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Unilateral Left Lung Aplasia in a 54-Year-Old Lady: Case Report of a Rare Adult Presentation Confirmed by Bronchoscopy and Imaging

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Abstract

Aim & Background: Congenital pulmonary aplasia is a rare embryological defect characterized by complete absence of pulmonary parenchyma and vasculature with a rudimentary bronchial stump. Most patients are diagnosed early in life due to respiratory distress or recurrent infections. Adult detection is extremely rare and often incidental.

Case description: We report a case of a 54-year-old female who presented with intermittent exertional dyspnea. Chest X-ray showed complete opacification of the left hemithorax. High- Resolution Computed Tomography thorax (HRCT) revealed absence of the left lung parenchyma and vasculature, and flexible bronchoscopy confirmed a blind-ending left main bronchus, consistent with left lung aplasia. The patient remains stable with conservative management.

Conclusion: This case highlights the rare adult presentation of congenital pulmonary aplasia, emphasizing the role of imaging and bronchoscopy in diagnosis.

Clinical significance: Early recognition is essential to prevent unnecessary interventions and provide optimal supportive care.

Keywords: Pulmonary aplasia, congenital lung anomaly, unilateral lung absence, adult diagnosis, bronchoscopy

1. Introduction

Congenital pulmonary aplasia is a rare anomaly resulting from incomplete development of the lung bud during the fourth week of gestation. It is classified into three types: agenesis (complete absence of lung and bronchus), aplasia (absent lung with rudimentary bronchus), and hypoplasia (underdeveloped lung). Unilateral lung aplasia accounts for fewer than 0.01% of congenital anomalies, and adult diagnosis is exceptionally rare. We

present a unique case of left lung aplasia diagnosed in a 54-year-old woman through computed tomography and bronchoscopy.

2. Case Description

A 54-year-old lady presented with mild exertional breathlessness for six months. She denied cough, fever, hemoptysis, wheezing, or weight loss. There was no history of childhood respiratory illness, tuberculosis, or prior thoracic surgery. She was a non-smoker and worked as a teacher. On examination, there was tracheal shift to the left, decreased chest expansion on the left side, dullness to percussion and absent breath sounds over the left hemithorax. Her oxygen saturation was 96% on room air.

Chest X-ray showed complete homogenous opacity of the left lung field with ipsilateral mediastinal shift. HRCT thorax [**Figure 1 and 2**] showed total absence of the left lung, left main pulmonary artery and left bronchial tree. Right lung was hyperinflated with herniation into the left thorax. Flexible Bronchoscopy [**Figure 3**] confirmed an abrupt, blind-ending left main bronchus with no segmental bronchi and right bronchial tree was normal. Pulmonary Function Test (PFT) showed mild restrictive defect. Echocardiogram showed normal pulmonary pressures with no evidence of congenital heart disease. Hence was diagnosed with congenital left lung aplasia. She was educated on vaccination, respiratory hygiene, and advised regular follow-up. No surgical intervention was required.



Figure 1: HRCT Chest Lung window showing total absence of the left lung, left main pulmonary artery and left bronchial tree. Right lung is hyperinflated with herniation into the left thorax.

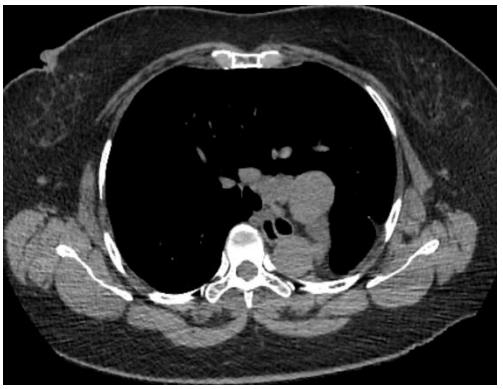


Figure 2: HRCT Chest Mediastinal window showing total absence of the left lung, left main pulmonary artery and left bronchial tree. Right lung is hyperinflated with herniation into the left thorax.

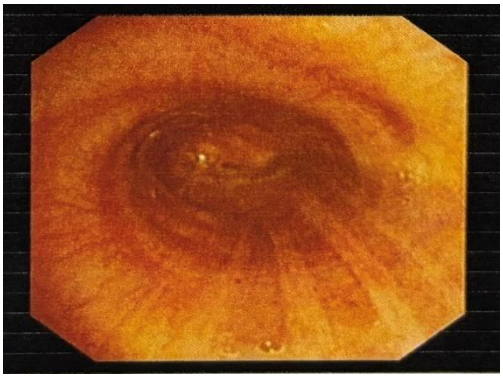


Figure 3: Flexible Bronchoscopy showing an abrupt, blind-ending left main bronchus with no segmental bronchi

3. Discussion

Pulmonary agenesis is a rare congenital anomaly. It was first observed by Pozze in 1673 during an autopsy It was classified into three major types. Type I is Agenesis, which is characterized by a complete absence of lung and bronchus and there is no vascular supply to the affected side. Aplasia is Type II, where there is a rudimentary bronchus with complete absence of pulmonary parenchyma, and Type III is Hypoplasia, where there are variable amounts of bronchial tree, pulmonary parenchyma and supporting vasculature. [1-3] Pathology has been hypothesized as abnormal blood flow in the dorsal aortic arch during the fourth week of gestation. As a result, the contralateral lung may develop as much as twice as many alveoli. [1,2] The major findings in radiography involve the presence of hemithorax whiteout and ipsilateral lung volume loss with ipsilateral shift of mediastinal structures on chest X-ray. [1-4] Absence of lung parenchyma and ipsilateral absence of pulmonary arteries were evident on CT. Contralateral lung hyperinflation is a common finding with herniation of normal lung into contralateral hemithorax. [3,4] Pulmonary aplasia results from incomplete division of the primitive foregut. The left side is less commonly affected but is associated with better prognosis due to preserved right lung function and favourable cardiovascular orientation. [3,4]

This case met the criteria for Type II pulmonary aplasia:

- Absence of pulmonary parenchyma
- Absent pulmonary vasculature
- Presence of a rudimentary, blind-ending main bronchus

Bronchoscopy remains essential in confirming aplasia and excluding differential diagnoses such as post-infective lung destruction, bronchial obstruction, or pneumonectomy.

Differential diagnoses considered included:

- Old pulmonary tuberculosis with fibrotic sequelae
- Destroyed lung from bronchiectasis
- Lung hypoplasia
- Unreported pneumonectomy

However, the absence of prior history, surgical scar, and the presence of a rudimentary bronchus on bronchoscopy confirmed congenital aplasia.

Conclusion

Congenital pulmonary aplasia should be considered in adults with unexplained unilateral chest opacification. Bronchoscopy and CT play a central role in accurate diagnosis. These patients can have a favourable prognosis with conservative management and regular monitoring.

Clinical Significance

Early recognition is essential to prevent unnecessary interventions and provide optimal supportive care.

List of abbreviations

High Resolution Computed Tomography - HRCT
Pulmonary Function Test - PFT

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Declaration of Conflicting Interests

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