Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Radiology

Giant Congenital Lobar Emphysema Presenting as Acute Dyspnea in a 2-Year-Old: A Case Report

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DOI: https://doi.org/10.36347/sjmcr.2025.v13i11.040 | **Received:** 15.09.2025 | **Accepted:** 03.11.2025 | **Published:** 20.11.2025

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Abstract **Case Report**

Giant lobar emphysema (GLE), also known as congenital lobar overinflation (CLO), is a rare pulmonary anomaly in infants and young children characterized by hyperinflation of one or more pulmonary lobes. It may present acutely or be discovered incidentally. We report a case of a 2-year-old child who presented with acute dyspnea and was incidentally found to have giant emphysema of the left upper lobe on imaging. Surgical lobectomy was performed with favorable outcome. Although rare, GLE should be considered in the differential diagnosis of dyspnea in young children. Early recognition and appropriate management, including surgery, can lead to excellent outcomes.

Keywords: Giant Lobar Emphysema (GLE), Congenital Lobar Emphysema (CLE), Dyspnea, Hyperinflation, Lobectomy, Mediastinal Shift.

INTRODUCTION

Giant lobar emphysema (GLE), also known as congenital lobar emphysema (CLE), is a rare pulmonary developmental anomaly characterized by overdistension of one or more anatomically normal lobes secondary to partial bronchial obstruction. This results in air trapping and compression of the adjacent pulmonary parenchyma. The condition most commonly manifests during the neonatal period or early infancy but may also be identified later in childhood, either through symptomatic presentation or as an incidental finding [1].

The etiology of GLE is multifactorial and may include intrinsic bronchial cartilage malformations, extrinsic compression by adjacent vascular structures, or idiopathic factors [2]. The left upper lobe is most frequently involved, followed by the right middle lobe [3].

This report presents a case of GLE incidentally detected in a 2-year-old child who presented with dyspnea, highlighting the critical role of imaging in the evaluation of pediatric respiratory distress.

OBSERVATION

A 2-month-old male infant, the younger of two siblings born to non-consanguineous parents, was admitted to the pediatric emergency department with afebrile respiratory distress accompanied by intermittent cyanotic episodes. On clinical evaluation, the patient was conscious, tachycardic with a heart rate of 152 beats per minute, and tachypneic with a respiratory rate of 62 cycles per minute. Oxygen saturation was 90% while receiving 2 L/min of supplemental oxygen.

Respiratory examination revealed signs of increased work of breathing, including subcostal and intercostal retractions, as well as bilateral wheezing. Cardiovascular examination demonstrated rightward deviation of the cardiac sounds and the presence of a grade 2/6 systolic murmur. The remainder of the physical examination was within normal limits.

A chest X-ray revealed significant hyperinflation of the left upper lobe with mediastinal shift to the right and compression of the remaining lobes **[Fig. 1]** Transthoracic echocardiography showed no abnormalities.).

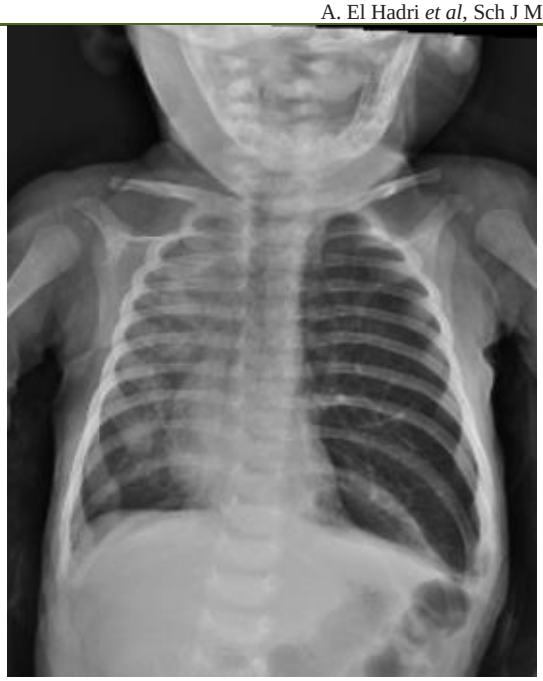

Figure 1: A chest X-ray revealed significant hyperinflation of the left upper lobe with mediastinal shift to the right A contrast-enhanced computed tomography (CT) scan of the chest confirmed giant emphysematous changes in the left upper lobe with attenuated vasculature and compression atelectasis of the adjacent lobes [Fig. 2]. No endobronchial foreign body or vascular anomaly was identified

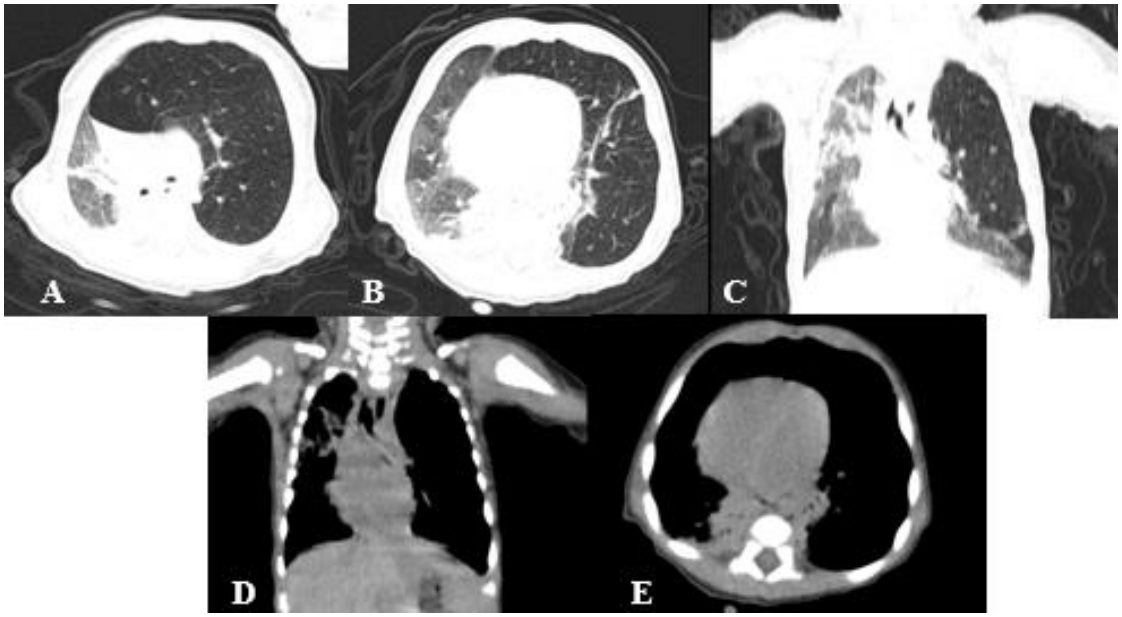

Figure 2: Thoracic CT scan parenchymal window: axial (A-B) and coronal section (C) and mediastinal window coronal (D) ans axial section (E), Giant emphysematous changes of the left upper lobe with vascular attenuation and compressive atelectasis of the adjacent lobe

A diagnosis of congenital giant lobar emphysema was established. The patient underwent a left upper lobectomy via thoracotomy. Intraoperative findings revealed a markedly hyperinflated left upper lobe that failed to deflate adequately, with preservation of normal architecture in the surrounding pulmonary parenchyma. Histopathological examination demonstrated alveolar overdistension without evidence of inflammation, fibrosis, or malignancy.

The postoperative course was uneventful. The patient was discharged on the seventh postoperative day and remained asymptomatic at a 6-month follow-up evaluation.

DISCUSSION

Congenital giant lobar emphysema (CGLE) is a rare but potentially life-threatening pulmonary anomaly of early childhood. Its incidence is estimated at

approximately 1 in 20,000 to 30,000 live births. The condition was first described by Nelson in 1932 and later characterized in greater detail by Robertson and James in 1951 [3,4]. CGLE demonstrates a male predominance, with one-third of cases presenting at birth, and nearly all diagnosed within the first six months of life [5–7]. Several etiological factors have been implicated in the pathogenesis of CGLE. In approximately 50% of cases, a deficiency in bronchial cartilage tissue has been identified, resulting in a "ball-valve" mechanism that leads to progressive air trapping and lobar overinflation.

Clinically, respiratory distress is the most common presenting feature. Other associated manifestations may include dyspnea, wheezing, grunting respirations, tachypnea, and progressive cyanosis [8].

The diagnosis of congenital giant lobar emphysema is primarily clinical and is supported by radiological imaging. Chest radiography and computed tomography (CT) typically reveal the characteristic appearance of a hyperinflated lobe causing compression of adjacent pulmonary lobes. Bronchoscopy may provide additional diagnostic value by identifying intrinsic airway anomalies or detecting extrinsic bronchial compression [6].

In the present case, the diagnosis was made incidentally during the evaluation of dyspnea, underscoring the importance of considering structural pulmonary anomalies in the differential diagnosis of respiratory symptoms in infants and young children.

Management strategies are guided by the severity of clinical presentation. Asymptomatic or mildly symptomatic patients may be managed conservatively with close monitoring. However, in cases associated with significant respiratory compromise, surgical intervention in the form of lobectomy remains the treatment of choice, as demonstrated in our patient [9].

Postoperative outcomes are generally favorable, with low morbidity rates and rapid recovery. Long-term prognosis following lobectomy is excellent, with most patients achieving complete resolution of symptoms and normal pulmonary function [10].

SUMMARY

Giant lobar emphysema represents a rare yet significant cause of respiratory distress in infants and young children. It should be considered in the differential diagnosis of dyspnea, particularly when imaging studies reveal characteristic features of lobar hyperinflation and compression of adjacent lung tissue. Early recognition and timely surgical intervention in symptomatic cases are crucial, as they can result in complete recovery and restoration of normal pulmonary function.

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