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# Congenital lobar emphysema in a newborn- A rare case report

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## Abstract

Congenital lobar emphysema (CLE) is a rare congenital malformation of lung which is characterized by over distension of a lobe of a lung due to developmental anomaly of the lower respiratory tract. CLE is a rare condition having a prevalence of 1 in 20,000 to 1 in 30,000 with male predominance of M:F ratio of 3:1. Although a rare lung disease, it can cause severe respiratory distress in the newborns and infants. Herein, we report a case of one month male child admitted in the pediatric emergency unit with respiratory distress since 5 days. Radiological investigation revealed hyperinflation of the left upper lobe with trachea-mediastinal shift towards right and a partial compression atelectasis of left lower lobe and right lung. Our histopathological findings confirmed the clinical diagnosis of congenital lobar emphysema with foci of interstitial pneumonia and collapse. The baby was treated with surgical management and the condition was improved. Congenital lobar emphysema often presents as a diagnostic and therapeutic dilemma but because it is a potentially reversible condition if diagnosed and treated on time, high index of suspicion in newborns presenting with respiratory distress should be made to avoid morbidity and mortality.

**Keywords:** Congenital lobar emphysema, Over inflation, Lung

## Introduction

Congenital lobar emphysema (CLE) is also known by many other synonyms like congenital alveolar over distension, congenital large hyper lucent lobe and congenital lobar over inflation. It is a rare congenital developmental lung malformation which is characterized by hyperinflation of one or more of the

pulmonary lobes occurring as a result of developmental anomaly of the lower respiratory tract [1,2]. During the third week of gestation, the development of the respiratory system begins and aberrations in this developmental stage may cause parenchymal lung malformations. [2,3]

CLE is a rare congenital malformation that has a prevalence rate of 1 in 20,000 to 1 in 30,000.[1] It shows male predominance with a males to females ratio of 3 to 1. The most commonly involved lobe is the left upper lobe (43%), followed by involvement of the right middle lobe (32%) and right upper lobe (21%). The rarest to involve are the lower lobes (2%)[4]. The affected lobe is essentially non-functional because of over distention and air trapping and causes respiratory distress.

Approximately 25% cases present at birth, 50% by one month of age and nearly all by six months of age. It is a potentially reversible condition if diagnosed and treated on time and therefore high index of suspicion in newborns presenting with respiratory distress should be made to avoid morbidity and mortality.

**Case Report**

A 35 days male child came to pediatric OPD with complaints of fast breathing since 5 days, cough and cold since 3 days along with history of lethargy and not accepting feed since 2 days. The child was a first born and the antenatal and birth history was uneventful. Other history was not significant.

**Physical Examination**

General examination was normal. There was no malnutrition. Respiratory examination showed moderate tachypnoea with symmetrical chest expansion. Apex beat was felt at left 4th intercoastal space at mid clavicular line. On percussion there was hyper-resonance in upper zone of left lung whereas this area had decreased breath sounds on auscultation. No adventitious sounds were heard. Other vitals and systemic examination was normal.

**Investigation**

All the routine blood investigations were within normal limit.

X-ray Chest (PA View) showed hyperlucency of left upper lobe with reduced lung marking and collapse of the ipsilateral remaining lung field. There was mediastinal shift to the right. (Fig.1)

CT Scan Thorax (Plain) revealed hyperinflation of the left upper lobe with trachea-mediastenal shift towards right and a partial compression atelectasis of left lower lobe and right lung. Major bronchus and segmental bronchus appeared normal. (Fig.2)

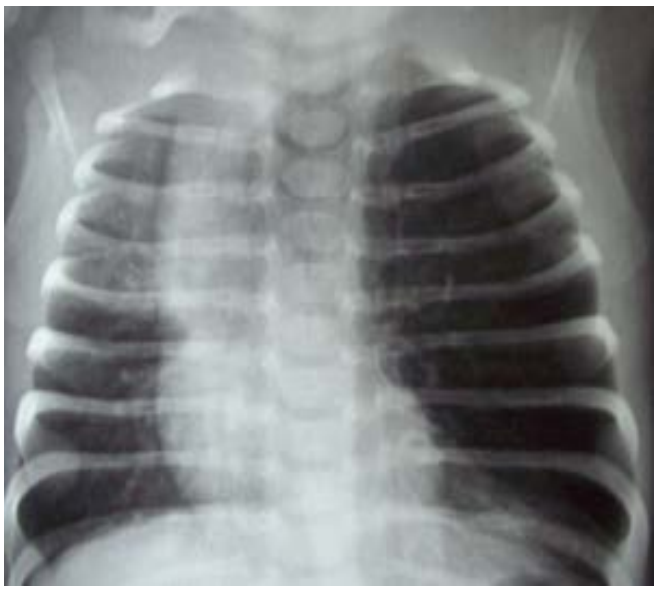

Fig.1: Chest X-ray PA view. Showing marked hyperlucency of the the left upper lobe and collapse of the ipsilateral remaining lung field with mediastinal shift to the right

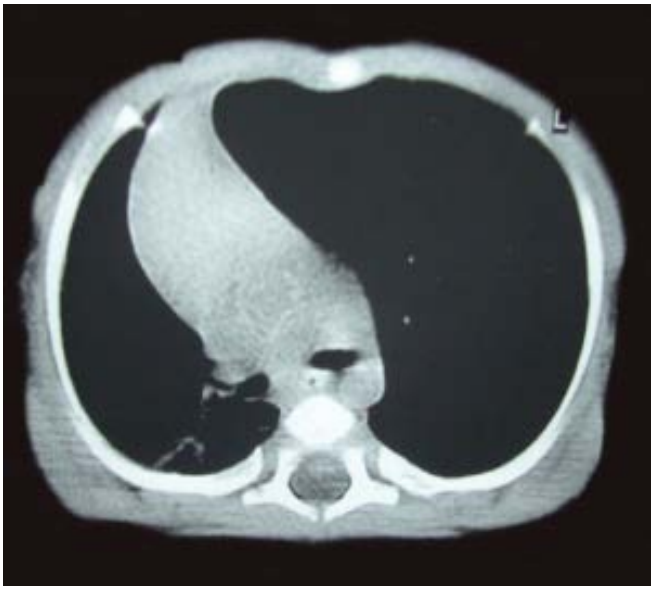

Fig.2: CT scan showed a hyperinflation of the left upper lobe with trachea-mediastenal shift towards right. Patient underwent surgical lobectomy of the involved lobe and a gross specimen of single greyish brown soft to firm lobular tissue mass of size 6.8 x 4. x 2cm was received.(Fig.3) Cut surface was spongy and brownish white.(Fig.4)

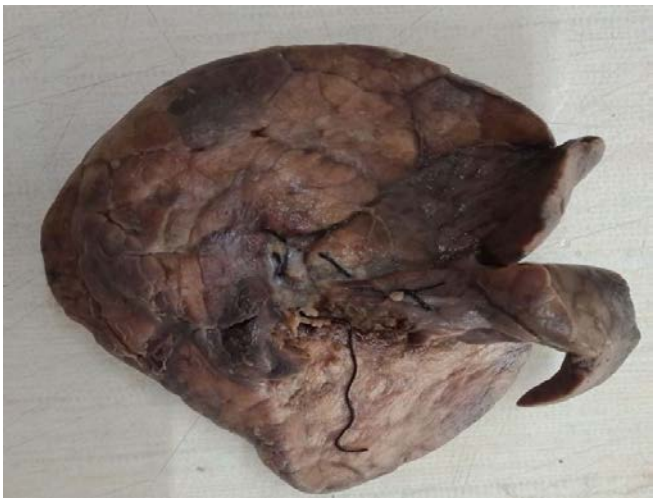

Fig. 3: Gross- Single greyish brown soft to firm lobular tissue mass.

Fig. 4: Cut surface was spongy and brownish white

**Microscopically** multiple sections were studied. They revealed dilated air spaces with thin alveolar septae.(Fig.5). Focal areas of collapse(Fig.6) and foci of interstitial pneumonia were seen showing thickened interstitial septae with collection of mononuclear inflammatory infiltrate and congested blood vessels.(Fig.7) Many bronchi with cartilage in the wall were also seen.(Fig.8)

The muscular layer appeared poorly formed.

There were no evidence of destruction of alveolar walls and alveolar septum as seen in acquired emphysema.

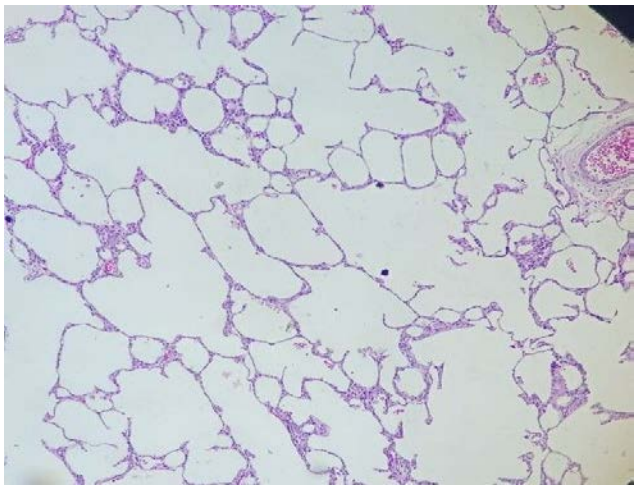

Fig. 5: Photomicrography of dilated air spaces with thin alveolar septae. (Hematoxylin and Eosin stain 10X)

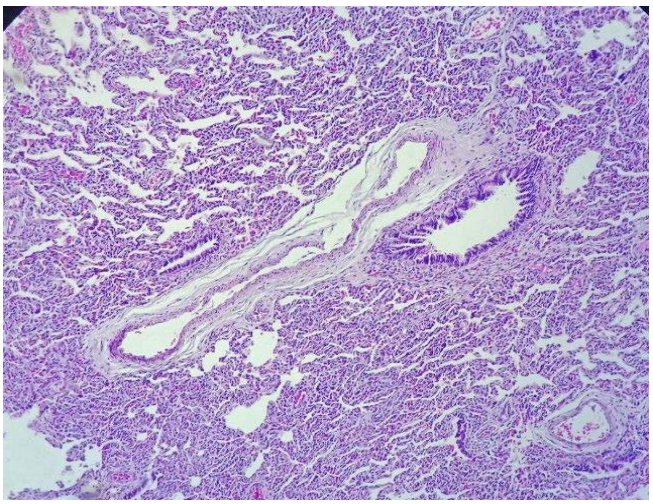

Fig. 6: Photomicrography from focal areas of collapse (Hematoxylin and Eosin stain 10X)

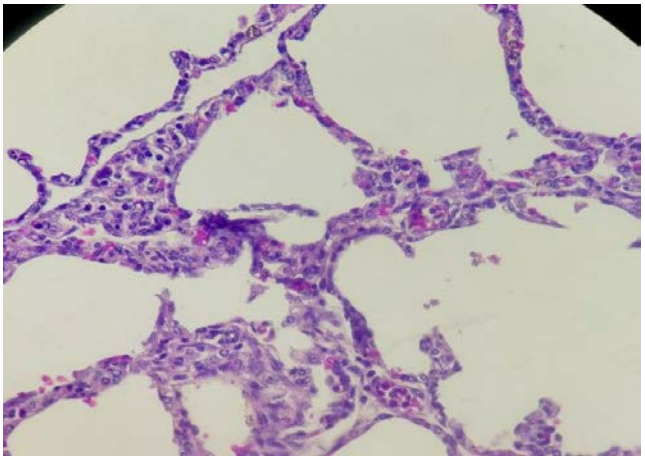

Fig. 7: Photomicrography from focal areas having mononuclear inflammatory infiltate in thickened interstitial septae (Hematoxylin and Eosin stain 40X)

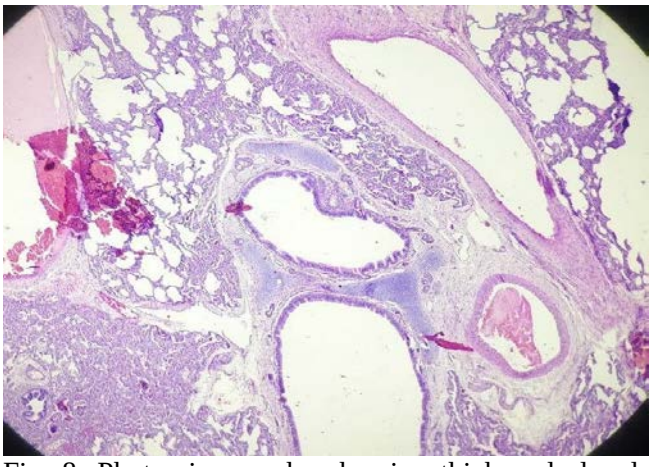

Fig. 8: Photomicrography showing thickened alveolar septae, areas of collapse and bronchial cartilage. (Hematoxylin and Eosin stain 4X).

Final microscopic diagnosis of left upper lobe of lung was given as histological features consistent with clinical and radiological diagnosis of congenital over inflation with foci of interstitial pneumonia and collapse.

**Discussion**

Our above discussed case had all the features suggestive of CLE where the child presented in the neonatal period which is most common. This case is of particular interest for pathologists, radiologists, pediatricians and pediatric surgeons.

The etiology of congenital lobar emphysema is idiopathic in 50-55% of the patients. The formation of CLE in the remaining 25% cases may be caused by abnormal lobar bronchus development which is most often due to a cartilage defect, less frequently a muscular coat or mucous membrane fold defect.[6] Due to absent or hypoplastic defective cartilage, there occurs bronchial collapse thus leading to air trapping during expiration. Our case did not show any cartilaginous defect however poorly formed muscular layer at places were noticed. Other causes includes parenchymal disease along with internal and external bronchial obstruction in the form of bronchial stenosis and compression of the abnormally located vessel, tumor etc.

There is typically a history of chronic cough and recurrent respiratory infections and the patient usually presents with difficulty in feeding and breathing, wheezing and cyanosis. Low incidence rate and chances of co-infection often cause misdiagnosis and delay in treatment.[8] The overlapping symptoms can

also leads to wrong diagnosis of some patients as just pneumonia or pneumothorax.

Children with congenital lobar emphysema could even be prenatally diagnosed with ultrasonography which was missing in our case. [9] A study of Eber et al showed that the lungs of a fetus affected with congenital lobar emphysema show hyper-echogenicity without any abnormal blood flow. However histopathological confirmation will be required to differentiate CLE from other radiological differentials like congenital pulmonary lesions such as bronchopulmonary sequestration, congenital cystic adenomatoid malformation, congenital diaphragmatic hernia and bronchogenic cyst. [8]

A histopathological distinction is necessary to make between a polyalveolar lobe, in which the number of alveoli is markedly increased from congenital lobar over inflating, in which the alveoli are markedly distended [3] The pathological change in CLE comprises of massive over distension of the alveolar spaces which is not accompanied by destruction of tissue. Therefore it is a not a true cystic or an emphysematous process.[10]

Although not seen in our case, 12 to 20% cases of CLE are associated with concomitant congenital heart disease (CHD) . Uncommonly, renal, gastrointestinal, musculoskeletal and cutaneous malformations may also occur.[3]

Early surgical management in case of congenital lobar emphysema helps to avoid irreversible consequences. Although surgical removal of the affected lobe is the most commonly accepted treatment, controversy exists regarding surgical and conservative management of this malformation. One opinion is in favor of conservative management for small number of cases who are are not clinically in respiratory distress and able to feed and grow but then stringent follow up is necessary. In our case, patient presented with moderate respiratory distress where surgical management was opted.

**Conclusion**

A high index of suspicion is required to diagnose congenital lobar emphysema and it should always be considered as a differential diagnosis in neonates or infants presenting with respiratory distress. Although CLE is an uncommon and a neglected disease of uncertain etiology, the radiological and histopathological findings assist in diagnosis. Protocol for surgical or conservative management should be individualized according to the clinical presentation and status of the child and the status of the family

**References**

1. Aditya K, Parvathi K, Prabhakar V.A case report of congenital lobar emphysema. International Journal of Contemporary Pediatrics. 2020 Apr;7(4):945-947
2. Demir OF, Hangul M, Kose M. Congenital lobar emphysema: diagnosis and treatment options. International journal of chronic obstructive pulmonary disease. 2019;14:921
3. Correia-Pinto J, Gonzaga S, Huang Y, Rottier R. Congenital lung lesions—underlying molecular mechanisms. InSeminars in pediatric surgery 2010 Aug 1 (Vol. 19, No. 3, pp. 171-179). WB Saunders.
4. Datta AK, Mandal S, Jana JK. Congenital lobar emphysema: a case report. Cases journal. 2009 Dec;2(1):1-3.
5. Chandran-Mahaldar D, Kumar S, Balamurugan K, Raghuram AR, Krishnan R. Congenital lobar emphysema. Indian journal of anaesthesia. 2009 Aug;53(4):482.

6. Minira B, Anna M, Aigul B. Case of congenital lobar lung emphysema. J Histol Histopathol. 2014;1:13.
7. Cataneo Cataneo DC, Rodrigues OR, Hasimoto EN, Schmidt AF, Cataneo AJ. Congenital lobar emphysema: 30-year case series in two university hospitals. Jornal Brasileiro de Pneumologia. 2013 Jun;39:418-26.
8. Lei Q, Zeng W, Ju R. Congenital lobar emphysema in bilateral lung lobes: a case report. Translational Pediatrics. 2020 Jun;9(3):266.
9. Eber E. Antenatal diagnosis of congenital thoracic malformations: early surgery, late surgery, or no surgery? Semin RespCrit Care Med 2007, 28(3):355-66.
10. Goldblum JR, Lamps LW, McKenney JK, Myers JL. Rosai and Ackerman's Surgical Pathology E-Book. Elsevier Health Sciences; 2017 Oct 25.
11. Ulku R, Onat S, Ozçelık C. Congenital lobar emphysema: differential diagnosis and therapeutic approach. Pediatrics International. 2008 Oct;50(5):658-61.
12. Olutoye OO, Coleman BG, Hubbard AM, Adzick NS. Prenatal diagnosis and management of congenital lobar emphysema. Journal of pediatric surgery. 2000 May 1;35(5):792-5.
13. Suryawanshi K, Nikumbh D, Singhavi S, Damle R, Dravid N. Congenital lobar emphysema with pulmonary extramedullary hematopoiesis. Turk Patoloji Derg. 2017 Jan 1;33(1):74-6.
14. Mulvany JJ, Weatherall A, Charlton A, Selvadurai H. Congenital lobar emphysema: diagnostic and therapeutic challenges. Case Reports. 2016 Jun 22;2016:bcr2016214994.
15. Thakral CL, Maji DC, Sajwani MJ. Congenital lobar emphysema: experience with 21 cases. Pediatric surgery international. 2001 Mar 1;17(2-3):88-91.
16. Lincoln JC, Stark JA, Subramanian S, Aberdeen EO, Bonham-Carter RE, Berry CL, Waterston DJ. Congenital lobar emphysema. Annals of surgery. 1971 Jan;173(1):55.