especially useful in demonstrating extracardiac anatomy. Echocardiography continues to be the preferred modality for imaging intracardiac anatomy and hemodynamical studies. The 64-slice CT which is a minimally invasive examination with fast scanning is suitable for the neonates and infants. A thorough preoperative understanding of the complex cardio-pulmonary and vascular anatomy facilitates a directed and prepared surgical approach.

Informed written consent from the patient for publication has been obtained.

DIFFERENTIAL DIAGNOSIS LIST

Pulmonary sequestration
Unilateral absence of pulmonary artery
Swyer james syndrome

Meandering pulmonary vein
Right middle lobe atelectasis

Final Diagnosis: Congenital venolobar syndrome.

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