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Congenital Lobar Emphysema Mimicking Pneumothorax in a 6-Month-Old Infant With Acute Respiratory Distress: A Case Report.

Introduction

2 Congenital lobar emphysema (CLE), increasingly referred to as congenital lobar overinflation, is a rare congenital lung malformation characterized by abnormal hyperinflation of a pulmonary lobe. Despite the term emphysema, the condition is not primarily a destructive alveolar disease. Instead, the affected lobe becomes progressively overdistended, compressing adjacent normal lung parenchyma and potentially causing mediastinal shift, respiratory distress, and impaired ventilation [1,2].

The mechanism is most often related to a ball-valve effect caused by intrinsic bronchial abnormality, such as deficient bronchial cartilage, bronchomalacia, mucosal folds, or bronchial stenosis. Extrinsic bronchial compression by vascular structures or mediastinal lesions may also occur, although in many patients no precise cause is identified [1-3]. CLE usually presents in the first months of life with tachypnea, retractions, wheezing, cyanosis, poor feeding, or recurrent respiratory infections. Severe cases can present as acute respiratory emergencies [1,4].

The diagnosis can be challenging because the radiographic appearance may resemble pneumothorax. Both conditions may present with unilateral hyperlucency and contralateral mediastinal displacement. However, treatment is different: pneumothorax may require urgent pleural drainage, whereas chest tube insertion is not therapeutic in CLE and may delay appropriate treatment [5,6]. Computed tomography (CT) is essential when the diagnosis is uncertain because it confirms the intrapulmonary nature of the hyperinflation, identifies the affected lobe, excludes pleural air, and assesses associated lung disease [2,7].

We report a case of congenital lobar emphysema in a 6-month-old infant presenting with acute respiratory distress, in whom the initial chest radiograph suggested pneumothorax, while CT confirmed CLE and demonstrated associated contralateral pulmonary involvement.

Case Presentation

A 6-month-old male infant was admitted to the emergency department for acute respiratory distress. He had previously required hospitalization in the pediatric intensive care unit during early infancy for severe respiratory distress in the context of recurrent lower respiratory tract infection. There was no known history of thoracic trauma, foreign body aspiration, congenital heart disease, or previous thoracic surgery.

The current episode began with cough, fever, progressive tachypnea, and feeding difficulty, followed by worsening respiratory effort. On admission, the infant was conscious but irritable and dyspneic. He had marked 3 intercostal and subcostal retractions, nasal flaring, and oxygen saturation of 86% on room air, improving to 95% under supplemental oxygen. Body temperature was 38.3 degrees C, heart rate was 164 beats/min, and respiratory rate was 62 breaths/min. Chest examination revealed decreased breath sounds over the left hemithorax with scattered crackles 1 on the right side.

Initial laboratory tests showed inflammatory features, with leukocytosis at 18,200/mm3 and C-reactive protein at 64 mg/L. Blood gas analysis showed mild respiratory acidosis with hypoxemia before oxygen supplementation. Because of the acute presentation and asymmetric chest findings, an urgent chest radiograph was obtained.

Chest radiography demonstrated marked hyperlucency and overexpansion of the left hemithorax, flattening of the left hemidiaphragm, compression of adjacent lung parenchyma, and mediastinal shift toward the right side. Pneumothorax was initially suspected because of the hyperlucent hemithorax and mass effect. However, careful review showed persistence of fine vascular markings within the hyperlucent area and no clear pleural line. In view of this diagnostic uncertainty, thoracic CT was performed before any pleural drainage.

Thoracic CT excluded pneumothorax and showed major overinflation of the left upper lobe, with stretching of bronchovascular structures, compression atelectasis of the remaining left lung, and rightward mediastinal displacement. These findings were consistent with congenital lobar emphysema of the left upper lobe. CT also demonstrated contralateral

pulmonary abnormalities, including patchy right basal consolidation and peribronchial thickening, suggesting associated bronchopneumonia. No endobronchial foreign body, pleural air collection, diaphragmatic hernia, or obvious mediastinal mass was identified. Echocardiography did not show significant congenital heart disease.

The infant was admitted for close monitoring and stabilization. Initial treatment included humidified oxygen therapy, careful hydration, antipyretics, respiratory physiotherapy when tolerated, and avoidance of unnecessary pleural drainage. Because of fever, inflammatory markers, and contralateral parenchymal abnormalities on CT, empirical intravenous antibiotic therapy with cefotaxime and gentamicin was started, then continued according to clinical response. Respiratory distress improved partially after supportive and antibiotic treatment, but persistent left upper lobe hyperinflation and mediastinal mass effect remained evident.

After multidisciplinary discussion involving pediatrics, pediatric intensive care, radiology, anesthesia, and pediatric surgery, operative management was indicated because of symptomatic CLE with significant mass effect. The infant underwent left posterolateral thoracotomy. Intraoperatively, the left upper lobe was markedly overinflated and compressed the remaining ipsilateral lung. There was no pleural air leak or mechanical obstruction. Left upper lobectomy was performed. The remaining lung re-expanded progressively after resection.

The postoperative course was favorable. The infant was monitored [1] in the pediatric intensive care unit during the early postoperative period and was extubated without difficulty. Oxygen therapy was progressively weaned. Follow-up chest radiography showed satisfactory re-expansion of the remaining left lung and improvement of mediastinal position. Histopathological examination confirmed features compatible with congenital lobar emphysema, including overdistended alveolar spaces without malignant lesion. [1] The infant was discharged in good clinical condition after 9 postoperative days. At 1-month follow-up, he had no respiratory distress, good feeding tolerance, and no clinical evidence of recurrence.

Discussion

This case illustrates a major diagnostic pitfall in pediatric respiratory emergencies: congenital lobar emphysema can closely mimic pneumothorax on initial chest radiography. In both conditions, the radiograph may show a hyperlucent hemithorax, mediastinal displacement, and apparent compression of the opposite lung. In an unstable infant, this similarity can lead to urgent chest tube insertion. However, pleural drainage does not treat CLE and may cause complications or delay definitive management [5,6].

Several radiographic signs may suggest CLE rather than pneumothorax. In CLE, the hyperlucency is intrapulmonary and vascular markings may still be visible within the affected area, although they are stretched and attenuated. A definite pleural line is absent. The affected lobe is overexpanded and may cause compression of adjacent lung, diaphragmatic depression, and mediastinal shift. In pneumothorax, peripheral vascular markings are absent beyond the pleural line. These distinctions are important but may be subtle on emergency radiographs, particularly in infants with severe respiratory distress [2,7].

CT was decisive in our patient. It confirmed that the hyperlucency represented lobar overinflation rather than pleural air, identified the left upper lobe as the affected lobe, and demonstrated compression of adjacent lung. It also showed contralateral parenchymal disease, which was clinically relevant because the infant had fever and inflammatory markers. The contralateral inflammatory changes justified antibiotic therapy and careful stabilization before surgery. CT also helps exclude other differential diagnoses, including congenital pulmonary airway malformation, pneumatocele, bronchogenic cyst, diaphragmatic hernia, Swyer-James syndrome, and foreign body obstruction [1,2,7].

The left upper lobe is the most commonly involved lobe in CLE, followed by the right middle lobe and right upper lobe. Bilateral or multilobar disease is less frequent but has been reported and may complicate management [2,3,8]. Associated congenital heart disease has also been described, so echocardiographic evaluation is often useful before surgery, especially when the infant presents with severe symptoms or when anesthesia is

planned [1,3].

Treatment depends on clinical severity. Conservative management may be considered in selected mildly symptomatic or incidentally diagnosed cases under close follow-up [9]. However, severe or persistent respiratory distress, progressive hyperinflation, recurrent infection, or marked mediastinal shift usually favors surgery. Lobectomy of the affected lobe remains the standard treatment for symptomatic CLE and generally provides excellent outcomes, as observed in our patient [2-4,10].

The present case has several practical messages. First, the absence of a clear pleural line on chest radiography should prompt reconsideration of pneumothorax before chest tube insertion. Second, CT should be obtained when the infant is stable enough and the diagnosis is uncertain. Third, associated infection or contralateral lung disease may influence timing and perioperative management. Finally, multidisciplinary decision-making is essential because these patients may require intensive respiratory support, specialized anesthesia, and pediatric thoracic surgical expertise.

Conclusion

Congenital lobar emphysema is a rare but important cause of respiratory distress in infants. It may mimic pneumothorax on chest radiography, especially when unilateral hyperlucency and mediastinal shift are present. Thoracic CT is essential to confirm the diagnosis, avoid inappropriate pleural drainage, identify associated pulmonary lesions, and plan treatment. In symptomatic infants with significant mass effect, surgical lobectomy after stabilization is an effective treatment with favorable outcome.

Learning Points

• Congenital lobar emphysema should be considered in any infant with respiratory distress and unilateral hyperlucency on chest radiography.

• Persistence of vascular markings and absence of a clear pleural line favor lobar overinflation rather than pneumothorax.

• Thoracic CT is the key investigation when pneumothorax and congenital lobar emphysema are difficult to distinguish.

• Contralateral pulmonary infection or inflammatory disease may justify antibiotic therapy before definitive surgery.

• Symptomatic congenital lobar emphysema with significant mass effect is usually treated by lobectomy, with good postoperative outcomes.

Declarations

Ethical approval: This anonymized single-patient case report does not include experimental intervention. Local institutional requirements should be followed for case report publication.

Consent: Publication of any real patient case and clinical images requires written informed consent from the parent or legal guardian before journal submission.

Conflicts of interest: The author declares no conflicts of interest.

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Data availability: The clinical data supporting this case report are available from the treating team and are not publicly shared in order to preserve patient confidentiality.

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