

## A three month old infant with severe respiratory distress

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### Abstract

This case report discusses the diagnostic challenge of congenital lobar emphysema (CLE) in a three-month old infant with severe respiratory distress. The infant was initially misdiagnosed and managed as a case of pneumothorax. This case highlights the importance of CT scans as a diagnostic tool for early diagnosis and lifesaving management of CLE. It also signifies the need for adequate funds and infrastructure in the health care system especially in rural areas of developing countries like Pakistan.

**Keywords:** Congenital lobar emphysema, respiratory distress, CT scan.

**DOI:** <https://doi.org/10.47391/JPMA.9553>

### Introduction

Congenital Lobar Emphysema (CLE) is a rare obstructive pulmonary disorder, usually presenting in the neonatal period or in early infancy, characterised by unilateral hyperinflation of the affected lung lobe. The aetiology of this disease is mostly unknown, and it may be associated with other developmental anomalies, including the absence of cartilage in the airway and small sized alveoli. CLE can present with respiratory distress in infancy, enlarged chest due to hyperinflation, and cyanosis. However mild forms of CLE can be asymptomatic and may appear later in life <sup>1</sup>. CLE most commonly affects the upper lobe of the left lung and is more common in males <sup>1</sup>. Along with hyperlucency, the initial chest x-ray may show atelectasis and increased density of the adjacent lung, which can be mistaken for pneumonia <sup>2,3</sup>. It can also be mistaken for pneumothorax.

### Case Report

A three-month-old male infant presented at Medicenter General Hospital in Karachi in December 2019, with a three-day history of severe, worsening respiratory distress and a chest tube placed on the left side of his chest. The

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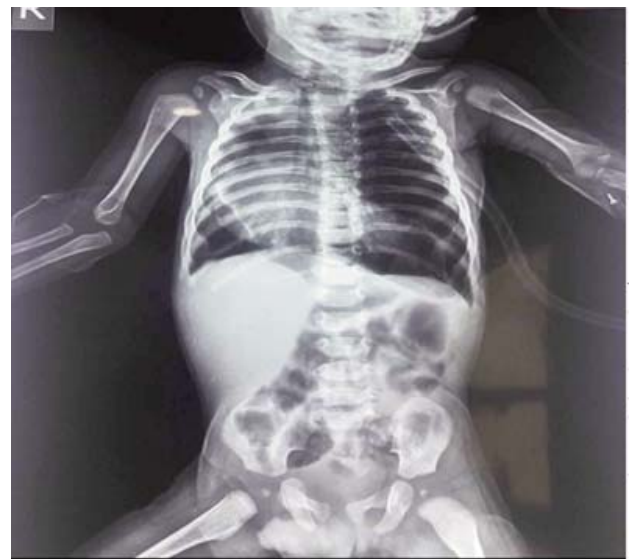
**Submission complete:** 28-04-2023

**Review began:** 07-06-2023

**Acceptance:** 23-12-2023

**Review end:** 05-11-2023

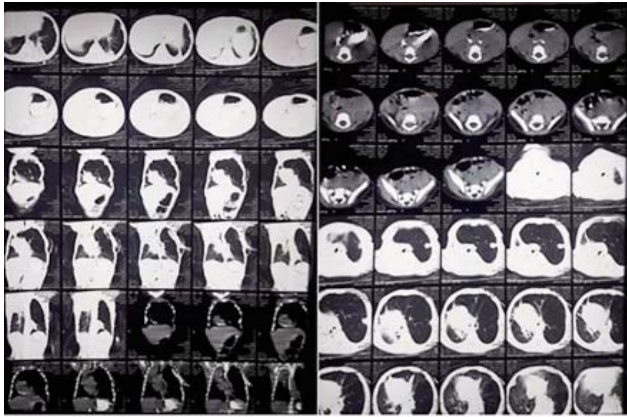
chest tube was inserted two days prior to presentation at a tertiary care hospital based on a frontal plane chest x-ray showing a hyperlucent and hyperinflated left hemithorax with shifting of the mediastinum to the opposite side (Figure 1).



**Figure-1:** Chest x-ray showing a hyperlucent and hyperinflated left hemithorax (1) with shifting of the mediastinum to the opposite side (2)

The infant was an only child and was born to a 30-year-old mother via spontaneous vaginal delivery in a rural hospital. The pregnancy was uneventful, but no ultrasound scans were done during the antenatal period. The immediate postnatal period was normal. The infant was breastfed and immunised according to local health guidelines. The parents denied any history of respiratory diseases in the child.

On examination, the infant was tachypnoeic, restless, and irritable. Other vitals showed systolic blood pressure of 60mmHg, respiratory rate of 68 breaths per minute, and heart rate was 150 beats per min. Flaring of ala nasi with subcostal and intercostal recessions and blue discoloration of lips were observed. On chest examination, the infant had a hyper-resonant percussion note in the left hemithorax and the apex beat was shifted towards the right side at fifth intercostal space lateral to the right midclavicular line. Remaining systemic examinations were insignificant.



**Figure-2:** CT scan chest showing hyper-inflation of the left lung with a hyper-lucent region and visible lung markings

Arterial Blood Gases (ABGs) showed a saturation of 80%, pH of 7.28, PaCO<sub>2</sub> of 60 mmHg, PaO<sub>2</sub> of 55mmHg, and HCO<sub>3</sub><sup>-</sup> 20mEq/L. The complete blood count was within normal parameters.

On admission, the infant was administered IV bolus of normal saline and was continued on maintenance fluid. He was also given inotropic support (dopamine) and was started on empirical antibiotics as part of conservative management. The infant was referred to radiology for a CT scan. The CT scan chest showed hyper-inflation of the left lung, with a hyper-lucent region and visible lung markings (Figure 2). A diagnosis of congenital lobar emphysema of the left lung was established.

The infant had undergone severe respiratory compromise due to delay in diagnosis and mismanagement. A surgical consult therefore advised surgical resection of the left emphysematous lobe but, unfortunately, the patient expired the next day before he could be taken for surgery.

## Discussion

Congenital lobar emphysema is a rare but important developmental anomaly of the lung, characteristically presenting in neonates and infants. The exact cause of this life-threatening condition is unknown. Despite advanced diagnostic techniques, pitfalls in the diagnosis and management of congenital lobar emphysema exist<sup>3</sup>.

Conventional chest X-rays are typically used to establish the diagnosis of CLE and usually show a unilateral hyperlucent hemithorax with shifting of mediastinum to the opposite side. However, this finding is also present in tension pneumothorax – the main differential diagnosis on routine chest radiography<sup>4</sup>. Therefore, while chest radiography remains the first investigation of choice in patients presenting with respiratory distress, a CT scan is considered the gold standard to diagnose CLE. CT scans

not only evaluate the affected lobe, but the surrounding structures as well<sup>5</sup>. A retrospective study by Abdel-Bary and colleagues determined that although chest radiography can be used to identify congenital lobar overinflation, a CT scan is required to verify the radiographic findings and to rule out other anomalies<sup>6</sup>.

We describe the case of a three-month-old male infant who was initially misdiagnosed and managed as a case of pneumothorax based on his presentation and his chest radiographic findings. A CT scan was not done initially, and a chest tube was inserted in the left hemithorax, but it showed no improvement in the condition of infant.

The infant was referred to a private hospital due to continued worsening of his condition where a CT scan of the chest was performed which subsequently led to the diagnosis of CLE. The scan demonstrated a hyperlucent and hyperinflated left hemithorax with mediastinal shift to the right side; but it also revealed lung markings and septae in the area, which confirmed the presence of hyperinflated lung parenchyma rather than a pneumothorax. It also demonstrated that the changes were due to left upper lobe emphysema.

Management of pneumothorax and congenital lobar emphysema differs greatly. The diagnosis of CLE requires a high degree of suspicion on clinical grounds. In such cases, the insertion of an intercostal drainage tube may be detrimental to the patient<sup>4</sup>. Early diagnosis and immediate surgical intervention can prove to be lifesaving in symptomatic cases of CLE<sup>7</sup>. While there is no definitive cure for emphysema and patients continue to live with shortness of breath that can be challenging, surgical treatment is a highly effective therapy with low rates of morbidity and mortality. Prompt removal of affected lobe of the lung is indicated due to risk of infection along with interference with normal lung growth which worsens the case and makes the surgery more difficult<sup>8</sup>. Newer therapies such as bronchoscopic lung volume reduction using Zephyr valves are being developed and studied. Some trials have shown that these newer therapies have clinically meaningful benefits in terms of post-operative lung function, exercise tolerance, dyspnoea, and improved quality of life<sup>9</sup>. In the case of our patient, the diagnosis was made too late to provide appropriate treatment. CLE should be suspected whenever there is respiratory distress, cyanosis, asymmetrical breath sounds, displaced cardiac sounds, and tympanitic chest percussion, all occurring within a few days to weeks of birth<sup>3</sup>.

CT scans, although a diagnostic tool of choice for confirmation of CLE, are expensive<sup>10</sup>. In developing

countries such as Pakistan, CT scans are often not ordered initially to avoid an excessive financial burden on the patient's family. About 64 % of Pakistan's population lives in rural areas, here CT scan facilities are not widely available and long waiting times for such scans can pose additional problems. This leads to misdiagnoses and mismanagement of atypical cases such as this one. Antenatal ultrasounds can also help detect lung changes in utero which can be tracked postnatally and treated accordingly<sup>11</sup>. But the role of such investigations in early detection of the disease, especially in district and rural areas, is not emphasized enough. Increased awareness and guidelines for physicians in these areas should be created and promoted.

### Conclusion

In summary, CLE remains commonly misdiagnosed due to lack of suspicion on clinical grounds and inadequate investigational measures because of a lack of funds and infrastructure in the health care systems, especially in rural areas, leading to mismanagement of patients. Prompt surgical intervention can be lifesaving. As presented in the case above, lack of access to appropriate infrastructure has devastating consequences, as this patient expired before appropriate management could be employed. Further steps should be taken to help overcome the diagnostic challenge and improve patient management in such cases.

**Human subjects:** Consent was obtained from the infant's parents for publishing the case.

**Disclaimer:** None to declare.

**Conflict of Interest:** None to declare.

**Financial relationships** All authors have declared that

they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work.

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### Authors' Contributions

**ZA, SS:** Drafting the work, interpretation, revising it critically.

**SQ:** Final approval, editing, revising it critically.

**SM:** Data acquisition.