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Case Report

Congenital lobar emphysema: a rare cause of respiratory distress in a neonate

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ABSTRACT

Congenital lobar emphysema (CLE) is a congenital condition characterized by distension and air trapping of the affected lobe of the lung. It is one of the causes of infantile respiratory distress, which may require surgical resection of affected lobe. Case characteristics: 3-day-old neonate with ventilation refractory respiratory distress. Imaging was suggestive of decreased lung tissue on the right side with ipsilateral mediastinal shift. Intervention/ outcome: Early surgical lobectomy was done to improve lung functions and the child improved dur to early intervention. Message: An early diagnosis with high index of suspicion helps patients with this rare congenital anomaly. Early intervention is the key to good long-term outcome. More awareness about the entity and treatment options available would greatly help improving the outcome and disease burden.

Keywords: Congenital anomaly, Congenital lobar emphysema, Lobectomy

INTRODUCTION

Congenital Lobar Emphysema (CLE) or Congenital Lobar Over-inflation (CLO) or infantile lobar emphysema is a rare entity causing acute onset respiratory distress in a newborn.[1,2] Incidence has been reported to between 1 in 70000 to 1 in 90000 live births.[3,4] There is post-natal hyperinflation of one or more lobes of the lung, presenting in infancy. This results in compression atelectasis, mediastinal shift and subsequent cardio pulmonary compromise. Its early recognition and surgical intervention can be lifesaving.[3-5] Even today, despite advanced diagnostic techniques, pitfalls in diagnosis and management are not uncommon. The initial imaging may not show typical features and may masquerade as pneumonia.[5,6] Authors here report a 3-day-old neonate with severe respiratory distress, requiring surgical resection of the affected lobe.

CASE REPORT

A late preterm 2 kg was born via LSCS in view of polyhydroamnios to a P2L1A1 mother. Antenatally, the mother had been hospitalized for polyhydroamnios thrice in a medical college, details not available. Baby cried immediately after birth. The first 3 postnatal days were also uneventful. The previous conceptus had IUD at full term gestation, only available history of polyhydroamnios. This baby developed acute onset respiratory distress on day 3 of life. CPAP was initiated and in view of respiratory distress and impending respiratory failure, the baby was referred for mechanical ventilation. Sepsis screen including a blood culture were negative. Chest X-ray showed an image of reduced lung tissue on right side with shift of mediastinum to the right (Figure 1).

A diagnostic dilemma of dextrocardia versus dextroposed heart was faced and further imaging was ordered. CT chest was done to pin point the diagnosis, but the picture was unequivocal. Baby improved significantly on antibiotics and good supportive care. The baby was not put on CPAP anytime in our centre.

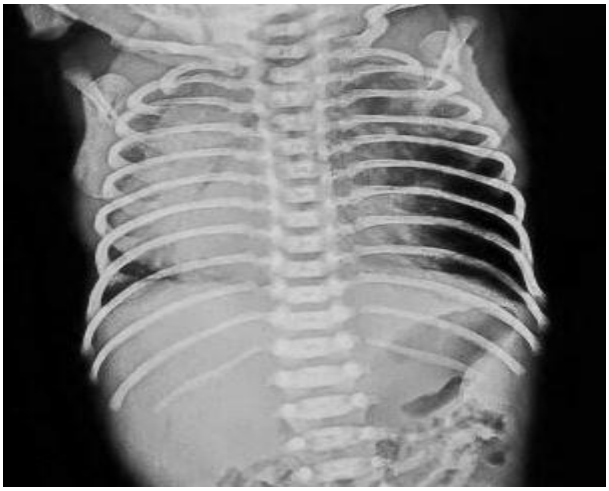

Figure 1: Chest X-ray shoeing hyper-lucent lung fields with mediastinal shift to the right.

By day 10 of life, the baby had improved significantly but was still requiring minimal oxygen supplementation by nasal prongs. Due to persistent oxygen requirement, congenital structural abnormalities were suspected. Repeat contrast CT chest was suggestive of significant hyperinflation with attenuation of the vascularity in the left upper lobe (Figure 2). A diagnosis of congenital lobar emphysema (CLE) was made. The baby underwent surgery, which was suggestive of CLE of the left upper lobe sparing the lingua. The baby improved post-surgery, oxygen requirement came down and feeds were established.

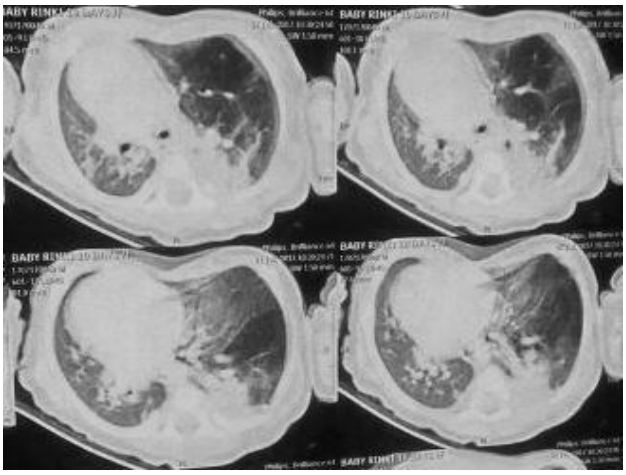

Figure 2: Contrast CT Chest in the axial cuts lung window showing hyper-inflated and hyper-lucent left upper lobe with associated mediastinal shift to the right.

DISCUSSION

Congenital lobar emphysema (CLE) is a rare congenital abnormality characterized by excessive aeration of a part of the lung. Progressive pulmonary hyperinflation may be caused by abnormal interactions between embryonic endodermal and mesodermal components due to a ball-valve effect.[7] Respiratory distress maybe due to massive over distension of the affected lobe and subsequent compression of surrounding structures.[8] Involvement of all lobes has been reported but left upper lobe involvement is most common. Cardiovascular defects occur in approximately 14% of infants with CLE. Renal agenesis, renal cysts, and limb anomalies may associate this disease.[9] Depending on the degree of bronchial obstruction, clinical presentation may vary from mild tachypnea to severe life-threatening respiratory distress.[10] There may or may not be mediastinal shift. Decreased bronchial cartilage tissue producing a ball valve effect with consequent over-inflation is seen in 50% of the patients.[11] The age of onset of symptoms ranges from a few days after birth to 6 months.[5] Most patients develop symptoms in the neonatal period and the male: female ratio is 3:1.[12] However, some patients may be asymptomatic and present later in life. Santra et al, reported such a case in which CLE was diagnosed at the age of 15 years.[13]

As in this baby, respiratory distress is the commonest mode of presentation. The diagnosis of CLE requires a high degree of suspicion. CLE may be misdiagnosed as pneumothorax and in such cases insertion of an intercostal drainage tube may be detrimental to the patient. On the other hand, in most cases such babies may be misdiagnosed as pneumonia leading to unnecessary antibiotic treatment and loss of valuable time in diagnosis.[14] In an Indian series, a correct diagnosis on chest radiography before referral was made in only 4 out of 10 cases even though the diagnosis could be arrived at in the 10 cases using the initial radiographs.[5] In our case, the lesion was suspected due to persistent oxygen requirement, absence of clinical and laboratory signs of septicemia and suggestive radiological findings.

Treatment of CLE is essentially surgical, i.e., lobectomy. Early surgical intervention has been found to significantly improve the condition of the patient.[15] Nazem et al found an unusually high mortality of 13.3% in their study of 30 operated cases of CLE. They concluded that the number of lobes affected and base deficit at presentation were associated with a high mortality rate in their study.[16]

CONCLUSION

CLE/ CLO is an extremely rare congenital lung abnormality which maybe missed on routine antenatal scans. It may or may not present at birth. Hence a high index of suspicion is required to look for causes of clinical distress in the absence of more common causes

like hyaline membrane disease and septicaemia. Careful radiological interpretation is imperative for diagnosis. A prompt surgical resection may be life saving.

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