The Unilateral Pulmonary Aplasia: Radiologic Clues to a Congenital Anomaly

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Abstract

Pulmonary aplasia is a rare congenital developmental malformation characterized by total absence of lung with rudimentary bronchus. Although the exact cause is not known, several factors have been proposed. Patients can be asymptomatic or can present with recurrent respiratory infections. Multimodality imaging may be needed for confirmation and needs to be differentiated from pulmonary agenesis or severe hypoplasia. This case highlights the key imaging findings of pulmonary aplasia and the need for early diagnosis so as to prevent further complications.

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Introduction

Pulmonary aplasia is a rare congenital anomaly of lung development, characterized by complete absence of pulmonary parenchyma and bronchi distal to a rudimentary main stem bronchus, which ends in a blind pouch. It results from arrest in bronchopulmonary development during the embryonic phase and accounts for a small subset of unilateral lung agenesis/aplasia spectrum disorders. The reported incidence is approximately 1–2 per 100,000 live births (1). Multimodality imaging, particularly X-ray, CT and MR, plays a pivotal role in differentiating aplasia from agenesis and severe hypoplasia, as well as excluding acquired causes of lung volume loss. This case report highlights the key imaging findings of pulmonary aplasia and underscores the role of radiology in accurate diagnosis and clinical management.

Case Presentation

A 4 year-old male child, developmentally normal and fully immunized for his age presented to the emergency department with complaints of fever, cough, uprolling of eyes and stiffening of bilateral upper and lower limbs. He has a past history of admission at 10 months of age which was treated as a lower respiratory tract infection. Chest x-ray taken showed tracheal shift to left, volume loss in the left lung with compensatory hyperinflation of the right hemithorax, crowding of ribs on left, and whiteout left lung with a homogenous radio opacity in the left hemithorax causing silhouetting of left heart border, dome of diaphragm, costophrenic angle (Figure 1). Contrast enhanced CT thorax with pulmonary angiogram was taken for further evaluation which showed mediastinal shift to left and non visualization of the entire left lung parenchyma with hyperinflation of right lung fields (Figure 2). Left main bronchus was tapering distally and blind ending (Figure 3). Big bronchus

was noted with right upper lobe bronchus arising directly from the right margin of distal trachea (Figure 4). Left pulmonary artery and veins were not visualized. Double Superior Vena Cava (SVC) noted with left SVC draining into coronary sinus (Figure 5). Both brachiocephalic veins noted to join and form right side SVC. Overall features were suggestive of left lung aplasia. Bronchoscopy was performed which confirmed bronchus suis (Figure 6) and blind ending left main bronchus. The child was started on antibiotics, became symptomatically better and got discharged after advising regular chest physiotherapy and prompt treatment of infections.

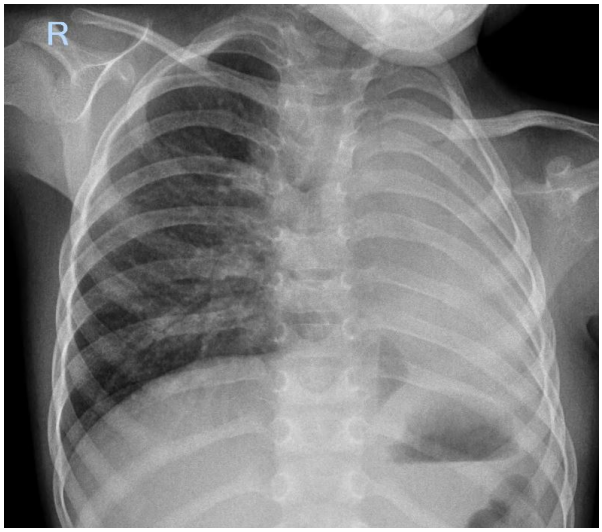

Figure 1: *Chest X-ray showing whiteout left lung.*

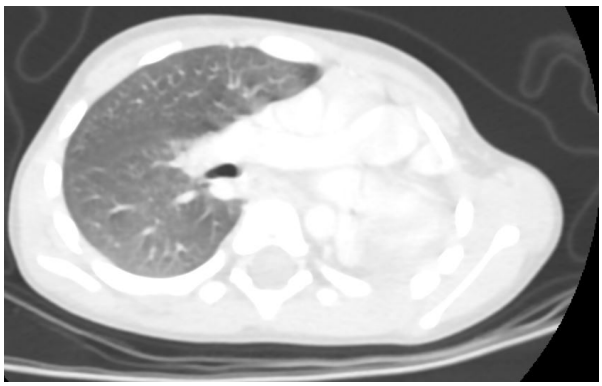

Figure 2: *Contrast enhanced CT Thorax axial image showing non visualization of entire left lung parenchyma, hyper inflated right lung field.*

Discussion

Pulmonary agenesis is an extremely rare congenital entity. Pulmonary agenesis was originally classified by Schneider and later revised by Boyden, who divided the condition into three types based on the developmental stage of the primitive lung bud. Type I, known as pulmonary agenesis, involves the complete absence of lung tissue, bronchus, and associated blood vessels on one side. Type II, referred to as pulmonary aplasia, is characterized by the total absence of

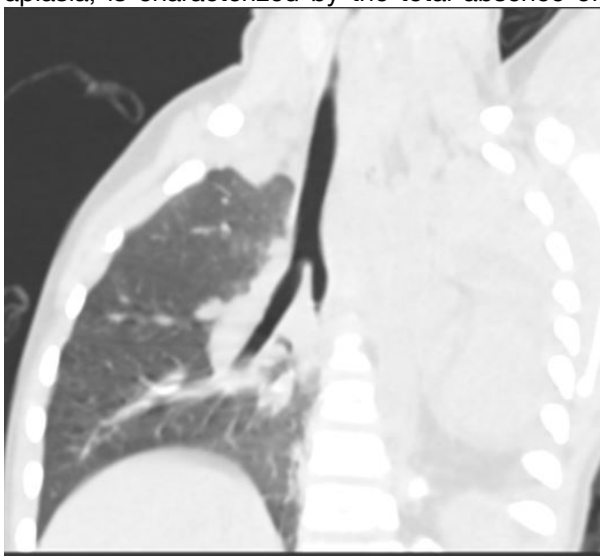

Figure 3: *Contrast enhanced CT Thorax Coronal image showing blind ending left main bronchus.*

one lung but with a rudimentary bronchus still present. Type III, called pulmonary hypoplasia, features partial development of the bronchial tree along with some lung tissue and blood vessels on the affected side (2). Although the exact cause of the condition remains unclear, several factors

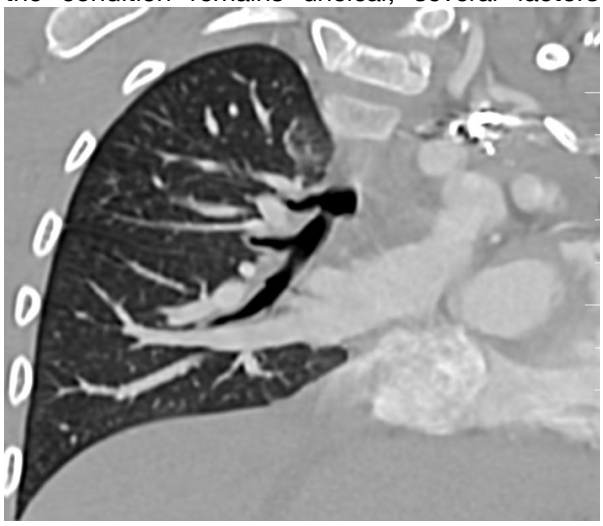

Figure 4: *Contrast enhanced CT Thorax Coronal image showing right upper lobe bronchus arising from right margin of distal trachea.*

have been proposed, including vitamin A deficiency during pregnancy, viral infections, genetic influences, and medical interventions (2). It can be associated with other congenital malformations also that involve the cardiovascular, gastro-

intestinal, skeletal, and genitourinary systems (3). Early recognition is critical, as many patients remain asymptomatic and may be incidentally diagnosed, while others present with recurrent infections or respiratory distress.

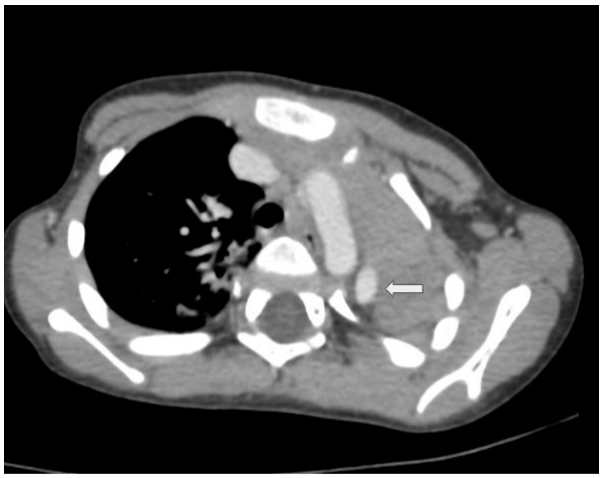

Figure 5: *Contrast enhanced CT Thorax Coronal image showing double SVC(arrow).*

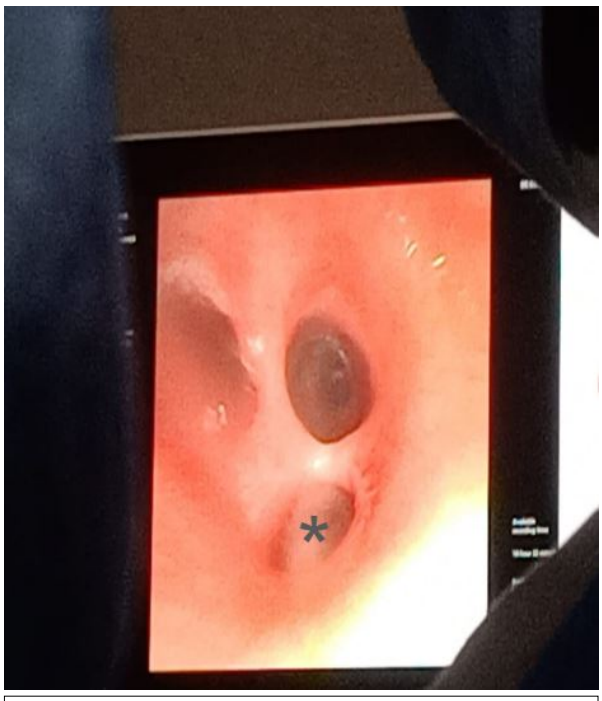

Figure 6: *Bronchoscopy image showing bronchus suis (asterix).*

Pulmonary aplasia can sometimes be asymptomatic and patients can survive upto adulthood. Predisposition to pulmonary infections is a risk factor which can even be fatal. Chest X-ray is the primary radiographic evaluation which shows complete absence of hemithorax, crowding of ribs, hyperinflation of contralateral lung field, mediastinal shift. This can mimic complete atelectasis from any cause. CT provides exquisite details about the anatomy of tracheobronchial tree, lungs, heart, and major mediastinal vessels (1). The presence of rudimentary bronchus, absent ipsilateral pulmonary vasculature confirms the diagnosis as in our case. Treatment depends on symptoms with majority of patients treated conservatively and surgery is rarely required (4).

Conclusion

Pulmonary aplasia is a rare congenital pulmonary anomaly that may remain undiagnosed until adulthood, especially in asymptomatic individuals or those with nonspecific respiratory complaints. Cross-sectional imaging, particularly CT, is crucial in establishing the diagnosis by revealing the absence of lung parenchyma, bronchial development, and pulmonary vasculature on the affected side. Radiologists should be familiar with this entity to differentiate it from more common causes of whiteout lung such as collapse, pneumonectomy, or pulmonary hypoplasia. Accurate diagnosis can prevent unnecessary interventions and guide appropriate clinical management, especially in cases with associated anomalies.

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