Case Report

Case of lung aplasia

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Abstract

Pulmonary aplasia is a rare congenital pathology in which there is unilateral or bilateral absence of lung tissue. Here we report a case of a case of lung aplasia who came with repeated respiratory tract infection with difficulty in breathing.

Keywords: Absent pulmonary artery, VACTERL ,White out hemithorax

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Introduction

It is a rare congenital pathology in which there is unilateral or bilateral absence of lung tissue & is distinguished from pulmonary agenesis, although similar, the main difference being that there is a short-blind ending bronchus in aplasia.[1]

It is usually unilateral, as bilateral pulmonary aplasia is not viable. It is frequently associated with other congenital abnormalities, mainly cardiovascular, and has been reported to occur with vacterl syndrome.[1]

Case Report

A one year & five month old female was admitted in pulmonary medicine ward with complaints of gradually progressive breathlesness with repeated episodes of respiratory tract infection.

Computed tomography (CT) showed whiteout of left sided hemithorax or left sided lung volume loss along hyperinflation of right sided lung , left sided mediastinal shift here we couldn't see left sided main bronchus. High resolution CT (HRCT) scan of thorax confirmed the absence of left lung parenchyma and ipsilateral mediastinal shift. There is a left sided absence of pulmonary artery. Volume rendering images also showed absensce of left lung & no left pulmonary bronchus.we have also included virtual bronchoscopy images which showed absence of left main pulmonary bronchus which is replaced by shallow groove.

Discussion

Pulmonary aplasia usually presents with neonatal respiratory distress of variable intensity .In rare cases, it may go unnoticed until later in childhood / adolescence .It can also be discovered during prenatal ultrasound screening, on which it presents as an hyperechoic hemithorax.[2] The diagnosis is frequently hard to make, but mediastinal shift will usually be prominent.Abnormal blood flow in the dorsal aortic arch during the 4th week of gestation had been hypothesized to cause pulmonary agenesis . The contralateral lung may develop as much as twice more alveoli in response to pulmonary aplasia / agenesis The main differential diagnosis is pulmonary hypoplasia. and complete lung atelactasis[2].

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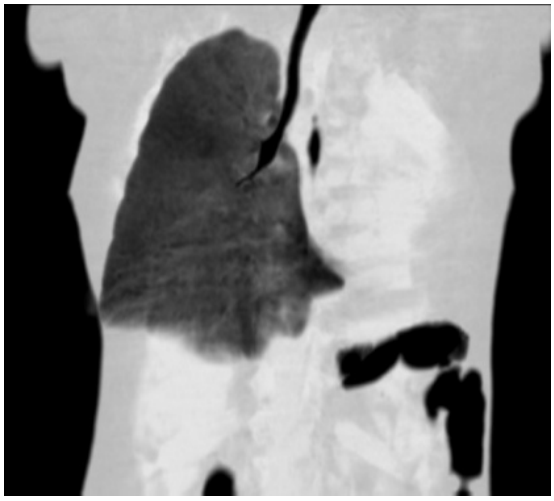

Figure 1. CT MinIP Showing whiteout of left hemithorax & absent left main bronchus

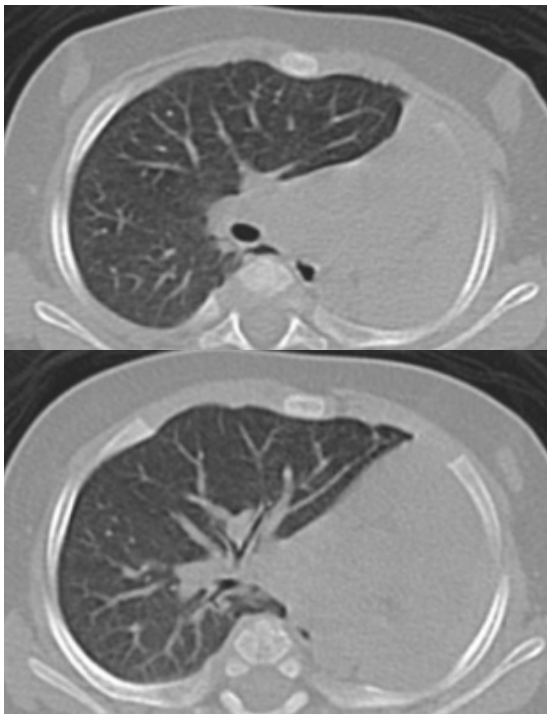

Figure 2a & 2b. HRCT thorax showing absent left lung parenchyma with evident absence of left main pulmonary bronchus

Interruption of the left or right pulmonary artery is an uncommon anomaly. In this congenital anomaly the proximal portion of the main pulmonary artery (arising from the primitive 6th aortic arch) fails to appear during embryologic development.

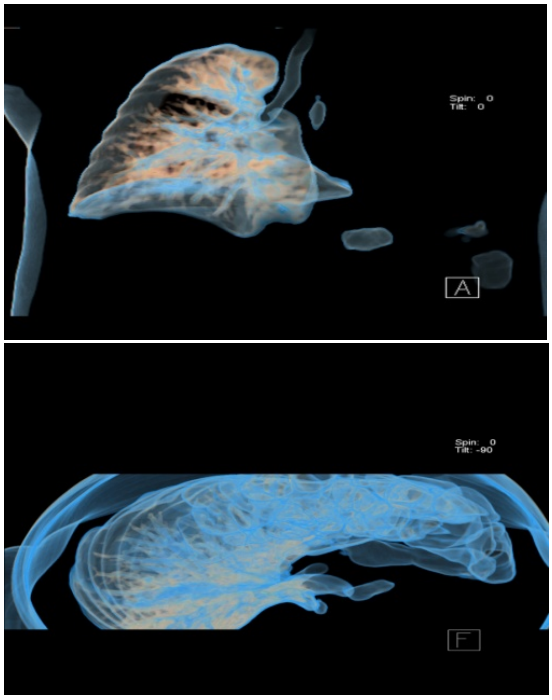

Figure 3a & 3b. Volume Rendering Technique showing no left lung parenchyma with normal right side lung & bronchus

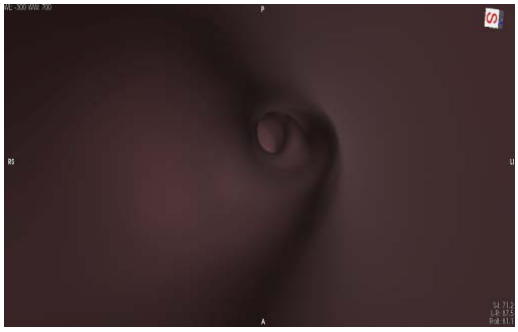

Figure 4. Virtual bronchoscopy showing 'crescent' which determined a shallow groove replaced with a normal main bronchus (of left side)

The term interruption is preferred to absence in view of the fact that it is usually only the proximal section of the vessel that is absent while the more peripheral intrapulmonary arterial network remains intact . This is explained by the different embryologic origins of the proximal and distal pulmonary artery branches. The intact intrapulmonary vessels receive oxygenated blood through systemic collaterals such as the bronchial,

intercostal, or internal mammary arteries or via a PDA.[3]

In most cases the interrupted pulmonary artery lies on the side opposite the aortic arch- therefore, right sided interruption is more common. Interruption of the left pulmonary artery is usually associated with a right sided aortic arch and other cardiovascular anomalies- most commonly tetrology of Fallot . When the interruption is right sided, there may even be an anomalous artery arising from the ascending aorta.[2] Patients with right sided interruption have been grouped into three categories: 1- those having an isolated anomaly (most common); 2- those having an associated left-to-right shunt (usually a patent ductus arteriosus); and 3- those having associated pulmonary hypertension (PAH affects 19-25% of patients and is the most important prognostic indicator) . Individuals in groups 2 and 3 are unlikely to survive beyond infancy, while those in group 1 often present as adults. Recurrent pulmonary infection, hemorrhage, and mild dyspnea on exertion are the most common symptoms. The hemoptysis is usually minor and self-limited.[3]

References

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