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Case Report

Congenital lobar emphysema presenting as recurrent pneumonia in an infant

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ABSTRACT

Congenital lobar emphysema (CLE) is a developmental anomaly of the lower respiratory tract, which is characterised by hyperinflation of one or more lobes of lung. CLE is potentially reversible, though possible life threatening cause of respiratory distress in neonate. Here we present a 4 months baby who presented with respiratory distress related to CLE.

Keywords: Congenital lobar emphysema, Chest x-ray, CT scan, Lobectomy

INTRODUCTION

CLE is a rare congenital malformation, with a prevalence of 1 in 20,000 to 1 in 30,000.[1-3] Over distension of the pulmonary lobe is secondary to partial bronchial obstruction.[4] Left upper lobe is the most common affected lobe followed by right upper lobe and right middle lobe but any lobe may be affected. Patients often present within the first 6 months of life with recurrent respiratory distress.[5]

Chest x-ray and CT scan of thorax are diagnostic investigation and show the hyperluscent affected lobe, shifting the mediastinum to the opposite side and remaining part of the ipsilateral lung will be collapsed.[6] Concomitant congenital heart disease (CHD) can be associated in CLE. In the literature, a 12 to 20% concomitant rate is given.

CASE REPORT

A 4 months old male infant got to our hospital with complaints of cough and hurried breathing of 3 days duration. History of recurrent episode of respiratory distress in the past 2 months of admission. He was from a poor socioeconomic status family. Antenatal, natal, postnatal history was uneventful. Baby was breast fed and immunized as per NIP.

On examination, the baby was found in respiratory difficulty, respiration rate was 75 per minute, heart rate120 per minute, weight was 5.8 kg, length 59 cm and head circumference 51 cm. Head to toe examination was normal. There was no pallor, cyanosis, jaundice or any signs of heart failure.

Examination of respiratory system revealed less movement of left upper chest, trachea was shifted to the right, hyper resonant note on percussion in left side infraclavicular and mammary region and on auscultation diminished breath sound was found on left side. Right side normal vesicular breath sounds were heard and crepitation were heard in the right infraclavicular region and mammary region .Other systems appeared normal. Blood investigation were normal. Chest X-ray showed marked overdistension of the left upper lobe with mediastinal shift to the right, patchy opacity in the right upper lobe (Figure 1).

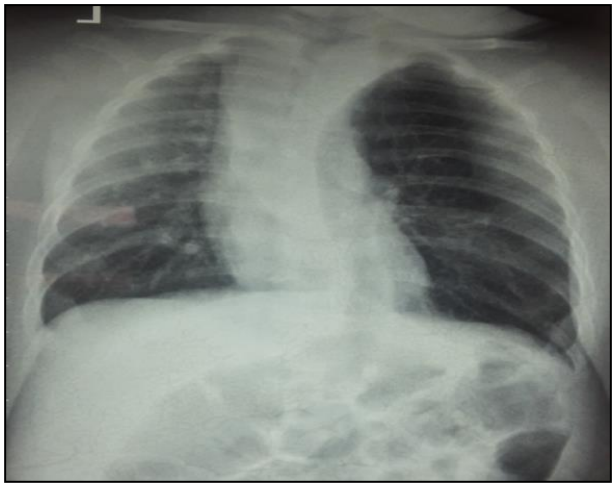

Figure 1: Chest x-ray preoperative.

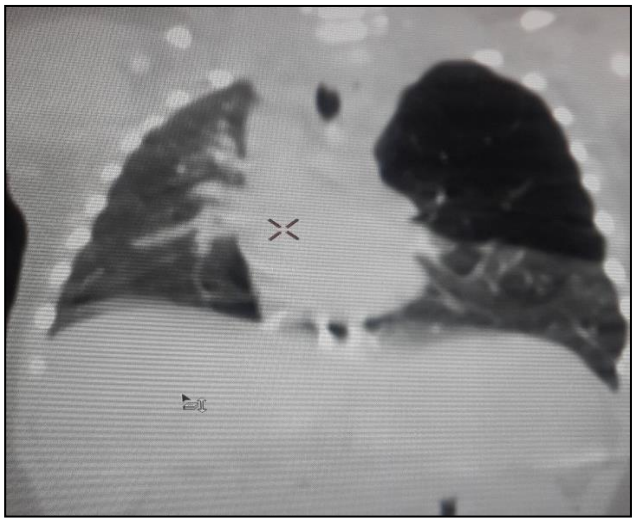

Figure 2: CT scan -hyperluscent, hyperinflation of left upper lobe with paucity of bronchovascular marking.

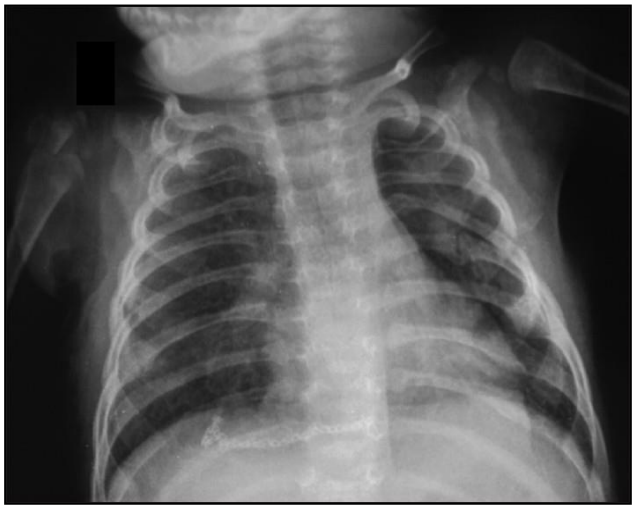

Figure 3: Postoperative chest x-ray showing expansion of lungs in the upper left lobe.

CT scan thorax showed a hyperluscent, hyperinflation of left upper lobe with paucity of bronchovascular marking with mediastinal shift to the right side likely to be CLE ill-defined patchy consolidation in the posterior segment of the right upper lobe likely to be infective etiology (Figure 2).

Child was treated with injection augmentin for 5 days and nebulization was given. After the clearance of pneumonia child was referred for surgery to higher centre. Lobectomy was done. Post-operative X-ray revealed no mediastinal shift, normalization of left lung. After 15 days of surgery child was followed up in our hospital. clinically improved and breathsounds were heard on both side normally. Child is feeding well no complaints.

DISCUSSION

In few cases there will be weakened or absent bronchial cartilages so that there is inspiratory air entry but collapse of the narrow bronchial lumen during expiration. This bronchial defect results in lobar air trapping7. Microscopically, cartilage plates in the bronchi are absent at the level where the cartilage is expected.

Clinically subtle or obvious respiratory distress is observed in an otherwise normal infant, asymmetry of chest and abdominal retractions on inspiration. Hypoxemia (in severely affected patients) may occur. The diagnosis is often suspected upon in utero sonography if an overexpanded lobe filled with fluid is identified. Progressive respiratory distress from birth reflects the degree of emphysema; occasionally, patients present in later childhood or even during adulthood.[8-10]

The basic investigation in CLE is the chest radiograph from which a diagnosis can be made and is readily available. CT, bronchoscopy and angiopulmography are also used in the diagnosis.[8,11] For infants with typical CLE and progressive respiratory distress, immediate surgical intervention i.e lobectomy is indicated.[12] Without surgery, the mortality rate among these patients is 50%, and 75% of the survivors have persistent respiratory distress; children with mild or asymptomatic form of disease have been managed conservatively without serious sequel.[13,14] Most surgically treated patients will be asymptomatic and will have normal growth and development.[13]

CONCLUSION

The early diagnosis of CLE can be done Antenatally. Children with mild or asymptomatic disease can be managed conservatively. Lobectomy should be performed in patients with severe respiratory distress which is life-saving surgery. The importance of follow-up should also be stressed to assess for growth and development.

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