

## Left upper lobar agenesis of lung: A rare case report

Sir,

Underdevelopment of lung is a group of congenital anomalies classified into agenesis, aplasia, and hypoplasia of lung, on the basis of presence or absence of lung parenchyma, bronchial tree, and pulmonary artery. The pathogenesis of such anomalies is not accurately known with several proposed theories. It is often associated with anomalies of the cardiovascular, genitourinary systems. An isolated left upper lobe pulmonary agenesis is rare and more commonly involves the right upper and middle lobes.<sup>[1]</sup> It is associated with the absence of lung parenchyma, bronchial tree, pulmonary vasculature, and remains undiagnosed until adulthood. Treatment depends on patient's symptoms, with most patients being treated conservatively.

A 29-year-old male, with no major illness (nondiabetic; nonhypertensive) or significant past surgical history, got a chest radiograph done as a part of routine health checkup. The radiograph revealed mild volume loss of left lung with abnormal homogenous opacity in the left upper zone. His respiratory examination revealed relatively reduced breath sounds in the left upper chest as compared to right side. His cardiovascular examination was unremarkable. Later, the patient underwent a contrast-enhanced computed tomography scan (CECT) chest for further evaluation of the radiographic findings.

The chest radiograph posterior-anterior view [Figure 1] revealed volume loss of left lung field with the collapse of left upper lobe suggested by the left upper zone opacity, superior migration of the hilum, fissure, and elevation of the left hemidiaphragm. Secondary changes were seen in

the form of hyperinflated right lung, herniation across the midline, and shift of mediastinum to the left.

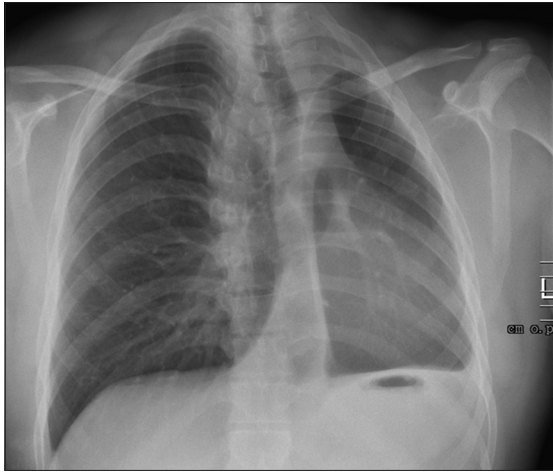
CECT chest [Figure 2] revealed the absence of left upper and lingular bronchus with complete agenesis of the left upper lobe (along with its lingular segment). The left main bronchus was seen continuing as the left lower lobe bronchus [Figure 3]. Compensatory hyperinflation with herniation of the right lung across the midline was also seen. The main pulmonary artery was short in length with early bifurcation [Figure 4]. There was hypoplasia of the left pulmonary artery (diameter of 9 mm at origin) which continued as the left lower lobe pulmonary artery without any upper lobe branch. No other associated congenital anomaly was found.

Lobar agenesis is a very rare entity, resulting from the anomalous formation of primitive lung bud. The incidence of the disease is 0.0034% with no gender predisposition. The first case was reported in 1673 by De Pozze in the autopsy of an adult female.

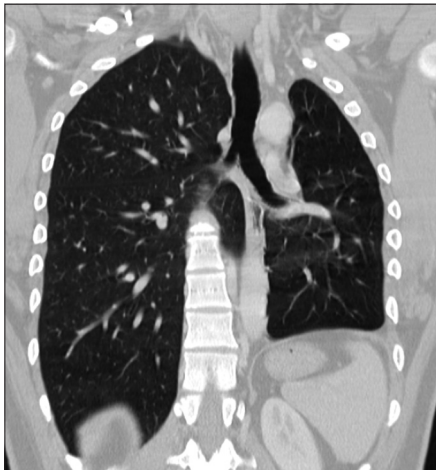
Lobar agenesis is included in "underdevelopment of lung" and is classified into three categories:

1. Agenesis shows complete absence of bronchus, lung parenchyma, and associated vessels
2. Aplasia has a rudimentary bronchus with absent lung parenchyma
3. Hypoplasia has hypoplastic bronchus and lung parenchyma.<sup>[2]</sup>

The etiopathogenesis is unclear; however, genetic, teratogenic agents (allopurinol),<sup>[3]</sup> and Vitamin A deficiency during pregnancy<sup>[4]</sup> have been hypothesized as its causes.



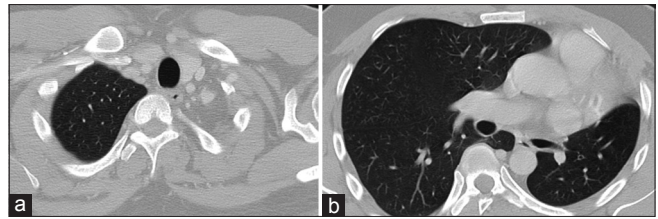
**Figure 1:** Chest X-ray shows volume loss on left side with homogenous opacity in the left upper zone with tracheal and mediastinal shift toward left side, compensatory hypertrophy of left lower lobe and right lung field, herniation of right lung field across midline, elevated left hemidiaphragm, and blunting of left costophrenic angle



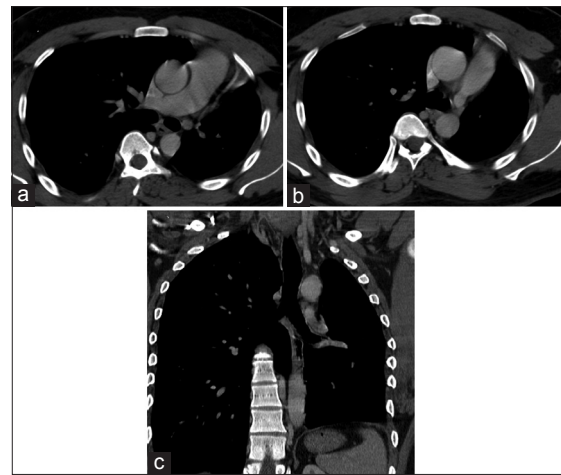
**Figure 3:** Computed tomography chest (lung window) coronal view reveals left upper lobar agenesis with the left main bronchus continuing as the left lower lobe bronchus, compensatory hyperinflation of left lower lobe and right lung field, herniation of the right lung across the midline, and elevation of left hemidiaphragm

The lung development starts at 4 weeks of gestation from the ventral wall of the foregut. A laryngotracheal groove develops from the ventral wall of the pharynx, dividing into the right and left lung buds at around 28<sup>th</sup> day of gestation. A tracheoesophageal septum separates the trachea and esophagus. Definitive human lung development is divided into five phases: Embryonic, pseudoglandular, canalicular, saccular, and alveolar. Insult during the phase of lung bud development is the probable cause of underdevelopment of lung.<sup>[2]</sup>

The abnormality goes undetected or is incidentally detected in the majority of cases. Symptomatic patients present in early childhood with symptoms of recurrent respiratory tract infection while others remain asymptomatic. Underdevelopment of lung is



**Figure 2:** (a) Computed tomography chest (lung window) axial view at T3 vertebral level reveals the absence of left upper lobe and visualization of normal right upper lung. (b) Computed tomography chest (lung window) axial view at T7 vertebral level reveals left main bronchus continuing as the left lower lobe bronchus with mediastinal shift toward ipsilateral side, compensatory hyperinflation of right lung field, and herniation of the right lung across the midline



**Figure 4:** Contrast-enhanced computed tomography chest axial and coronal views reveals. (a) The main pulmonary artery short in length and shows early bifurcation with normal right pulmonary artery, (b) hypoplasia of left pulmonary artery (diameter of 9 mm at origin), and (c) the hypoplastic left pulmonary continues as left lower lobe pulmonary artery without any upper lobe branch

usually associated with other congenital abnormalities of cardiovascular, musculoskeletal, and genitourinary system, more commonly toward the ipsilateral side. Mardini-Nyhan association<sup>[5]</sup> is seen in consanguineous marriages, comprising congenital heart disease, thumb abnormality, and complete/partial lung agenesis. Our case did not have any other associated congenital abnormality.

History of pneumonectomy, segmental or lobar collapse, and thickening of pleura are few differential diagnosis<sup>[4]</sup> to be thought off. Thus, proper history taking is critical in these cases. Chest CECT is the most conclusive examination<sup>[6]</sup> for the diagnosis of the spectrum of underdevelopment of lung diseases. The role of angiography is important as it clearly entails the associated vascular abnormalities. These patients are managed conservatively, and surgery is rarely required as in symptomatic patients.

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**Conflicts of interest**

There are no conflicts of interest.

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