Unilateral pulmonary aplasia: A rare case report

[1]Dr. Mayank Agarwal, [2]Dr Jaypal Singh Chahar

[1]Postgraduate Resident, [2]Assistant Professor
Jaipur national university

Abstract- **Background: Pulmonary aplasia is an infrequent congenital anomaly, the etiology of which is clearly unknown. Other systemic comorbidities such as cardiovascular, gastrointestinal, musculoskeletal, and urogenital system anomalies can be detected in more than half of the patients. It is usually detected during childhood. The patient usually presents in early childhood with recurrent lower respiratory tract infection and respiratory distress. Diagnosis in adulthood is very rare.**
Case report: A 34 years old female presented with complaints of dyspnoea on exertion, chest pain, dry cough and generalised weakness. Chest X ray revealed white out Right sided hemithorax. Rudimentary right bronchus was seen in FOB images. The patient was treated conservatively and the family members were counseled about the anomaly.
Conclusion: Presentation of unilateral lung agenesis in late adulthood, as seen in the current case is extremely rare. As its clinical presentation has a larger range of variability, any adult presenting with opaque hemithorax with ipsilateral mediastinal shift on radiography should always be considered in the list of differentials.

Keywords: **Pulmonary aplasia, congenital anomaly, opaque hemithorax, ipsilateral mediastinal shift.**

Introduction

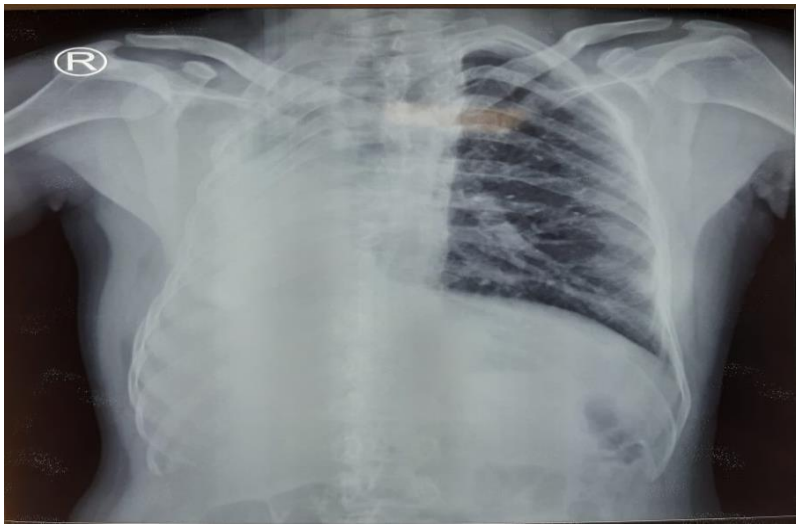
Pulmonary agenesis, aplasia and hypoplasia are few congenital abnormalities of the lung which are rare. Pulmonary agenesis is the complete absence of the lung parenchyma, its vasculature, and its bronchus. Pulmonary aplasia, the most common variant, consists of a carina and the main-stem bronchial stump with absence of the distal lung. Presentation is with usual respiratory symptoms like noisy breathing, fast breathing, repeated respiratory tract infections and respiratory distress.

Case Presentation

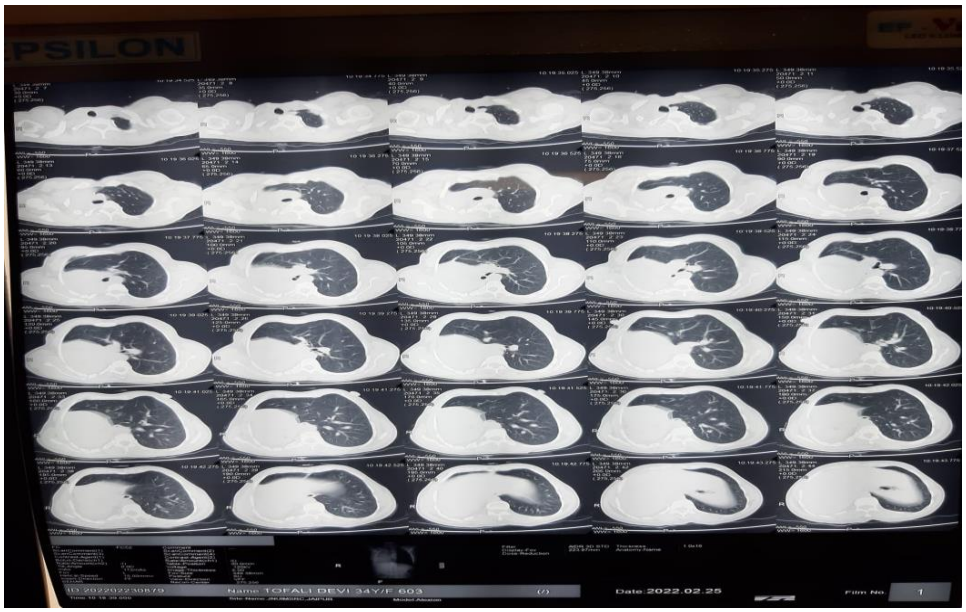
A 34-year-old hindu, female, housewife, resident of Alwar, Rajasthan was reported in the Department of respiratory medicine, JNU hospital with chief complaints of dyspnoea on exertion since 30 days, chest pain since 20 days and dry cough of 15 days. The patient also complained of generalised weakness and fatigability. General physical examination was normal. Routine haematological investigations (Blood counts, Liver function tests, renal function tests) were within normal limits except Haemoglobin (9gm/dl).

Respiratory system examination	
➢ **Inspection** • Trail sign: positive • Right supraclavicular fossa hollowness • Right flattening of chest wall • Asymmetrical Shaped chest • Movements reduced in right hemithorax • Thoraco-Abdominal type of breathing • Respiratory rate 20/min • Apical impulse not visible • No dilated veins, scars, sinuses	➢ **Palpation :** • Findings of inspection are confirmed • No tenderness, local rise of temperature • Trachea shifted to right • Apex beat not palpable • Right sided rib crowding, ICS narrowing • Chest wall expansion– 1cm right hemithorax,3cm on left hemithorax • Chest wall movements diminished over right hemithorax • TVF reduced on right sided hemithorax
➢ **Percussion :** • Direct percussion over clavicle is dull on right side, resonant note on left side • Direct percussion over upper and lower part of sternum was resonant ➢ **Comparative percussion**	➢ **Auscultation :** • Breath sound markedly reduced over Right hemithorax • Vocal resonance : reduced on right hemithorax. • Left hemithorax : normal vesicular breath sounds

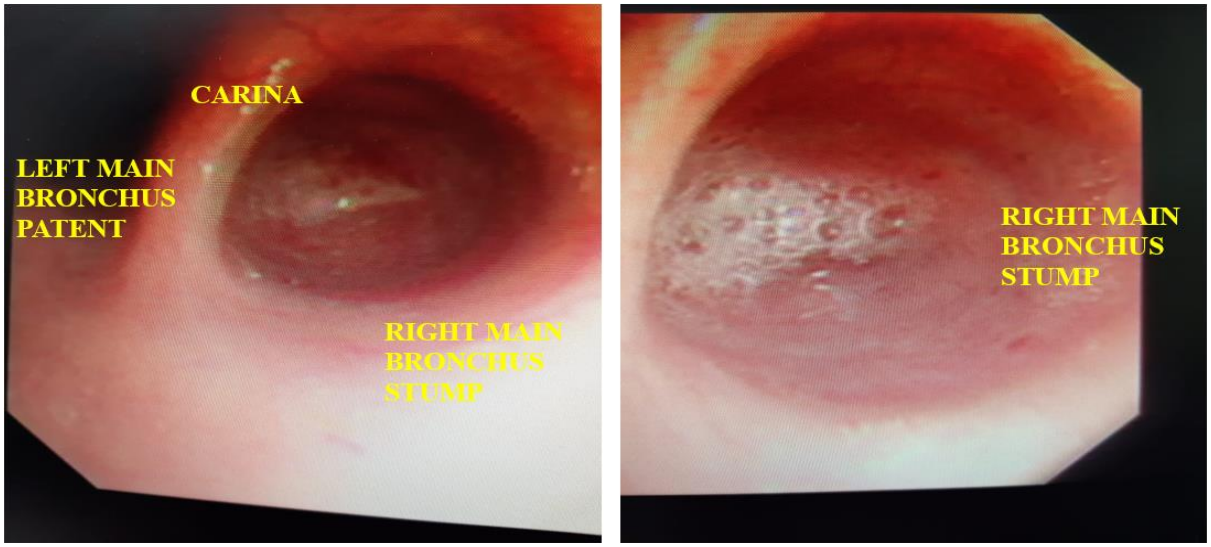
• Right hemithorax is dull, resonant note over Left hemithorax ✓ Cardiac dullness was absent on Left hemithorax	



Chest X ray revealed white out Right sided hemithorax. Trachea pulled towards opacified side.



CECT Thorax- Non-visualization of right lung and bronchial tree with rudimentary stump of right main stem bronchus, compensatory hypertrophy of left lung parenchyma and shift of mediastinal structures with aplasia / severe hypoplasia of right pulmonary artery suggestive of aplasia/ severe hypoplasia of right lung.



FOB images – Rudimentary bronchus seen in right bronchial tree.

Discussion

Pulmonary agenesis encompasses undeveloped pulmonary parenchyma, pulmonary vasculatures, and airways. There is failure of development of the primitive lung bud. Arrest in the development of respiratory system from the foregut corresponds to these embrogenic malformations. Respiratory anlage may develop unilaterally leading to lung agenesis. Unilateral aplasia and hypoplasia are better viable with life. Functionally, unilateral agenesis and aplasia are similar. Sole functional lung is larger than normal to compensate and this enlargement is true hypertrophy and not emphysema.Primary causes include deficiency in TTF-1, HNF310, and EGFR. Secondary causes include small fetal thoracic volume, prolonged oligohydramnios, and congenital heart diseases. A third diagnosed during adulthood are asymptomatic. Recurrent childhood respiratory infections are common symptoms leading to identification. Diagnosis is suspected in patients examined with tracheal deviation, in the presence of a clinically symmetric chest and massive atelectasis in chest xray. Chest CT scan has emerged as diagnostic modality of choice in diagnosis. There is no treatment needed for asymptomatic cases. Treatment is necessary in the presence of respiratory tract infections. Prognosis of pulmonary aplasia depends on severity of co-existing congenital malformations and involvement of normal lung in infection and large vessels kink due to mediastinal displacement. Normal life span is expected for those surpassing first five years without major infection.There is higher mortality with right lung agenesis than left due to greater compression of tracheobronchial tree by shifting of midthoracic structures into the right chest.

Pulmonary Underdevelopment

- Schneider and Schwalbe Classification
  - Group 1 - Bronchus and lung are absent (agenesis)
  - Group 2 - Rudimentary bronchus is present and limited to a blind end pouch without lung tissue (aplasia)
  - Group 3 - Bronchial hypoplasia with variable reduction of lung tissue (hypoplasia)

The present case belongs to the second group of Schneider and Schwalbe classification.

- Monaldi divided the mal-development of lung in four groups.

  - Group I: No bifurcation of trachea
  - Group II: Only rudimentary main bronchus
  - Group III: Incomplete development after division of main bronchus
  - Group IV Incomplete development of subsegmental bronchi and small segmental bronchi of the corresponding lobe.

The present case belongs to the second group of Monaldi classification.

- Boydens - three degrees of mal-development of Lung

  - Agenesis- there is complete absence of lung tissue
  - Aplasia- rudimentary bronchus is present but no lung tissue is present.
  - Hypoplasia-all the normal pulmonary tissues are present but are under-developed.

  The present case belongs to the second degree of Boydens classification.

Conclusion:

- The present case belongs to the second group of Schneider and Schwalbe classification i.e. rudimentary bronchus is present and limited to blind end pouch without lung tissue (aplasia).
- Patient with right lung agenesis have a shorter life expectancy than those with left lung agenesis.
- Bilateral pulmonary agenesis is extremely rare and uniformly fatal.
- >50% of children with pulmonary agenesis have associated other congenital anomalies.

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