

CASE REPORT

Congenital Lobar Emphysema: A Case Report of Rare Cause of Respiratory Distress in Neonate

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Abstract:

Congenital lobar emphysema is one of the many causes of the respiratory distress syndrome in infants, and its early recognition is important because proper surgical therapy can be lifesaving. Congenital lobar emphysema (CLE) is a rare condition characterized by over inflation of one or more lobes of the lung, resulting in compression of adjacent lung tissue and impaired respiratory function. We report the case of a one-month-old female infant, Faiza, who presented with progressive respiratory distress and feeding difficulties since the tenth day of life. Despite multiple hospital visits and empirical antibiotic therapy, her symptoms persisted. Upon admission to National Institute of Diseases of the Chest and Hospital (NIDCH), clinical evaluation revealed dyspnea, subcostal retractions, and decreased breath sounds over the left hemithorax. Radiological imaging studies confirmed the diagnosis of congenital lobar emphysema and underwent left upper lobectomy with successful outcomes. This case highlights the importance of prompt recognition and considering CLE in the differential diagnosis of persistent respiratory distress in early infancy with appropriate management of this condition to prevent complications and ensure favorable outcomes.

Keywords: Congenital lobar Emphysema, Neonate, surgical treatment, good prognosis and outcome.

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Introduction

Congenital lobar emphysema (CLE) is a rare congenital pulmonary anomaly characterized by overexpansion of one or more lobes of the histologically normal lung, without alveolar wall destruction. This condition results in compression of the adjacent lung parenchyma and impaired

ventilation, often manifesting as recurrent respiratory distress. There is air trapping in the lung during the expiratory phase of respiration due to deficient bronchial cartilage, which causes repeated episodes of respiratory distress. The affected lobe is essentially non-functional because of over distention and air trapping. It is frequently

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recognized in newborns; however, a few cases do not get evident until adulthood. This disease is potentially reversible if diagnosed and treated on time.¹⁵ CLE poses a significant diagnostic and therapeutic challenge due to its variable presentation and potential overlap with other pulmonary conditions such as pneumonia or pneumothorax. If diagnosed early, CLE is potentially reversible, with surgical resection of the affected lobe often resulting in excellent outcomes.

Congenital lobar emphysema (CLE) is a congenital anomaly of the lung, with a prevalence of 1 in 20,000 to 1 in 30,000¹. Most of the cases present in the neonatal period, with a male to female ratio of 3:1^{2,3}. CLE has also been reported with other associated anomalies with double superior vena cava and horse shoe kidney⁴. One case is reported with polysplenia, a syndrome characterized by bilateral bilobed lungs and bilateral pulmonary atria along with liver, which is symmetrically placed in the midline and multiple nodules of spleen⁵.

Case report:

Faiza, a 1-month-old female infant, second issue of non-consanguineous parents, presented with complaints of respiratory distress and feeding difficulty since 10 days of age. She was delivered at term by lower uterine cesarean section (LUCS) without any perinatal complications. The mother reported noticing progressive respiratory difficulty, particularly during feeding episodes. Despite receiving empirical antibiotic treatment at multiple local healthcare facilities, her symptoms failed to resolve, prompting referral to our tertiary care center. On admission to the National Institute of Diseases of the Chest and Hospital (NIDCH), Faiza appeared ill, lethargic, and dyspneic, with marked subcostal retractions and oxygen saturation was 88-89% at room air. Physical examination revealed reduced chest wall movement on the left side, tracheal deviation to the right, hyper resonance on percussion and markedly diminished breath sounds over the left hemithorax. A frontal radiograph of the chest shows marked over distention of the left upper lobe with mediastinal shift to the right. A subsequent computed tomography (CT) scan of chest confirmed hyper-aeration of the left upper lobe with associated

mediastinal displacement. Routine hematological and biochemical investigations were within normal limits. Further evaluation led to the diagnosis of left-sided congenital or infantile lobar emphysema.

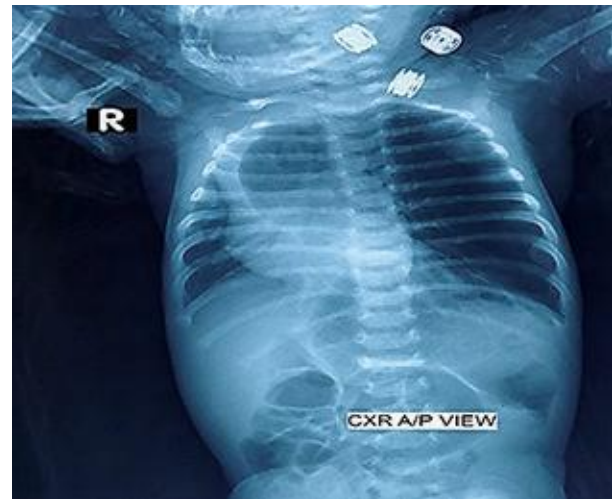


Figure 1: CXR

Management and Outcome:

The patient underwent a left upper lobectomy under general anesthesia with one-lung ventilation. Intraoperative findings revealed an emphysematous and markedly over expanded left upper lobe, while the adjacent left lower lobe

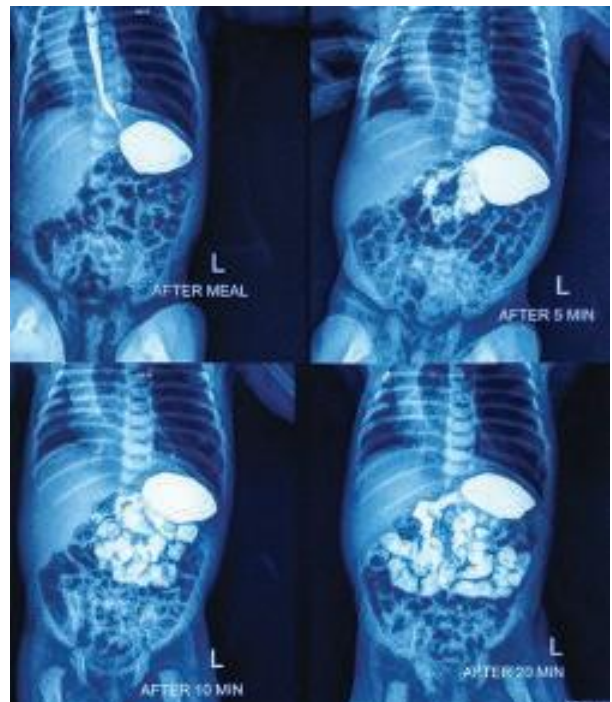


Figure 2: Contrast follow through

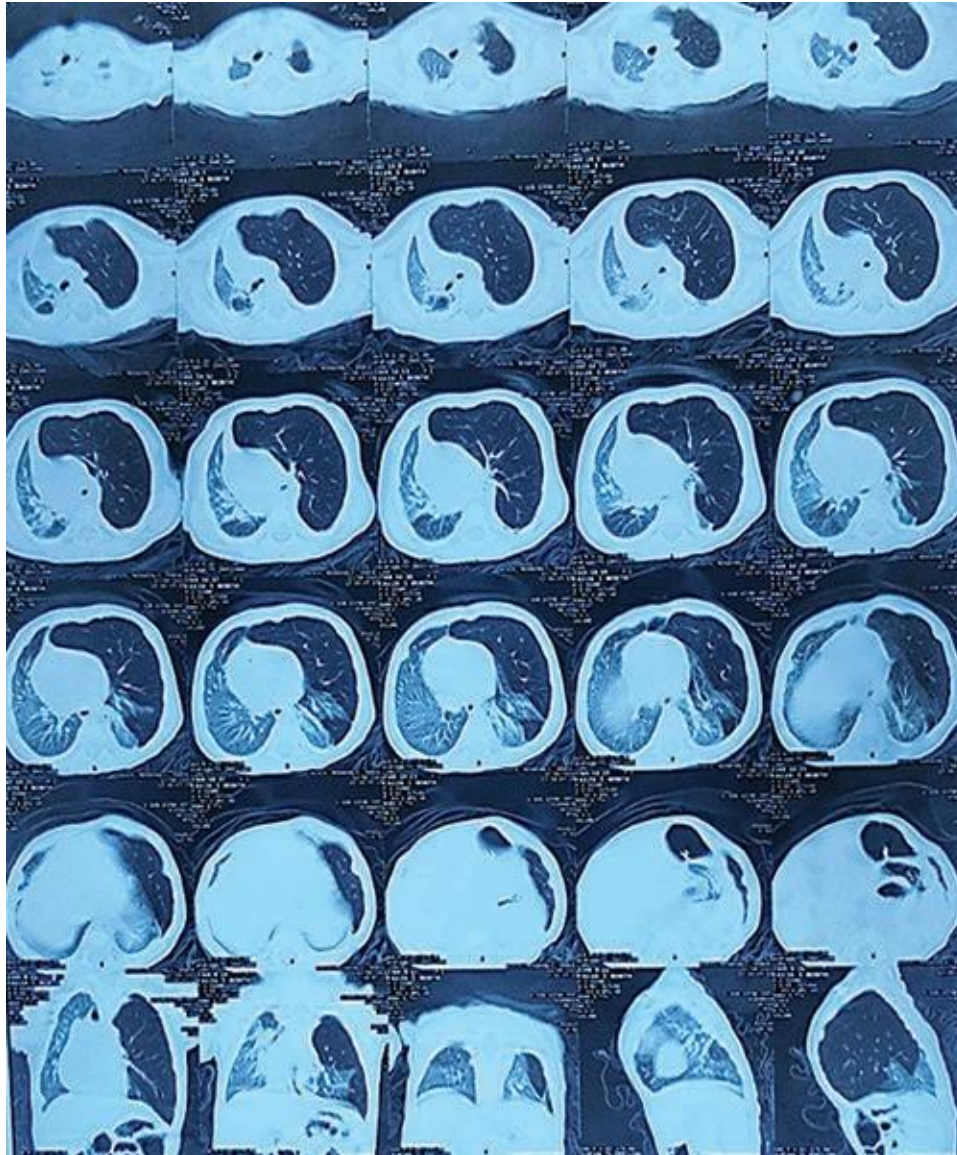


Figure 3: CT scan of chest

appeared partially collapsed due to compressive effects. The resection proceeded without complications.

Postoperatively, the patient experienced an uneventful recovery. Clinical and radiological assessments confirmed satisfactory re-expansion of the remaining lung parenchyma. The intercostal chest drain was removed on the third postoperative day. Faiza demonstrated significant clinical improvement, including resolution of respiratory distress and feeding difficulties, and was discharged in stable condition with scheduled follow-up.

Per operative pictorial



Figure 4: Emphysematous left upper lobe



Figure 5: After resection of Left upper lobe



Figure 6: specimen after resection



Figure 7: post-operative Chest X-ray



Figure 8: Post-operative status during discharge

Discussion:

Congenital lobar emphysema (CLE) is an uncommon congenital malformation, characterized by alveolar distension and pulmonary hyperinflation, with contralateral pulmonary atelectasis³. The clinical diagnosis of CLE can be challenging and chest radiography is often diagnostic, which typically shows hyperlucency of the affected lobe, widened intercostal spaces, diaphragmatic flattening, and mediastinal shift away from the affected side. In some cases, the mediastinum may herniate into the contralateral hemithorax.

Although most cases present in the neonatal period, approximately 5% are diagnosed by six months of age, and some are detected even later in childhood. Prenatal ultrasonography may identify CLE in utero, and familial cases have also been reported^{6,7,8}. The incidence of left upper lobe involvement is 43% of all cases, our infant belonging to this group; the middle right lobe is affected in 35% of cases, the upper right lobe in 21% of cases, while bilateral involvement occurs in 20% of cases⁹. Pneumonia & pneumothorax is a common misdiagnosis for CLE.²¹ It has some synonyms: infantile lobar emphysema, Congenital lobar overinflation, Congenital lobar or segmental bronchomalacia, Emphysema of infancy or childhood. Two histopathological forms of CLE exist: the hypoalveolar type (fewer than expected alveoli) and the polyalveolar type (greater than expected alveoli).

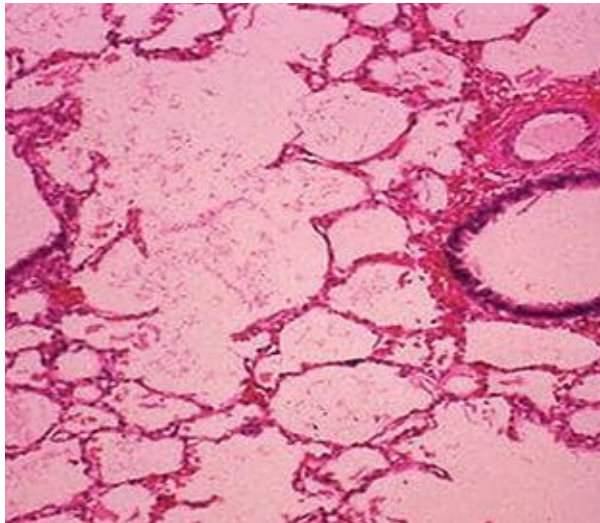


Figure 9: *Histopathology of congenital lobar emphysema with marked over distention of all alveoli.*

Pathophysiology

The fundamental mechanism involves defective bronchial cartilage, resulting in airway collapse during expiration and subsequent air trapping. This leads to progressive over inflation of the affected lobe, compression of adjacent lung tissue, and impaired pulmonary function.

Etiology

The etiology of CLE remains idiopathic in approximately 50% of cases. Identifiable causes include bronchial cartilage defects (25%), extrinsic vascular compression, bronchial stenosis, mucous plugging, abnormal bronchial secretions and mediastinal displacement¹⁰. In 10% of cases, congenital lobar emphysema can be associated with congenital cardiac malformations, which are excluded by echocardiography¹¹.

Clinical presentation

Typically, infants with CLE appear normal at birth. 50% of cases, Symptoms typically manifest within the first few days of life which includes- dyspnea, cyanosis, chest retractions, wheezing, and feeding difficulties. Physical examination may reveal asymmetric chest expansion, hyper-resonance, diminished breath sounds, and tracheal deviation. Progressive hypercapnia and respiratory failure may ensue if untreated.

Differential Diagnosis

1. Congenital cystic adenomatoid malformation.
2. Post pneumonic pneumatocele
3. Tension Pneumothorax
4. Congenital diaphragmatic hernia

Investigations / Diagnostic workup

1. Chest radiography- Chest X-ray is often the first diagnostic modality, revealing hyperinflation of the affected lobe, contralateral mediastinal shift, and adjacent lung atelectasis. Lateral and decubitus views help delineate non-collapsing emphysematous lobes.¹⁹
2. CT scan of chest- CT scan provides detailed anatomical and vascular information. It typically demonstrates a hyperlucent, hyper expanded lobe with preserved yet attenuated vascular markings, along with mediastinal displacement and compression of uninvolved lobes.²⁰
3. MRI of chest- Used selectively to evaluate vascular supply. CT/MRI, allows diagnosis and initiation of early treatment for a favorable prognosis ¹¹⁻¹⁴.
4. Prenatal USG- Can detect CLE as hyperechoic lung segments with normal vascular flow.
5. Bronchoscopy- Assesses bronchial anatomy and to remove the mucous plug or aspirated foreign body which should be performed at the same anaesthetic period for thoracotomy.
6. Ventilation-Perfusion Scanning: - Nuclear imaging shows delayed alveolar emptying and diminished perfusion of the affected lobe due to vascular compression.

Concomitant malformations

CLE is frequently associated with congenital cardiac anomalies (14–20%). Common abnormalities include Patent ductus arteriosus (PDA), Atrial septal defect (ASD), Ventricular septal defect (VSD), Total pulmonary venous return anomaly (TAPVR), and Tetralogy of Fallot (TOF). Embryologic overlap in the development of the heart and lungs during weeks 4–6 of gestation may account for this association. Therefore, disruptions during this critical window may result in simultaneous cardiopulmonary anomalies. A

thorough cardiovascular evaluation is warranted before surgical intervention.

Concomitant malformations accompanying congenital lobar emphysema*

Cardiac malformations 14-20%	Patent ductus arteriosus Atrial septal defect Ventricular septal defect Tetralogy of Fallot Pulmonary stenosis Pulmonary valve atresia Aortic coarctation Pulmonary hypertension Left aortic arch Right descending aorta Left ligamentum arteriosum Double superior Vena cava
Renal anomalies	Aplastic kidney Horseshoe kidney
Musculoskeletal anomalies	Pectus excavatum Hiatal hernia Diaphragmatic hernia
Gastrointestinal tract	Omphalocele Pyloric stenosis
Others	Cleft palate Chondroectodermal dysplasia Chondrodystrophy

Figure 10: Concomitant malformations

Treatment

Surgical lobectomy is the treatment of choice in symptomatic infants and usually results in complete recovery. Special attention must be given during anaesthetic induction to avoid rapid over-inflation of the affected lobe. Segmental resection, as an alternative to lobectomy, has been employed in select cases with localized disease. [14] Conservative management may be considered in asymptomatic or mildly symptomatic cases, particularly in older children with stable clinical status and normal bronchoscopic findings

Learning Points

- Early-Onset Respiratory Distress: CLE should be considered in the differential diagnosis of neonates and infants with unexplained or persistent respiratory distress, particularly when unresponsive to antibiotics or conventional therapy.
- Radiological Clues Are Key: Chest X-ray and CT scan are essential for diagnosis. Findings such as lobar hyperinflation, mediastinal shift, and compression of adjacent lung structures are characteristic of CLE.

- Mimics Other Conditions: CLE is frequently misdiagnosed as pneumonia, pneumothorax, or congenital cystic lung lesions, underscoring the importance of a high index of suspicion.
- Cardiac Evaluation is Crucial: Up to 20% of CLE cases are associated with congenital heart defects; echocardiographic assessment should be performed routinely before surgical planning.
- Definitive Management is Surgical: Lobectomy of the affected lobe is curative in symptomatic patients and typically results in excellent long-term respiratory outcomes.
- Conservative Approach Possible: Asymptomatic or mildly symptomatic patients may be managed conservatively with close monitoring, especially in older children.

Take-Home Message

Congenital lobar emphysema (CLE), though rare, should be considered in the differential diagnosis of neonates presenting with persistent respiratory distress, especially in cases unresponsive to conservative management. Congenital lobar emphysema is a potentially life-threatening condition characterized by over inflation of one or more lobes of the lung. Early recognition, accurate diagnosis, and prompt surgical intervention are pivotal in preventing complications and ensuring favorable clinical outcomes.

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Ethical Approval

The study is exempt from ethical approval in our institution.

Consent

Written informed consent was obtained from the patient or the publication of this case report and the accompanying images.

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