

## CONGENITAL LOBAR EMPHYSEMA : A RARE CASE REPORT

## General Surgery

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## ABSTRACT

**Introduction:** Congenital lobar emphysema is a rare developmental anomaly of lung, most of them presents within first 6 months of life. In this condition there is hyperinflation of one lobe of lung because air enters the lung but cannot escape causing overinflation of lobe or lung. Incidence of congenital lobar emphysema is 1/20,000-30,000 live birth. **Case:** In this case report we are presenting a case of 1 month 10 days old Male infant presented with features of tachypnea, subcostal retraction with respiratory distress. After investigation diagnosis of Congenital Lobar Emphysema was confirmed, for which excision of Left upper (emphysematous) lobe was done. Histopathological report was suggestive of congenital overinflation with foci of interstitial pneumonia and collapse. In such case surgical management is mainstay treatment.

## KEYWORDS

Congenital Lobar Emphysema, Hyperinflation, Surgical lobectomy.

## INTRODUCTION:

Congenital Lobar Emphysema is rare developmental anomaly of the lung seen in newborn or in young infant characterised by hyperinflation of one lobe of one lung compressing ipsilateral and/or contralateral lung tissue and mediastinal shift. Synonyms for congenital lobar emphysema is congenital lobar overinflation and infantile lobar emphysema.<sup>[1]</sup> Incidence of congenital lobar emphysema is 1/20,000-30,000 live birth with male to female ratio of 3:1.<sup>[2]</sup>

Airway obstruction can be intrinsic, intraluminal or extrinsic, with the former being more common. This leads to the creation of a "ball-valve" mechanism, in which a greater volume of air enters the affected lobe during inspiration than leaves during expiration, producing air trapping. Additionally, bronchial atresia and pulmonary sequestration have been identified as a common finding in Congenital Lobar Emphysema.<sup>[3]</sup>

Heart problems (15% cases) are seen associated with this condition.<sup>[1]</sup> Patient of Congenital Lobar Emphysema usually presents with Dyspnea, tachypnea, retraction, increased intrathoracic pressure, coughing. Misdiagnosis on initial x-ray chest with pneumothorax was common.<sup>[4]</sup>

Radiological investigations like Chest X-ray, CT scan is helpful for diagnosis of Congenital Lobar Emphysema. Surgical management is only treatment available for this condition.

## CASE REPORT:

A 1 month 10 days old male infant born by caesarean section [For Cephalopelvic disproportion] with birth weight of 2.2kg was admitted with complaints of tachypnoea and not accepting feed since 1 day.

On examination patient was afebrile, drowsy with Respiratory rate = 60/min and SpO<sub>2</sub> = 95% at room air. Patient was tachypneic with nasal flaring, subcostal retraction with respiratory distress. Patient had a previous history of respiratory distress with NICU stay for 1 month [sepsis with congenital pneumonia].

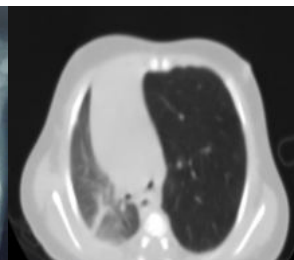
X-ray chest was suggestive of hyperinflated left lung with herniation into the Right thorax with mediastinal shift (Figure.1). C.T scan of Thorax was suggestive of Hyperinflation of left upper lobe with trachea mediastinal shift towards the right and atelectasis of left lower lobe and partial compression right lung (Figure.2).

Left posterolateral thoracotomy was done. Operative findings were hyperinflation of Left upper lobe with collapsed left lower lobe

(Figure.3, Figure.4 and Figure.5). Resection of affected left upper lobe was done. Surgical procedure was uneventful. Histopathological report of excised pathological upper lobe of left lung suggestive of congenital overinflation with foci of interstitial pneumonia and collapse (Figure. 7).



**Figure.1** Chest X-ray AP View Preoperative



**Figure.2** CT Thorax



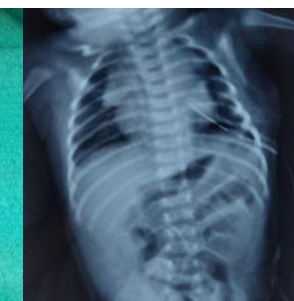
**Figure.3** Excised pathological upper lobe of left lung



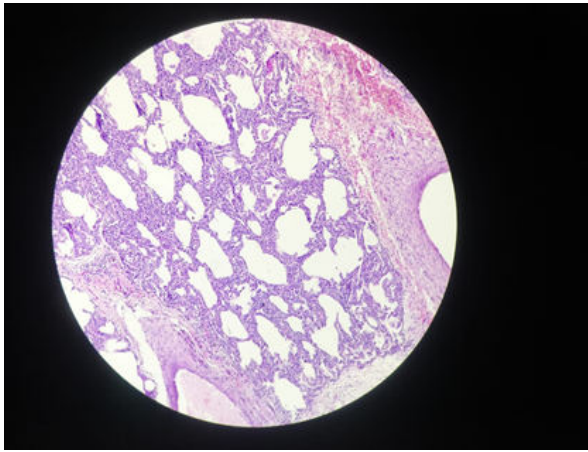
**Figure.4** Post thoracotomy image.



**Figure.5** Excised Left Upper Lobe Of Lung



**Figure.6** Chest X-ray AP view Post operative



**Figure.7** Histopathological Image Of Excised Pathological Upper Lobe Of Left Lung.

#### DISCUSSION:

Congenital lobar emphysema is congenital abnormality of lung which has a wide range of presentation most of them presents within first 6 months of life. In 25% of cases congenital lobar emphysema presents with life threatening acute respiratory distress in neonatal age group. Congenital lobar emphysema is not a true emphysema because in this alveolar distention is there without actual wall destruction. In congenital lobar emphysema hyperventilation occurs in one lobe of lung which worsens the functional capacity of other lobe of same lung or the other lung.

Usually, unilateral and upper left lobe most affected (42%), but the right middle (35%) also affected. Bilateral involvement is exceptionally rare.<sup>[1]</sup> Chest wall retraction, tachypnea, breathing difficulty, cyanosis, recurrent pneumonia and failure to thrive are the characteristic features of the congenital lobar emphysema. Imaging techniques like Chest Xray, CT Thorax and MRI Thorax are used for confirmation of diagnosis of Congenital Lobar Emphysema and ruling out other pathological conditions.

In our case the affected lobe (Left upper lobe) was resected and baby recovered well post operatively with good lung expansion. Early diagnosis and early surgical treatment have good outcome with good lung expansion and normal growth of the patient without any morbidities.

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