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Bilateral Giant Congenital Lobar Emphysema in an Infant: A Case Report

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Abstract **Case Report**

Congenital lobar emphysema (CLE) is a rare malformation of the bronchopulmonary tree characterized by progressive hyperinflation of one or more pulmonary lobes due to airway obstruction. Bilateral involvement is exceptional and typically presents with severe respiratory distress in early infancy. We report the case of a 45-day-old infant presenting with respiratory distress. Clinical symptoms began in the first days of life with tachypnea, cyanosis, and feeding difficulties. The patient was initially treated as having viral bronchiolitis. Due to the lack of clinical improvement, a chest radiograph was performed, revealing a right apical opacity associated with marked hyperlucency and hyperexpansion of the left lung, widening of the intercostal spaces, flattening of the left hemidiaphragm, and anterior mediastinal herniation. Chest computed tomography (CT) demonstrated consolidation of the apical segment of the right upper lobe, bilateral ground-glass opacities involving the lower lobes and the lingula, and marked hyperlucency of the left upper and middle lobes with vascular attenuation. These findings were highly suggestive of bilateral giant congenital lobar emphysema with superinfection. Surgical lobectomy was performed, and histopathological examination confirmed the diagnosis.

Keywords: Congenital lobar emphysema; Bilateral giant CLE; Infant; Respiratory distress; Chest CT; Lobar hyperinflation; Bronchial obstruction; Pulmonary imaging.

INTRODUCTION

Congenital lobar emphysema (CLE) is a rare pulmonary malformation resulting from partial bronchial obstruction, leading to air trapping and progressive lobar overdistension. It typically affects a single pulmonary lobe, most commonly the left upper lobe, while bilateral involvement remains extremely rare.

The disease usually becomes symptomatic within the first days of life, presenting with respiratory distress, tachypnea, and cyanosis. Imaging plays a central role in establishing the diagnosis.

CASE REPORT

A 1-month-and-15-day-old female infant was admitted for febrile respiratory distress. Clinical history revealed tachypnea, cyanosis, and feeding difficulties since birth. The patient had initially been treated for bronchiolitis.

Chest radiography showed a right apical opacity associated with hyperexpansion of the left lung, widening of intercostal spaces, and mediastinal herniation.

A chest CT scan was performed for further evaluation, revealing consolidation of the right upper lobe, bilateral ground-glass opacities, marked hyperlucency with hyperexpansion of the left upper and middle lobes, and vascular attenuation.

The patient underwent lobectomy, and histopathological examination confirmed the diagnosis of congenital lobar emphysema.

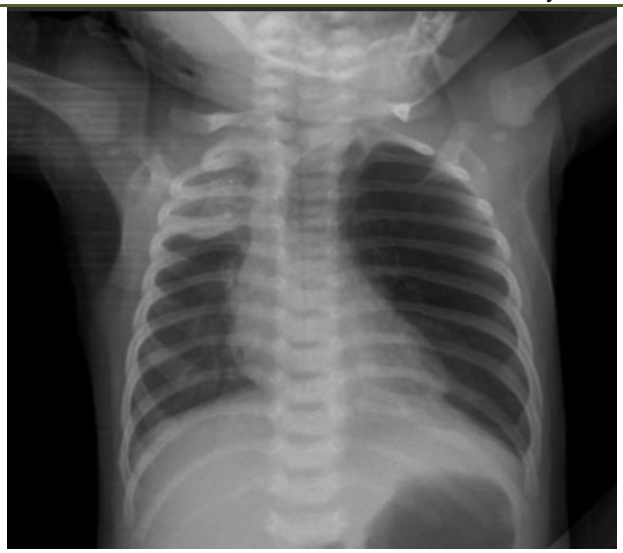

Figure 1 : Chest radiograph showing right apical opacity and left lung hyperexpansion with mediastinal shift.

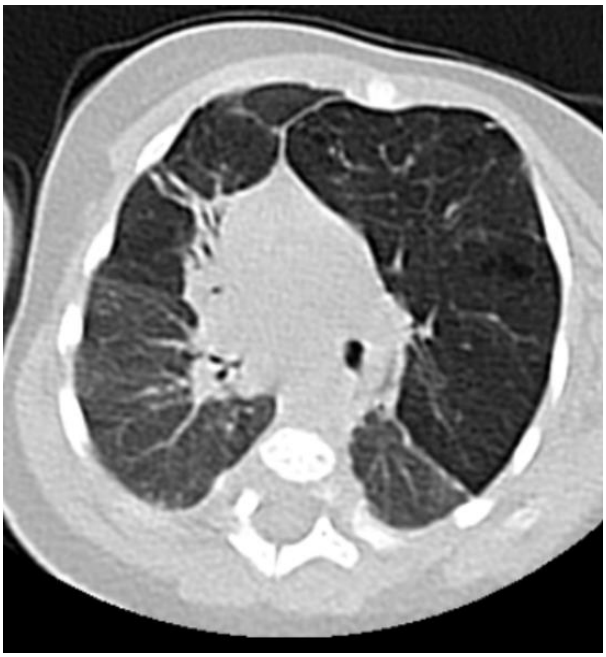

Figure 2 :CT scan showing hyperlucency of the left upper and middle lobes with vascular attenuation.

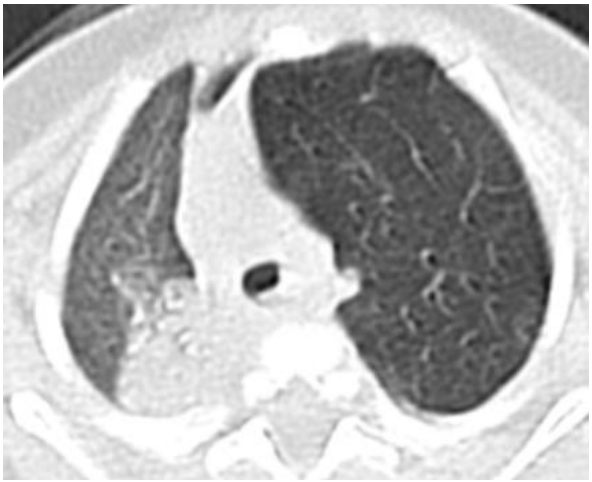

Figure 3: CT scan showing right upper lobe consolidation.

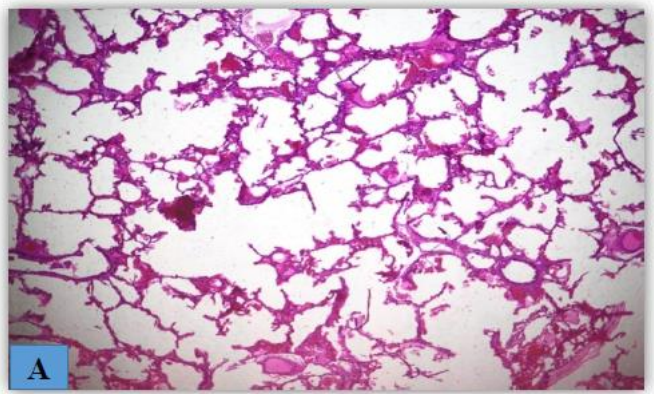

Figure 4: Histopathological confirmation of congenital lobar emphysema.

DISCUSSION

Congenital lobar emphysema (CLE) is a rare developmental lung disorder with an incidence ranging from 1 in 20,000 to 1 in 30,000 live births [1]. It most commonly involves a single lobe, particularly the left upper lobe, while bilateral involvement is extremely rare, accounting for less than 12% of reported cases worldwide [1,2].

This rarity requires increased diagnostic vigilance, as bilateral presentations often mimic more common neonatal conditions such as viral bronchiolitis, leading to delayed diagnosis and management.

The pathophysiology involves a partial bronchial obstruction acting as a "ball-valve" mechanism. Air enters during inspiration but becomes trapped during expiration due to airway collapse, resulting in progressive alveolar distension and lobar hyperinflation. Bilateral involvement may mimic tension pneumothorax [2,3].

Computed tomography is essential for diagnosis, showing hyperlucent lobes with vascular attenuation. In our case, CT also revealed consolidation and ground-glass opacities, suggesting superinfection.

The main differential diagnoses include CPAM and bronchial atresia. Early surgical management ensures an excellent prognosis.

CONCLUSION

Bilateral congenital lobar emphysema is an exceptionally rare but potentially life-threatening condition that can mimic more common neonatal respiratory diseases. This case highlights the crucial role of imaging, particularly CT, in achieving an accurate diagnosis. Early surgical intervention leads to rapid clinical improvement and excellent outcomes, even in severe bilateral form.

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