CONGENITAL LOBAR EMPHYSEMA. REPORT OF THREE CASES

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ABSTRACT

Congenital lobar emphysema is a rare cause of respiratory distress during infancy which is cured by surgery. We are reporting three cases of congenital lobar emphysema with different presentations of the disease. Chest x-ray was the basis of diagnosis but confirmatory lung computerized tomography was also used. The emphysematous lobe was left upper lobe in two infants and right upper and middle lobes in the third one. Resection of the affected lobes was performed with good results.

Key words: Congenital Lobar Emphysema, Left Upper Lobe, Computerized Tomography, Chest x-ray.

INTRODUCTION

Abnormal lobar distention can produce subtle or gross respiratory distress in normal newborn or infant(1). In this report three cases of congenital lobar emphysema (CLE) are being presented with specific attention to clinical presentation, diagnosis and treatment.

CASE 1

An 11 months old male infant was brought with tachypnea, retraction and wheezing. He had history of chronic cough and repeated pneumonia associated with wheezing and respiratory distress since 7 months of age. Chest x-ray (CXR) showed left upper lobe (LUL) emphysema, shift of the mediastinum to the right side and paracardiac infiltration (figure 1).

Bronchoscopy ruled out foreign body aspiration. High resolution computarized tomography (CT) of the chest revealed overinflated left upper lobe (LUL). He underwent surgery and the emphysematous lobe was resected. At the present time he is well and does not show any symptoms.

CASE 2

A 4.5 months old male infant who had been well up to a few days prior to referral, presented with severe respiratory distress and occasional wheezing. He had been admitted in paediatric intensive care unit due to severe respiratory distress and had been taken care of with mechanical ventilation. CXR revealed overinflated LUL, shift of the mediastinum, atelectasis of right upper lobe and paracardiac infiltration (figure 2). He responded well to supportive treatment but follow up CXR showed the LUL still overinflated. High resolution CT of

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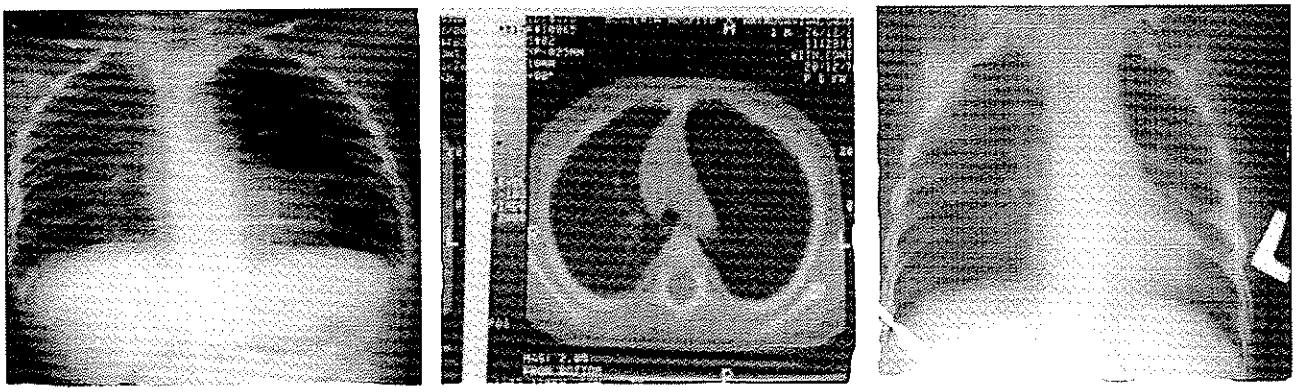

A B C

Figure 1. A: CXR of an 11 month old infant with congenital lobar emphysema of LUL.
B: CT of the same patient shows over inflated LUL.
C: CXR of the same patient after resection of the involved lobe

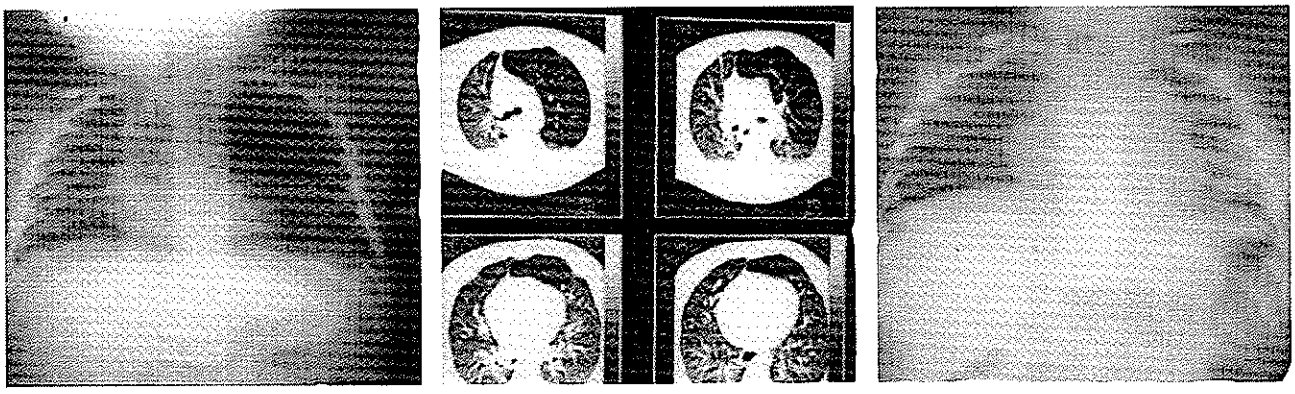

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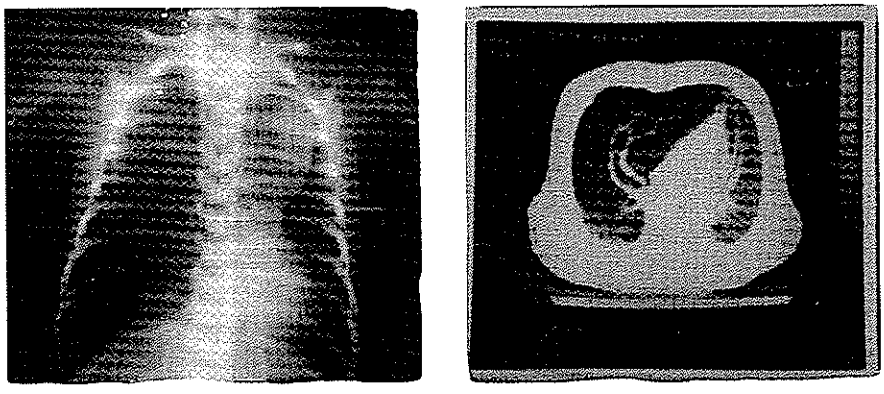
Figure 2. A: CXR of a 4.5 month old infant with severe respiratory distress.
B: CT of the same patient.
C: CXR after lobectomy.



A B

Figure 3. A: CXR of a 3 month old infant with respiratory distress. The mass which is seen in the left upper mediastinum disappeared after a short course of steroid therapy and turned out to be thymus.
B: Lung CT of the same patient, which shows emphysematous right lung.

the lungs showed overinflation of LUL which confirmed the diagnosis of CLE. The affected lobe was resected with a good outcome.

CASE 3

A three month old male infant was referred with episodes of respiatory distress since birth. His general condition was fair but had tachypnea, chest retractions and rales on auscultation. Chest radiography showed a mediastinal mass, shift of the mediastinum to the left side, hyperareation and infiltration of the right lung (figure 3). High resolution CT of the lungs was in favour of emphysema of the right lung. He had partial improvement after antibiotic therapy and the thymic shadow which appeared as mediastinal mass disappeared after a short course of steroid. He had an operation as a case of CLE and the emphysematous right middle and upper lobes were found and resected. He is well in his follow ups.

DISCUSSION

CLE accounts for about 15% of congenital bronchopulmonary malformation(1). It is more common in the boys. The disease is usually unilobar and the LUL is the most common site followed by right middle and upper lobes. The reports of congenital lung lesions, including 10 cases of CLE, revealed that the most common site of involvement was the right lung(2). Except for a few reported cases in older children and adults, all infants with CLE were less than 12 months of age at the onset of the first symptoms (3,4). They usually present in the first 4 months of life, often in the newborn period, with respiratory distress or poor feeding. Occasionally the presentation is more insidious, consisting of tachypnea, dyspnea, wheezing, cough, poor feeding, intermittent cyasnosis or exercise intolerance. In some patients CLE is identified incidentally during routine health care(1). At least 10% of patients also have congenital heart disease and a larger percentage have other anomalies(5) but none of our three patients had any associated anomaly.

The eitology of CLE is unknown. The current explanation for CLE involves partial bronchial obstruction or intrinsic alveolar disease. The bronchial obstruction can be due to complete or partial absence of cartilage, bronchomalacia, mucosal folds and extrinsic vascular compression (1,2).

Antero posterior CXR shows a large hyperlucent area containing vague lung and bronchovascular markings. Emphysematous lobe produces ipsilateral lobar atelectasis, diaphragmatic compression, mediastinal shift and contralateral lung atelectasis. On the lateral view, a translucent anterior mediastinum is suggestive of lung herniation.

In the typical setting of an infant under 4 months of age with characteristic clinical feature and CXR, no further work up is needed. In the older child with an atypical presentation, a CT scan may be useful in delineating the lesion. Ventilation perfusion scan can assess the function of the involved lobe and in some cases it is used for initial evaluation and follow up(6). Bronchoscopy usually is not necessary and may be life threatening. This procedure can be used if a foreign body is a possibility and may be warranted in children whose first symptoms occur after 6 months of age. Bronchoscopic appearance may be helpful in finding the specific cause, such as bronchomalacia (7).

Surgical therapy, is the treatment of choice when the diagnosis is accompanied by symptoms(2,4,5,8). Newborn presenting with severe respiratory distress requires immediate surgical excision(1). The operative mortality rate is low even in the face of early age, concomittant congenital heart disease or severe respiratory symptoms(8). However, recent reports demonstrate that children with mild or absent symptoms can be managed medically without significant progression of their respiratiory symptoms(1,6).

A controversy exists regarding the need for routine resection of CLE in asymptomatic or mildly affected patients, however early diagnosis and prompt surgical resection in infant with respiratory distress remains the primary approach to optimum outcome.

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