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Congenital Lobar Emphysema (Overinflation) Due to Bronchomalacia: A Rare Case Report

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ABSTRACT

Congenital lobar emphysema is a rare variety of congenital malformation of lung characterized by over distension of a lobe of a lung due to partial obstruction of the bronchus(1). We are reporting a 2month old female baby admitted in the pediatric emergency ward with respiratory distress. CT suggested congenital lobar emphysema. Lobectomy was done, diagnosed as Congenital lobar emphysema with bronchomalacia and is presented for its rarity.

Keywords: Congenital Lobar emphysema, Respiratory distress, Bronchomalacia,

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INTRODUCTION

Congenital lobar emphysema (CLE) is a congenital malformation of lung characterized by unilobar alveolar distension secondary to bronchomalacia or absent cartilage is a rare condition [1-5]. It is caused by the hyperinflation of the lung lobe with compression of the normal lung parenchyma and contra lateral displacement of the mediastinum [4]. Male babies are affected more often than female in the ratio of 3:1and most commonly involves left upper lobe [3,5]. The usual presentation is during the 1st six months of life in most cases. Breathlessness with or without cyanosis is the common presentation and management has been surgical [1,5,6].

Case Presentation

A 2month old female baby weighing 3.1kg was referred to pediatrics emergency ward of a tertiary care hospital in South Tamil Nadu with complaints of respiratory difficulty for 1week. On examination, baby was presented with difficulty in breathing, tachypnea, retraction of ribs and cyanosis. On examination of respiratory system , trachea was shifted to the left, vocal resonance was decreased in right side and on auscultation diminished breath sounds was found on right side.

Radiographs of the chest showed overinflated right middle lobe and collapse of the left lobe with slight mediastinal displacement to the left (Fig1). During the next few days the symptoms became more pronounced and gradually the percussion note over the right middle lobe became more and more hyper-resonant and the air entry into the left lung diminished.

CT scan showed a hyperluscent, hyper extended lobe with midline substantial herniation and compression of the remaining lung. The mediastinum is significantly shifted away from the side of the abnormal lobe (Fig2).So right middle lobectomy was done.

Gross: We received a formalin-fixed specimen of soft tissue pieces from overinflated right middle lobe (Fig 3) ,measuring 9x5x3cm showed spongy , foaming with spotty hemorrhages.

Microscopically (Fig 4-7), alveolar spaces were distended, some filled with erythrocytes. Bronchiolar ducts were distended. No cartilage seen in multiple sections studied.

The patient did well after lobectomy.

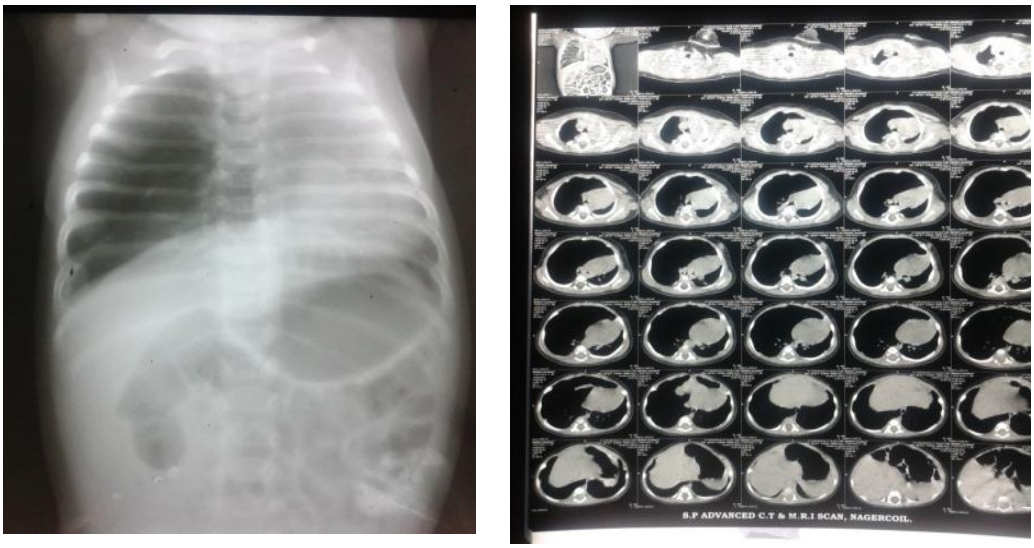

Figure 1,2: X-ray (Fig1) and CT.
(Figure 2 showed overinflated right middle lobe)

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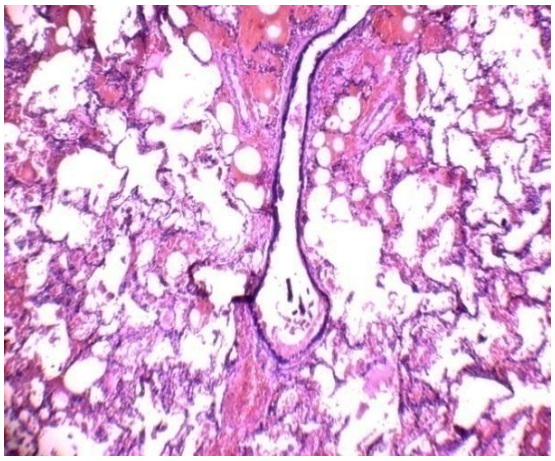
Figure 3: Gross appearance of excised right middle lobe.



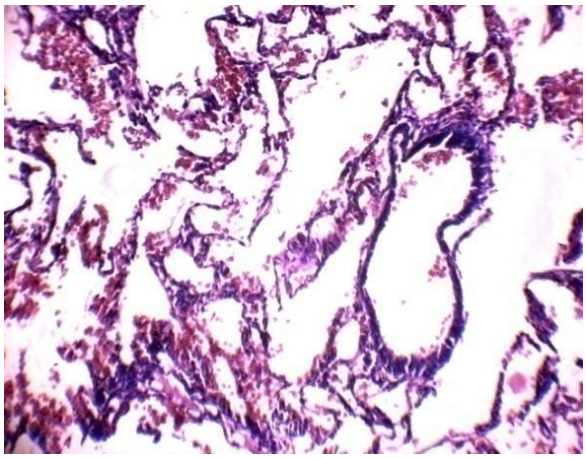
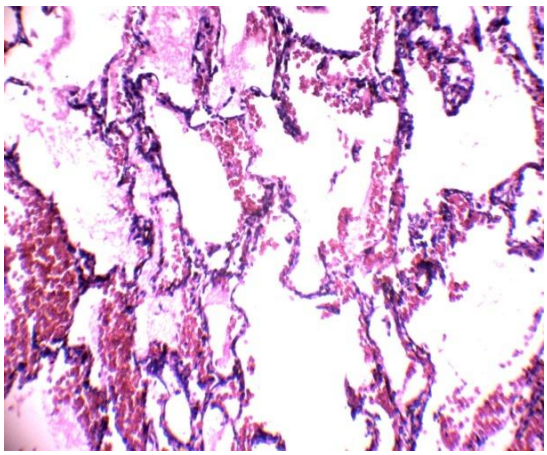


Figure 4,5: Microscopic appearance (H&E 5X and 10X) showing distended alveolar spaces and bronchiolar ducts.



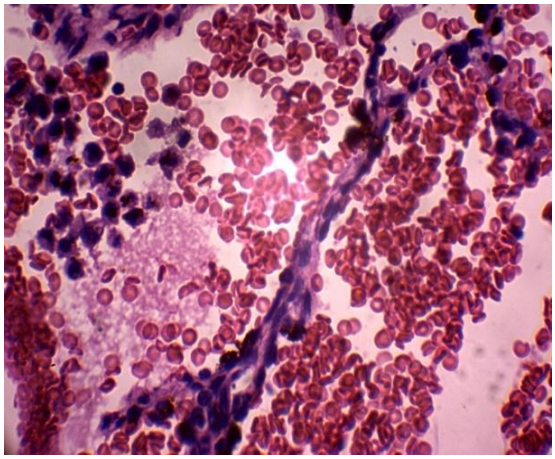


Figure 6,7: High power view (H&E 20X and 40X) showing distended alveolar spaces and some filled with erythrocytes

DISCUSSION

Congenital lobar emphysema (CLE) is a rare developmental anomaly of the lower respiratory tract, which is characterized by hyperinflation of one or more of the pulmonary lobes [1,3,5,7].

It is a rare disease with the incidence of approximately 1in 70,000 to 1in 90,000 live births, more common in neonatal period upto 6months of age and more common among boys and whites [1,3,5,8,9].

In congenital lobar overinflation, there are many presumed mechanisms for progressive overdistension of a lobe including , disruptions of bronchopulmonary development due to abnormal

interactions between embryonic endodermal and mesodermal components of the lung, dysplasia, check-valve mechanism at the bronchial level [5,7].

About 50% of cases have no exact etiology, however 25% of cases have bronchial cartilage dysplasia [3,5,7-9]. There is one previous report of mother and daughter both with CLE of right middle lobe and another report of father and son with CLE of right upper lobe, suggesting of inheritable basis of condition(5). By molecular genetics, normal branching of lung morphogenesis is regulated by FGF-10 gene and NK2homeobox1(also known as thyroid transcription factor1). If there is any mutation in these gene will lead to lung anomalies and deficiency of bronchial cartilage [5,7,10].

Most of the patients are asymptomatic or present with respiratory distress in the newborn period. Later, infants may experience dyspnoea (57%)or recurring respiratory infection (28%) [1,8,10].

On chest X-Ray, increased aeration of the affected lobe with widening of the ribs and interspaces is the characteristic features and herniation to the other side of the chest, mediastinal shift, collapse of the adjacent lobes on the ipsilateral side and flattening of the diaphragm on this side [1,6,8]. The diagnosis may be confirmed on chest CT, hyperluscent, hyper extended lobe attenuated but intact pattern of organized vascularity [1,3].

CLE may mimic other congenital cystic malformations of lung like cystic adenomatoid malformation, Bronchogenic cyst and Pulmonary sequestration [6,11]. Radiography and Histopathology helps in arriving at a final diagnosis.

The management of CLE has been immediate, surgical excision is required for patients with respiratory distress. First case of CLE successfully treated by lobectomy was reported in 1945 [5]. However, infants and older children who are asymptomatic or have minimal symptoms can be treated conservatively [1,8,9]. The prognosis is usually excellent after resection of congenital lung lesion [5].

This case is presented for its rarity and is a notable cause of respiratory distress in neonates. The outcome of surgery is good in most cases and symptomatically satisfactory despite of minor changes in pulmonary function test.

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