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Hypoplasia of Left Lung with Left Pulmonary Artery Diagnosed in Middle Age: A Case Study

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Abstract: ***A rare case of unilateral pulmonary hypoplasia of the left lung associated with left pulmonary artery hypoplasia in a 48 - year - old female is reported. The patient exhibited no significant congenital anomalies in other major organ systems. She had a history of recurrent respiratory tract infections dating back to childhood, suggestive of chronic pulmonary insufficiency. Despite the underdevelopment of the left lung and its vascular supply, there was no evidence of severe cardiopulmonary compromise or additional syndromic features. Pulmonary hypoplasia, a developmental disorder characterized by incomplete lung formation, can lead to varying degrees of respiratory impairment depending on the severity of lung tissue and vascular involvement. In this case, the persistent infections highlight the clinical implications of reduced pulmonary reserve. A thorough diagnostic evaluation, including imaging and pulmonary function assessment, was essential for confirming the diagnosis and guiding management. This case underscores the importance of recognizing rare congenital pulmonary anomalies in adults with chronic respiratory symptoms.***

Keywords: Left lung hypoplasia, left pulmonary artery hypoplasia, contrast - enhanced chest imaging, recurrent respiratory infection

1. Introduction

Unilateral pulmonary hypoplasia or agenesis occurs in approximately 1 to 2 per 12, 000 births, with the left lung being involved in 70% of cases. [1] In contrast, the combination of unilateral pulmonary artery agenesis and lung hypoplasia is rare, with an incidence of about 1 in 200, 000 births. [2] While it is typically diagnosed in childhood, it can remain asymptomatic for a long period. [3]

The symptoms linked to this malformation are not well - defined. The prognosis is influenced by factors such as the extent of lung underdevelopment, the underlying cause, the degree of functional impairment, and the presence of other concurrent congenital organ abnormalities. [4] Consequently, treatment strategies vary depending on these factors.

2. Case Description

Patient Information & primary concerns and symptoms of the patient
A 48 - year - old Hindu woman was admitted to the hospital due to worsening breathlessness on exertion, a productive cough, and intermittent fever persisting for three weeks. She has no history of wheezing, chest pain, loss of appetite, or weight loss. She reports a history of frequent mild respiratory infections since childhood. She has been diabetic for the past five years and is managing it with oral hypoglycemic agents. She is married and has six daughters.

On general examination, the patient exhibited mild pallor and a thin physique. There was no evidence of clubbing, cyanosis, lymphadenopathy, icterus, or edema. Additionally, polydactyly was noted, with the presence of six fingers on her left hand.

Lower respiratory system examination:
On inspection, the patient exhibited left - sided shoulder drooping along with reduced chest expansion on the left side. Palpation revealed a leftward deviation of the trachea and a significantly displaced apex beat, located in the anterior axillary line at the fourth intercostal space. Additionally, rib crowding and increased vocal fremitus were noted on the left side.

Percussion revealed a dull note over the entire left hemithorax, with obliteration of Kronig's isthmus. On auscultation, breath sounds were diminished over the left hemithorax, with bronchial breath sounds and whispering pectoriloquy noted in the left axillary, suprascapular, and both upper and lower interscapular regions. Normal vesicular breath sounds were present over the right hemithorax, with no added sounds bilaterally. Other systemic examinations were unremarkable.

Clinical examination suggested collapse of the left lung with gross mediastinal shift to the same side.

3. Diagnostic Assessment

Investigations:
Routine hematological investigations (Blood counts, Liver function tests, renal function tests, Fasting blood sugar) were within normal limits, HIV test was non - reactive.

Sputum smear for acid - fast bacilli was negative, TrueNAAT – MTB not detected.

Chest roentgenogram revealed dense radio - opacity in left hemithorax, shifting of the mediastinum to left, narrowed intercostal spaces on left side with hyperinflation of right lung (**Figure 1**). CECT chest (

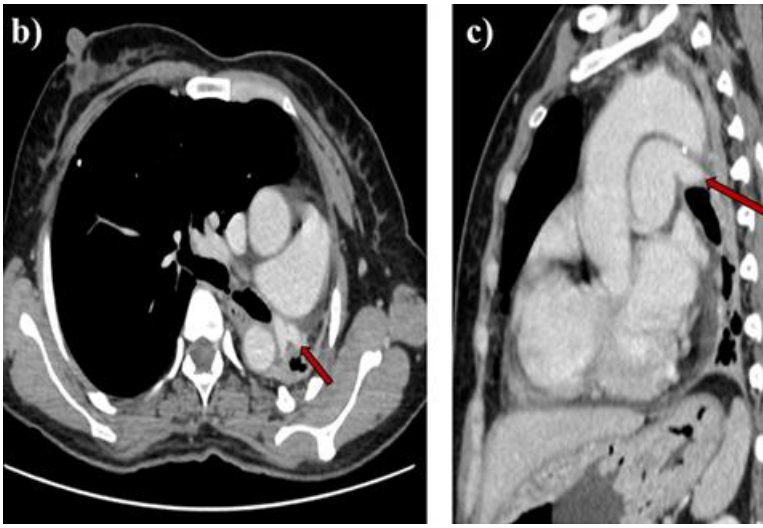


Figure *2*) revealed grossly reduced left lung volume, trachea and mediastinum shifted left with herniation of right lung to the left side and left pulmonary artery hypoplasia.

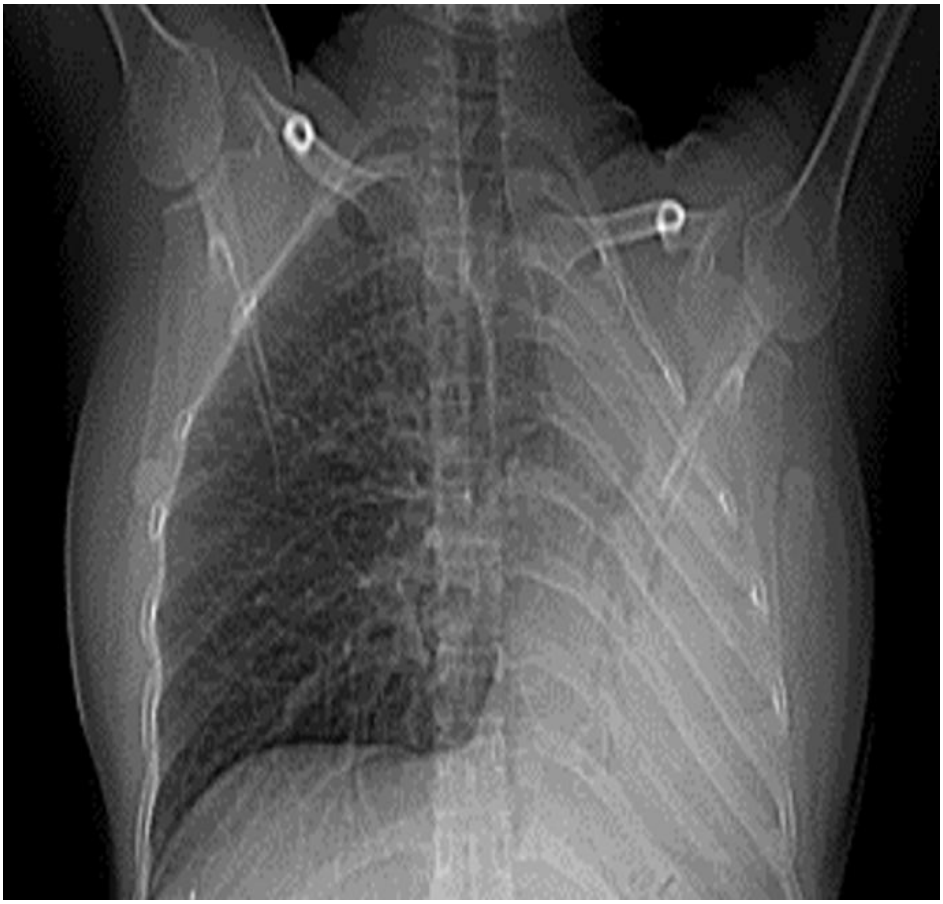


Figure 1: CXR showing radio - opacity in whole left hemithorax, shifting of trachea and mediastinum to left, narrowing of intercostal spaces on left side with hyperinflation of right lung.

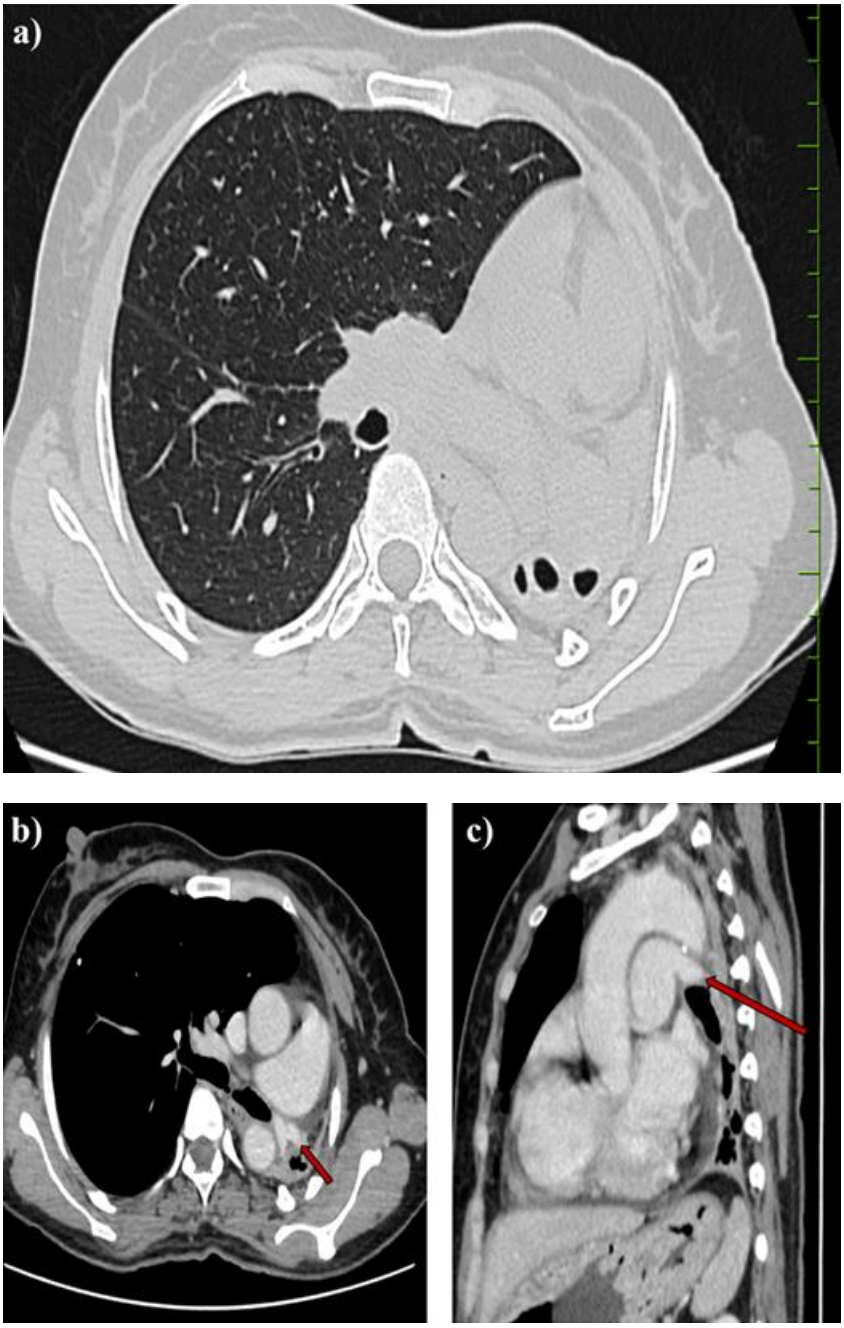
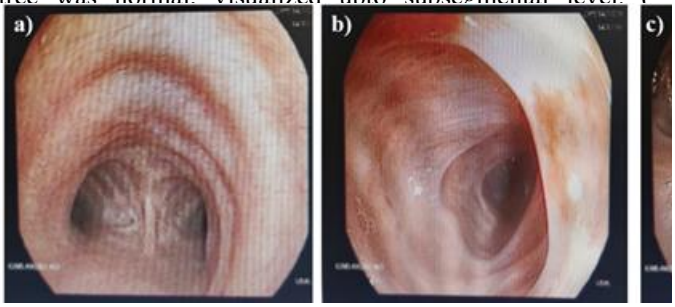


Figure 2: a) CECT thorax (long window) showing complete left lung volume loss, trachea and mediastinal shift toward left with herniation of right lung to left hemithorax, b) mediastinal window and c) sagittal reconstruction arrows (highlighted in red) showing hypoplastic left pulmonary artery.

Bronchoscopy showed normal trachea, narrowing of left mainstem bronchus with left upper lobe bronchus not visualized beyond segmental level and left lower lobe bronchus visualized upto subsegmental level. Right bronchial tree was normal, visualized upto subsegmental level. (**Figure *3***). Thus, the diagnosis of hypoplasia of left main pulmonary artery and left lung parenchyma with bronchial tree is confirmed.



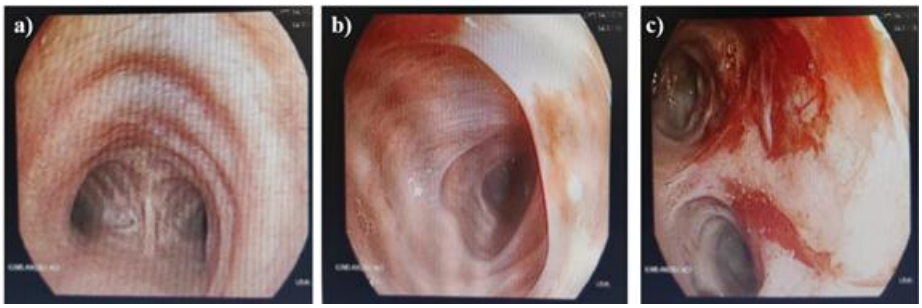


Figure 3: Bronchoscopy image showing a) carina, b) narrowed left main stem bronchi, and c) left upper and lower lobe bronchus

2 - D (Two - dimensional) echocardiography was done to exclude cardiac malformations and pulmonary hypertension; and was found to be normal.

Spirometry showed moderately severe restriction pattern (FVC - 52 % predicted). (**Figure 4**)

4. Therapeutic Intervention

Treatment:

Supportive treatment was given to the patient. She showed improvement with empirical antibiotics.

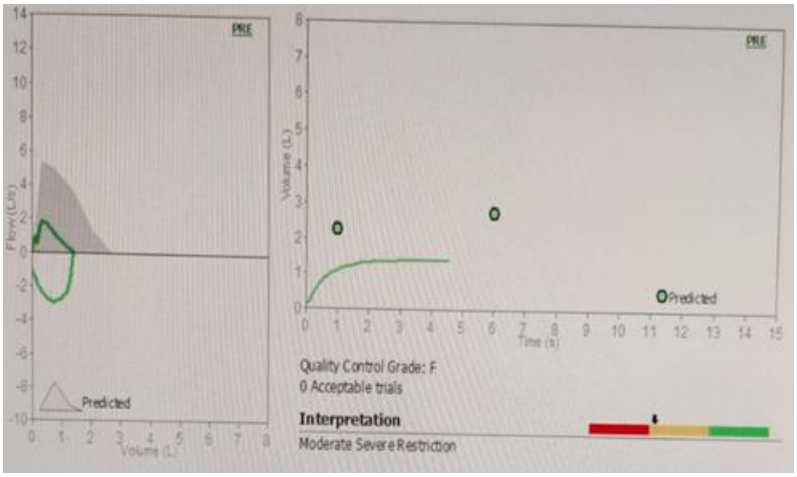


Figure 4: Spirometry showing moderate restriction pattern FVC - 52 % predicted

5. Discussion

Boyden categorizes lung maldevelopment into the following types:

- **Agenesis**: Complete lack of lung tissue, including the airways, parenchyma, and pulmonary artery.
- **Aplasia**: A rudimentary bronchus is present, but there is no lung tissue or pulmonary artery.
- **Hypoplasia**: Inadequate development of the airways, parenchyma, and pulmonary artery, resulting in a small, fibrotic, and nonfunctional lung due to incomplete formation of alveolar tissue. [4]

Monaldi further classified lung maldevelopment into the following categories:

- **Group I**: Lack of tracheal bifurcation.
- **Group II**: Only a rudimentary main bronchus is present.
- **Group III**: Development is incomplete beyond the main bronchial division.
- **Group IV**: Partial development of subsegmental bronchi, with a small part of the corresponding lobe. [5]

The case presented here involves hypoplasia of the left lung and left pulmonary artery, categorized as the third group in Boyden's classification and the fourth group in Monaldi's classification. This case is significant due to its delayed presentation. This case is notable due to its late presentation.

Pulmonary hypoplasia is categorized as either primary or secondary. Secondary pulmonary hypoplasia, which accounts for approximately 60% of cases, is commonly associated with fetal or maternal abnormalities, space - occupying lesions within the thoracic cavity, congenital chest wall defects, and disorders affecting the urogenital or neuromuscular systems. [6] In contrast, primary pulmonary hypoplasia is rare and typically arises from intrauterine events or embryological malformations of the lung or its vascular structures, such as the unilateral absence of the pulmonary artery. [7]

Survival rates are typically higher in cases of left lung hypoplasia, as the larger right lung compensates through hyperinflation. [8]

Unilateral hypoplasia of the pulmonary artery (HPA) results from malformations of the sixth aortic arch during embryogenesis[9], leading to two types: agenesis (proximal interruption of pulmonary arteries) and hypoplasia (rudimentary pulmonary arteries). The left pulmonary artery is most commonly affected. [10] Unilateral hypoplasia of the pulmonary artery (HPA) occurs due to malformations of the

sixth aortic arch during embryonic development[9]; resulting in two types: Agenesis refers to the proximal interruption of the pulmonary arteries, while hypoplasia involves the presence of rudimentary pulmonary arteries. The left pulmonary artery is most frequently affected. [10]

Pulmonary angiography is considered the gold standard for diagnosing vascular pulmonary malformations. However, less invasive methods like perfusion scintigraphy, cardiac catheterization, and MRI are also utilized for diagnosis. [11]

In this case, CT imaging played a crucial role in establishing the diagnosis. The differential diagnosis includes congenital pulmonary airway malformation (CPAM) and Swyer–James–MacLeod syndrome (SJMS). CPAM typically presents as a multicystic pulmonary mass with recurrent infections but lacks associated arterial anomalies. [8] In contrast, SJMS, which is associated with post - infectious bronchiolitis obliterans in childhood, is characterized by a hyperlucent lobe on chest radiography and a mosaic attenuation pattern on chest CT. [1]

Diagnosing the condition in adults is difficult due to the nonspecific symptoms, which often resemble those of acquired diseases. Clinical presentations can range from asymptomatic cases to recurrent pulmonary infections, caused by underdeveloped alveolar tissue, fibrotic lung, surfactant deficiency, and impaired mucociliary clearance, all of which increase the risk of infections. [12]

Early diagnosis is essential for effective monitoring and prompt treatment of complications, including pulmonary infections, hemoptysis, and pulmonary hypertension. Symptomatic treatment includes the use of expectorants, bronchodilators, empirical antibiotics, infection control measures, and vaccination with pneumococcal and influenza vaccines. Asymptomatic cases without additional anomalies may not require treatment. Surgical intervention is infrequently needed for agenesis or aplasia, but in cases with recurrent pneumonia, it may involve limited resection of blind - ending bronchi. [13]

6. Conclusion

This case report presents a rare instance of left lung and pulmonary artery hypoplasia diagnosed in middle age. Despite chronic respiratory symptoms since childhood, the patient adapted without severe cardiopulmonary compromise. It reinforces the need for awareness of congenital pulmonary anomalies, even in adult patients presenting with long - standing respiratory issues.

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