Case Report

Congenital lobar emphysema: Pitfalls in diagnosis

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ABSTRACT

Congenital lobar emphysema (CLE) is a rare but life-threatening congenital anomaly leading to respiratory distress in early childhood. Diagnosis requires a strong clinical suspicion. We report a case of a 31/2-month-old infant who was initially diagnosed with pneumonia requiring multiple hospital admissions. After computed tomography of the thorax, a diagnosis on CLE was made. The child was planned for surgery in the next available routine operation theatre. However, suddenly in the evening, she developed respiratory distress and needed emergency surgical intervention. The child improved dramatically after surgery, and the postoperative period was uneventful. Early diagnosis and treatment in such cases can lead to dramatic results.

KEY WORDS: Congenital lobar emphysema, lobectomy, pneumonia

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INTRODUCTION

Congenital lobar emphysema (CLE) is a rare but life-threatening congenital anomaly of the lower respiratory tract with an incidence of one in 20,000–30,000 deliveries. It is characterized by hyperinflation of a pulmonary lobe with compression of the surrounding lobes and in severe cases causing mediastinal shift.[1] The left upper lobe is most commonly involved followed by the right upper lobe and right middle lobe.[1] Usually known to affect a single lobe, reports of multilobar and bilateral involvement have also been reported in literature.[2]

CASE REPORT

A three and a half month old female child being treated in another hospital was referred to us. This child had complaints of cough, coryza, fever, and respiratory distress since day 10 of life. The child was admitted in another hospital and was treated for pneumonia. Chest X-ray (CXR) at that time showed collapse consolidation of the apical segment and lower lobe of the right lung which was misread as lobar consolidations [Figure 1]. She was given intravenous antibiotics and was discharged. Subsequently, this child required four more readmissions and was suspected of having recurrent pneumonia. During this course, her blood investigations were essentially normal. Two-dimensional echo was normal. A cystic fibrosis screening was done which also was normal. This child was then referred to Lok Nayak Hospital, New Delhi. On examination, the child was alert, active with stable vitals. Blood investigations were essentially normal. Venous blood gas showed pH 7.2, PCO_2 54, and BE 5.4. CXR showed hyperlucency in the right upper and middle zone with slight mediastinal shift to the left. An inhomogeneous opacity was noted in the right middle zone along with a thin linear opacity in the left hemithorax which led to the suspicion of right lobar emphysema [Figure 2]. Computed tomography (CT) scan was planned which showed collapse of apical and posterior segments of the right upper zone along with hyperinflation of the right middle lobe. Consolidation with subsegmental collapse was seen in the apical segment of both lower lungs [Figure 3].

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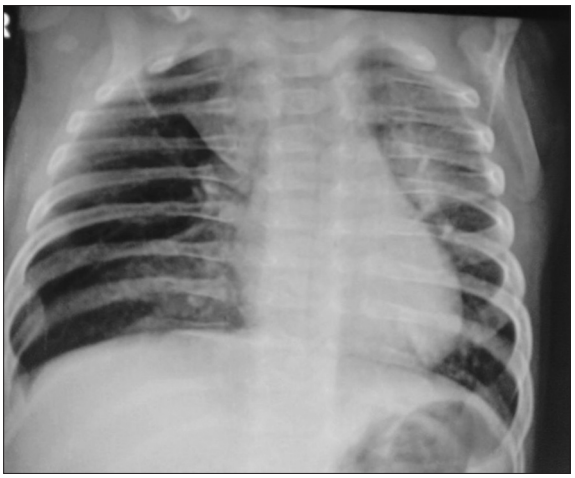

Figure 1: Chest X-ray done elsewhere showing consolidation in right upper and middle lobes

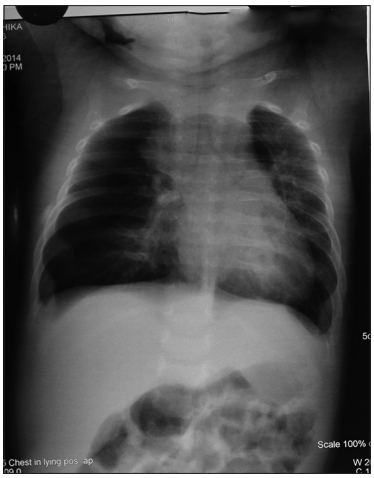

Figure 2: Chest X-ray showing hyperlucency in right upper and middle zone with slight shift of mediastinum to the left

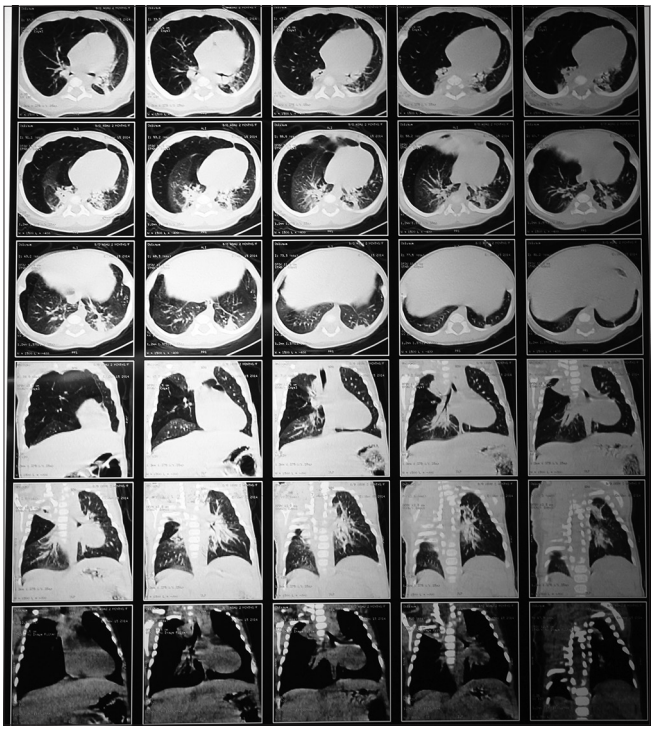

Figure 3: Computed tomography thorax showing collapse of apical/postsegment of the right upper zone. Hyperinflation of right middle lobe

Figure 4: Intraoperative photo of the emphysematous right middle lobe

A diagnosis of CLE was made. As the child was clinically stable, a decision to manage her conservatively was taken. However, on the 7th day of admission, the child's condition deteriorated with worsening respiratory distress and was unable to maintain saturation even with oxygen support. The child developed severe chest retractions. The child was then taken up to the emergency operation theatre for urgent thoracotomy and exploration. Right middle lobe was found emphysematous [Figure 4] and right middle lobectomy was done. There was no episode of desaturation during the operation. Postoperatively, the child was put on elective ventilation and shifted to pediatric Intensive Care Unit. The child was extubated on the 2nd postoperative day, and intercostal drain was removed on the 3rd day. The child remained stable thereafter and was discharged in stable condition. Histopathology report was consistent with the diagnosis of CLE.

DISCUSSION

The etiology of CLE is unknown in half of the cases. It is speculated that defect in the bronchial cartilage tissue may lead to hyperinflation and air trapping on expiration due to a ball-valve effect. This causes distortion of the terminal airspaces leading to ineffective ventilation.[3] This has led to this condition being also known as congenital lobar hyperinflation. The hyperinflated lobe may compress on the surrounding lobes and may even cause mediastinal shift. This causes a decrease in functional lung tissue resulting in increased intrathoracic pressure and reduced respiratory reserve causing ventilation/perfusion mismatch with resultant hypoxia.[4] Some congenital cardiac malformations (patent ductus arteriosus, atrial septal defect, ventricular septal defect, total anomalous

pulmonary venous return, and Tetralogy of Fallot) have been reported with CLE.[5] Hislop and Reid added that an increase in the number of alveolar lobe (polyalveolar lobe) may play a role in CLE.[6]

CLE generally manifests as breathlessness, cough, wheezing, and respiratory distress in neonatal and early infancy. 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.[7] 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. Cataneo *et al.* in their 30-year case series of CLE mentioned a case in which CLE was misdiagnosed as tension pneumothorax at birth, and chest tube drainage was done, without any success.[8] On the other hand, in most cases such children may be misdiagnosed as pneumonia leading to unnecessary antibiotic treatment and loss of valuable time in diagnosis. As in our case, the child had multiple hospital admissions since birth and was being treated for pneumonia. CXR and CT scan showed hyperinflated right middle lobe compressing upon the neighboring lobes with collapse of the right upper zone and also mediastinal shift leading to the diagnosis of CLE.

Treatment of CLE is essentially surgical, i.e., lobectomy. Asymptomatic patients may be managed conservatively but should be kept in close follow-up as they may require surgical intervention in cases of worsening respiratory distress as was the scenario in our case. Early surgical intervention has been found to significantly improve the condition of the patient.[9,10] However, the number of lobes affected may also determine the outcome of surgery. 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.[11]

CONCLUSION

A strong clinical suspicion is required to diagnose this rare anomaly which may mimic other causes of respiratory distress. CT scan is a helpful adjunct in making the diagnosis. Early identification and surgery can lead to dramatic improvement in the clinical condition of the patient.

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Conflicts of interest
There are no conflicts of interest.

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