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# Giant Congenital Lobar Emphysema: A Rare Cause of Persistent Respiratory Distress in Neonatal Intensive Care

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**Abstract** **Case Report**

Congenital lobar emphysema (CLE) is a rare congenital anomaly of the lung causing excessive aeration of one or more lobes of a histologically normal lung. It manifests in the first weeks of life and rarely in the neonatal period, with respiratory distress due to compression atelectasis and often associated with mediastinal displacement. CLE poses a challenge in terms of diagnosis and positive pressure ventilation due to air trapping. We report the case of a newborn presenting with persistent respiratory distress. A thoracic CT scan was performed, confirming the diagnosis of CLE. The definitive diagnosis is provided by histology of the operative specimen, revealing emphysematous lesions. Treatment involves lobectomy after stabilization of respiratory function. Surgical abstention is possible for asymptomatic or paucisymptomatic lesions.

**Keywords:** Congenital lobar emphysema (CLE), lung, Neonatal Intensive Care.



## INTRODUCTION

Bronchopulmonary malformations (BPM) result from developmental accidents in the bronchopulmonary system. During the third week of gestation, the development of the respiratory system begins and aberrations in this developmental stage may cause parenchymal lung malformations. They are rare and polymorphic conditions among which congenital giant lobar emphysema (CGLE) represents 3 to 15%. Congenital giant lobar emphysema (CGLE) is a pulmonary developmental anomaly resulting in hyperinflation of one or more pulmonary lobes, most often due to bronchial stenoses. It is a rare cause of neonatal respiratory distress. The diagnosis can be prenatal, but remains difficult due to clinical similarities with other pulmonary malformations.

X-ray and thoracic computed tomography confirm the diagnosis postnatally [1,2]. We report a 1-day-old term, male baby who was hospitalized in our neonatal care intensive,hospital mohamed 6 marrakesh Morrocco for persistent respiratory distress. on detailed clinical examination at admission, a possibility of congenital lobar emphysema (CLe) was considered. a Ct chest was performed, and diagnosis of CLe was confirmed.

## CASE REPORT

The newborn is a male infant born to a 19-year-old primiparous mother via vaginal delivery after an unmonitored pregnancy with a negative medical history. The newborn had Apgar scores of 8, 9, and 10 at 1, 5, and 10 minutes, respectively. On examination, the newborn appeared pale, minimally reactive, with limited movement, cyanosis of the extremities, heart rate of 144 bpm, respiratory rate of 50 breaths per minute, temperature of 36.4°C, capillary refill time less than 3 seconds, oxygen saturation of 96% on 1 liter of oxygen, and signs of respiratory distress including subcostal retractions, intercostal retractions, thoracoabdominal rocking, xiphoid funnel, and bilateral crepitations. The newborn also exhibited weak sucking reflex. Chest X-ray revealed a distended right lung with overall hyperlucency of the middle lobe, with visible vascularity (Figure 1).

Initially, the patient was placed on oxygen goggles and treated for maternal-fetal infection with pulmonary involvement. However, due to the lack of clinical improvement in the newborn, the patient was switched to continuous positive airway pressure (CPAP) and broad-spectrum antibiotics.

On day 4, a second chest X-ray revealed uniform hyperlucency with distension of the right hemithorax (Figure 2). Given this radiological appearance, giant congenital lobar emphysema, diaphragmatic hernia, dextrocardia, and pneumonia are to be considered.

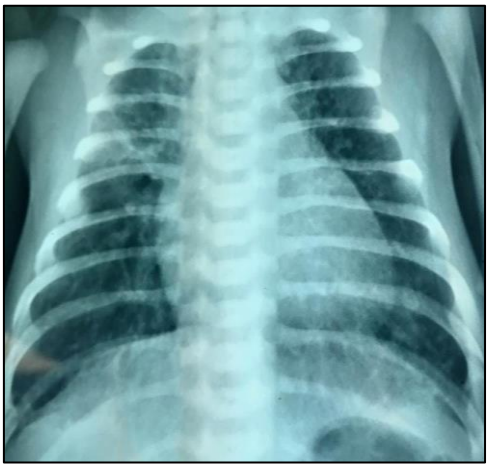

**Fig 1: Chest X-ray taken on day 1**

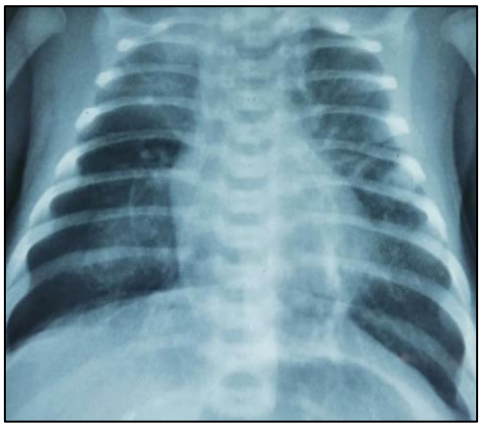

**Fig 2: Second chest X-ray performed showing Overinflation and atelectasis of right upper lobe**

At this stage, a thoracic computed tomography (CT) scan was necessary, revealing moderate parenchymal lobar hyperinflation leading to atelectasis of the right upper lobe, indicative of congenital lobar emphysema (Figure 3).

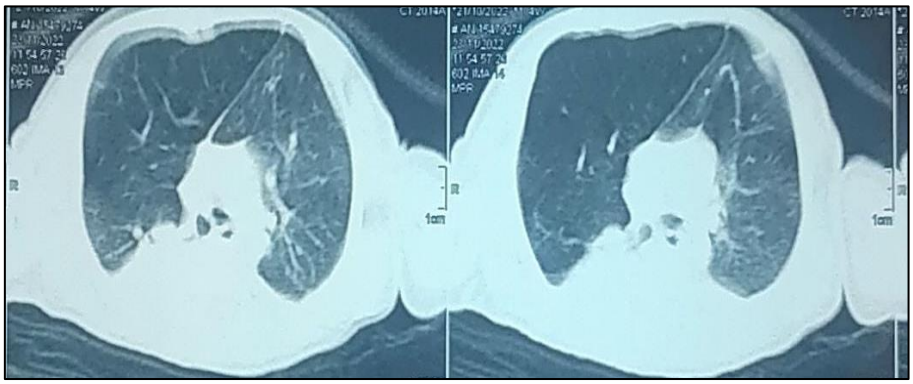

**Figure 3: Thoracic computed tomography (CT) scan showing midle lobar parenchymal hyperinflation responsible for atelectasis of the upper right lobe suggesting congenital lobar emphysema**

As part of the investigation for associated cardiopathy, an echocardiogram was performed, revealing left ventricular hypertrophy. Bronchoscopic examination and lung scintigraphy were not conducted. The newborn was placed on continuous positive airway pressure (CPAP) and required an FiO2 of 0.3 to maintain a normal oxygen saturation of 90% to 95%. Following investigation, sepsis screening was negative, and blood

culture yielded no growth. An indication for right upper lobectomy was proposed.

## DISCUSSION

Congenital lobar emphysema (CLE) is a rare congenital lung malformation with a prevalence of 1 in 20,000 to 1 in 30,000. Air trapping in the lungs during the expiratory phase of respiration due to deficient cartilage leads to excessive distension and hyperaeration of the affected pulmonary lobe, resulting from a check valve mechanism, leading to atelectasis of the remaining contralateral lung and cardiomedastinal displacement [2,3]. CLE manifests early in the neonatal period up to 6 months of age, with clinical manifestations appearing at birth in 33% of cases and before one month of age in 50% of cases. This condition occurs more frequently in boys. There is a prevalence of 43% of cases involving the left upper lobe, 32% of the right middle lobe, 20% of the right upper lobe, and 20% of bilateral involvement.

The disease may be caused by bronchial cartilage dysfunction (reduced or absent bronchial cartilage leading to intrinsic bronchial narrowing and bronchomalacia) or external bronchial compression due to various causes, resulting in air trapping. CLE can be diagnosed as hypoalveolar (with a reduced number of alveoli) or polyalveolar (with a higher number of alveoli) through histopathological examination [3].

The diagnosis of CLE may be established on prenatal ultrasound, but it is often overlooked and detected in infants when progressive hyperinflation leads to symptoms of compression of the remaining ipsilateral lung as well as the contralateral lung, resulting in cardiorespiratory compression. Less severe cases of CLE may manifest later in infancy or childhood. 10% of patients present with associated anomalies such as congenital heart disease. Typically, patients present with rapid respiratory rate, tachycardia, and thoracic retractions, which deteriorate progressively with gas accumulation in the affected lobe, leading to respiratory difficulty and insufficiency. Asymmetric chest wall dilation, rhonchi, displacement of apical impulse, markedly resonant percussion over the affected side, and reduced breath and heart sounds are observed. Chest X-ray reveals hyperinflation of the affected lobe, atelectasis of the contralateral lung, mediastinal shift, and flattening of the ipsilateral diaphragm. Several articles in the literature report diagnostic delays, with patients being managed for recurrent community-acquired pneumonia or recurrent pneumothorax [4,5].

For diagnosis, chest X-ray and thoracic CT scan can be performed. CT scan findings suggestive of CLE include herniation of the contralateral lobe, mediastinal shift to the opposite side, and atelectasis of a portion of the contralateral lung.

Despite their rarity, pulmonary malformations are a major cause of persistent respiratory problems in infants. Congenital lobar emphysema may mimic pneumothorax, underscoring the importance of definitive diagnosis to avoid unnecessary interventions such as needle aspiration. Postnatal chest X-ray and CT scan often differentiate between lobar emphysema and cystic pulmonary malformations. Definitive diagnosis is provided by histology of the operative specimen, which reveals emphysematous lesions. Treatment involves lobectomy after stabilization of respiratory function. Surgical abstention is possible for asymptomatic or paucisymptomatic lesions. However, due to the risk of recurrent bronchopulmonary infection, some authors recommend elective systematic lobectomy [6].

## CONCLUSION

Congenital giant lobar emphysema is a rare pulmonary malformation, characterized by a segmental, lobar, or multilobar pulmonary distension without parenchymal destruction, posing a life-threatening condition in newborns. Chest X-ray and CT scan are two essential tools for diagnosis.

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