

## CONGENITAL LOBAR EMPHYSEMA MIMICKING PNEUMOTHORAX: A CASE REPORT

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### ABSTRACT

A 2-month-old infant was admitted with a diagnosis of acute bronchopneumonia, respiratory failure and diminished air entry on one side of the chest. Chest x-ray revealed a hyperlucent hyperinflated left lung and a mediastinal shift. Chest tube drainage was done considering tension pneumothorax as a possibility. Subsequent chest x-ray did not show any improvement. On carefully examining x-rays, diagnosis of Congenital Lobar Emphysema (CLE) was considered which was confirmed by CT scan of the chest. Infant improved after surgical removal of the involved lobe of the lung. This case highlights the importance of sound clinical practices and careful radiological interpretation before embarking on any surgical intervention.

### KEYWORDS

Congenital Lobar Emphysema, Pneumothorax.

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### INTRODUCTION:

**CASE:** A 2-month-old male baby weighing 4.7 kg was referred to paediatrics emergency ward of our hospital with problem of worsening bronchopneumonia with left-sided pneumothorax. Infant was born full term, normal vaginal delivery with birth weight 2.9 kg without any prior known congenital anomalies. At admission, he was irritable, sick looking, had severe tachypnoea with respiratory rate 86 breaths/min., pulse rate of 180/min., and blood pressure of 90/60 mmHg. His temperature was normal. O<sub>2</sub> saturation was 80% in room air. A chest examination revealed an asymmetrical expansion of the left chest, nasal flaring, subcostal retraction, slightly decreased air entry on left side, with diffuse wheeze and few crepitations. Blood gas at admission showed pH 7.47, PO<sub>2</sub> 62 mmHg, PCO<sub>2</sub> 62 mmHg, and HCO<sub>3</sub> of 23.8 mmol. A clinical diagnosis of acute bronchopneumonia with wheeze with left-sided tension pneumothorax with respiratory failure was made. The infant was started on nebulised epinephrine, oxygen via head box and supportive care. Percutaneous needle insertion done in second left intercostal space revealed pneumothorax which was followed by chest tube insertion. After one hour of admission, the clinical picture deteriorated. Repeat blood gas showed pH 7.13 with PCO<sub>2</sub> 92. Infant was connected to ventilator on SIMV mode. Radiographs of the chest revealed a hyperlucent hyperinflated left lung and a mediastinal shift (Figure 1). During the next few days, baby did not show any improvement with failure to extubate. Repeat chest x-ray revealed same radiological findings along with herniation of the left lung to right side raising suspicion of CLE (Figure 2).

Thoracic CT showed that there was a highly localised hyperlucent region in the LUL with attenuated bronchovascular markings; with a mediastinal shift to the contralateral right side likely related to LUL over-inflation suggesting left upper lobe CLE (Figure 3). After consultation with a paediatric surgeon, surgical intervention was suggested. Left upper lobe CLE was noted during the operation (Figure 4). A histopathological examination of the excised left lower lobe revealed changes consistent with CLE. The infant was successfully extubated within 48 hours after surgery. He was discharged on seventh postoperative day. Chest x-ray revealed normal lung expansion with resolution of collapse of left lower lobe and resolution of right upper lobe pneumonia.



**Fig. 1: Chest X-ray showing Hyperinflated Left Lung and Mediastinal Shift**



**Fig. 2: Chest X-ray showing Herniation of the Left Lung to Right Side and Chest Tube in situ**

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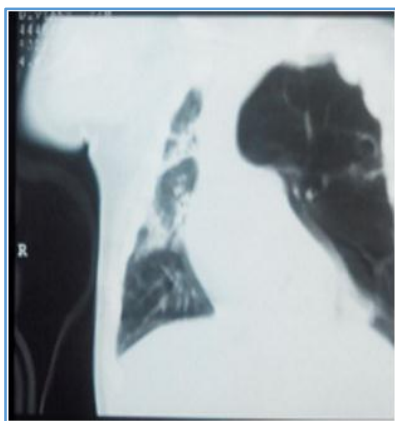
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**Fig. 3: CT Thorax showing Left Upper Lobe Congenital Lobar Emphysema (CLE) with Left Lower Lobe Collapse and Right Upper Lobe Pneumonia**



**Fig. 4: Left Upper Lobe CLE noted during Surgery**

**DISCUSSION:** Congenital lobar emphysema (CLE), also known as infantile lobar emphysema or congenital lobar hyperinflation, is a congenital lung anomaly that is characterised by progressive hyperinflation of a lobe. The prevalence of CLE is 1 in 20,000-30,000 and the incidence is 1 in 70,000 to 1 in 90,000. Male babies are affected more often than female in the ratio of 3:1 and most commonly involves left upper lobe (In 42% cases).<sup>(1,2,3)</sup> Approximately 25 to 33 percent of cases present with respiratory distress at birth, 50 percent by one month of age, and nearly all by six months of age; although rare cases are reported in adults also.<sup>(4,5)</sup> The characteristic radiographic appearance of CLE is asymmetric hyperlucency of the lobe. The prominent differentials for cystic lucent chest lesion on chest radiograph in infants and young children (Under 3 years of age) with respiratory distress are post-infective pneumatocele, pneumothorax, congenital diaphragmatic hernia (CDH), congenital cystic adenomatoid malformation (CCAM), congenital lobar emphysema (CLE), pulmonary sequestration, bronchogenic cyst, and bullous lung disease. In developing countries, the incidence of chest infection and infection-related complications (pneumothorax and pneumatoceles) is high, and they are considered to be the more common causes of lucent cystic lesions on chest radiograph in patients with respiratory distress. Chest tube insertion into congenital cystic lung lesions, mistaking them

for an infection-related complication, is a recurring clinical problem which has been reported in the literature as isolated case reports/series and is associated with increased rate of complications in the patients. The major errors in diagnosis leading to chest tube insertion are due to the inability to differentiate CCAM from tension pneumothorax/pneumatocele and CLE from a tension pneumothorax.<sup>(6,7)</sup> In our patient too; hyperlucency of one side on chest radiograph in an infant with severe respiratory distress was mistaken as tension pneumothorax with resultant chest tube insertion. It is important to look at the x-rays carefully so as to distinguish it from pneumothorax. Both CLE and pneumothorax share a common feature in a prominent mediastinal shift towards the unaffected lung (opposite the hyperlucent lobe/region). The presence or absence of bronchovascular markings in the hyperlucent area is the main distinguishing factor between the two conditions on plain chest x-ray.

The overinflated lobe acts as a space-occupying lesion that produces a mass effect on adjacent structures leading to collapse of other lobes. CLE compression of the adjacent lobes pushes the cephalad or caudal toward the diaphragm, whereas in pneumothorax, the lung collapses toward the hilum.<sup>(5,6,7)</sup> CT is an excellent imaging modality to exclude an underlying hilar mass or intraluminal bronchial process. It typically shows both the hyperinflated lobe and the attenuated bronchovascular bundle, which runs at the periphery of the expanded alveoli. Additionally, CT can precisely delineate and localise the lesion, which is particularly helpful for preoperative evaluation. Treatment approach is different in the CLE and tension pneumothorax; whereby severely symptomatic CLE requires surgical intervention with a lobectomy while large symptomatic pneumothoraces require aspiration or chest tube drainage, which may worsen the outcome of CLE patients.

The prognosis is usually excellent after resection of congenital lung lesion.<sup>(2,8)</sup>

We report this case to elucidate that CLE can mimic tension pneumothorax. Clinicians must have a high index of suspicion for CLE in the context of respiratory distress in infancy and should not mistake CLE for pneumothorax or tension pneumothorax leading to inadvertent insertion of chest tube in CLE which can be harmful. Early recognition and effective treatment of CLE can be life-saving.

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