Case Report

Congenital Lobar Emphysema: A Misleading Entity

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Abstract

Congenital lobar emphysema is a rare disorder of unknown aetiology that may present a diagnostic challenge. We report two cases of congenital lobar emphysema in paediatric age group who were initially treated as pleuro-pulmonary tuberculosis and recurrent pneumonias, respectively. Eventually they were diagnosed as congenital lobar emphysema following chest radiography, computed tomography and bronchoscopic evaluation. **[Indian J Chest Dis Allied Sci 2019;61:207-209]**

Key words: Lobar emphysema, Unilateral hyperlucency, Lung.

Introduction

Congenital lobar emphysema (CLE) is a developmental anomaly of the lower respiratory tract that is characterised by marked over distension of one or more of the pulmonary lobes.[1] Other terms used for CLE are congenital lobar over inflation and infantile lobar emphysema.[2] The CLE is an uncommon congenital anomaly of the respiratory system with an incidence estimated to be one in 70,000 to 90,000.[3] Males appear to be affected more than females, with a ratio of 3:1.[1] Definite cause of the overinflation of the lobe is not identified in majority of the cases, however most frequently identified cause remains the obstruction of the developing airways.[1] We hereby describe two cases of congenital lobar emphysema detected in toddlers, which were being managed as pleuro-pulmonary tuberculosis and recurrent pneumonias in peripheral care facilities.

Case Reports

Case 1

A 3-year-old male delivered at full-term by normal vaginal delivery, breast-fed baby, resident of rural India, initially became symptomatic at the age of four months with cough with expectoration, fever and breathlessness on exertion. Initial chest radiograph showed hyperlucency of the left hemithorax which was probably mis-diagnosed as pneumothorax (left) and he was put on intercostal chest tube drainage for seven days, but without any radiological improvement. He was also given a short course of oral antibiotics, to which he responded partially. At the age of one year he was again symptomatic with similar complaints. He was suspected to be a case of pleuro-pulmonary tuberculosis and anti-tuberculosis therapy was administered empirically for the total duration of six months (2HRZE+4HR). However, he continued to remain symptomatic in-between with complaints of recurrent episodes of breathlessness, fever and productive cough. He was being treated symptomatically by the general practioners, without reaching a definite diagnosis. The patient was brought to tertiary care hospital with similar complaints. On examination, he was 36 inches tall, with weight of 10kg, systemic examination revealed pectusexacavatum, hyperresonance over the left infra-clavicular, mammary area, obliterated cardiac dullness and reduced breath sounds over the above-mentioned regions. Routine haemotological and biochemical parameters were within normal limits. Chest radiograph showed marked over distension of the left upper lobe with a mediastinal shift to the right and collapse of the ipsilateral remaining lung field (Figure 1). Computed tomography (CT) showed a hyperlucent, hyper-extended lobe (attenuated but intact pattern of organised vascularity) with midline herniation, compression of the remaining lung and the mediastinum significantly shifted away from the side of the abnormal

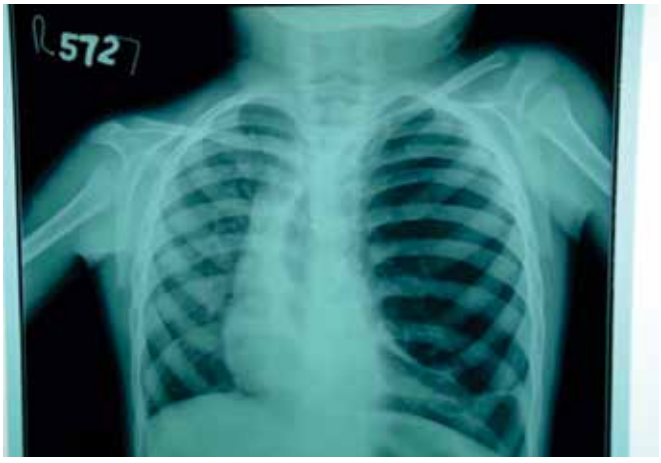

Figure 1. Chest radiograph (postero-anterior view) showing hyperinflation of the left upper lobe with shifting of the mediastinum to the right.

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lobe (Figure 2). Fibreoptic bronchoscopy revealed no obstruction with normal tracheobronchial tree. Based on the clinical and radiological profile, he was diagnosed as a case of congenital lobar emphysema and was treated conservatively, with close follow-up in the out-patient department.

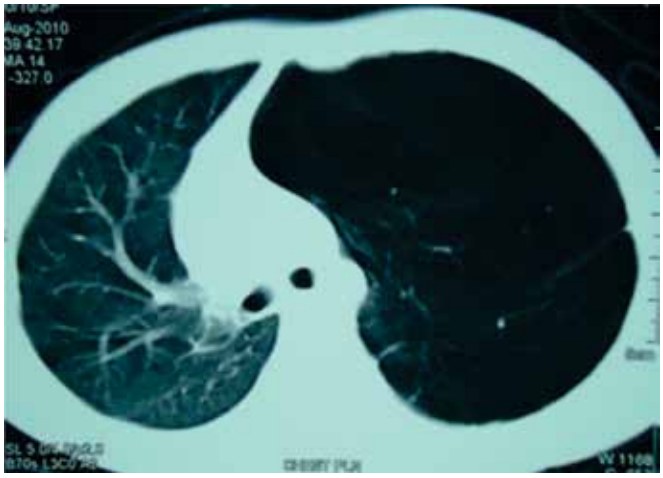

Figure 2. Computed tomography showing, marked over inflation of the left upper lobe with hyperlucensy, preserved vasculature, shifting of the mediastinum to the right side and compression of the right lung.

Case 2

A 2-year-old female toddler born at full-term by caesarean section was symptomatic since two months of age with complaints of cough and difficulty in breathing. She was diagnosed as a case of pneumonia and was prescribed oral antibiotics by the local practioner. Thereafter parents brought the child to us. Chest radiograph revealed hyperinflated left lung with mediastinal shift to the right and CT showed hyperlucent left lung with contralateral mediastinal shift (Figure 3). Fibreoptic bronchoscopy detected normal tracheobronchial tree with no endobronchial obstruction. Consequently, she was diagnosed as case of congenital lobar emphysema and was advised left upper lobe lobectomy in view of the symptomatology.

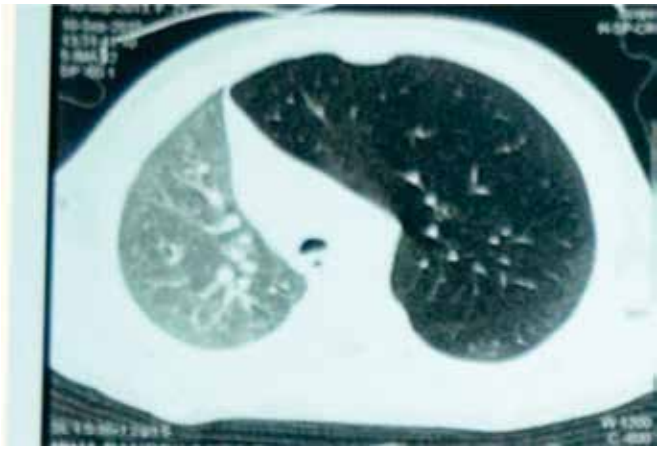

Figure 3. Computed tomography showing hyperlucent left lung with contralateral mediastinal shift.

Discussion

Congenital lobar emphysema is a developmental anomaly which is seen almost always unilateral and there is distinct predilection of the left upper lobe.[4] Progressive lobar hyperinflation is likely the final common pathway that results from a variety of disruptions in bronchopulmonary development.[1] Airway obstruction can be due to intra-mural or extra-mural causes, with the former being more common. Among the intra-mural causes, bronchial cartilage abnormalities and mucosal folds are the most common abnormalities.[5] Extra-mural compression of airways most commonly occur from anomalous vessel.[4] 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. Patients often present within the first six months of life with recurrent respiratory distress. Chest radiograph and CT of thorax are diagnostic and show the hyperlucent affected lobe with herniation of the lobe to the opposite side, shifting of the mediastinum to the opposite side and collapse of the remaining part of the ipsilateral lung.[4] Concomitant congenital heart disease (CHD) is seen in 12% to 20% cases of congenital lobar emphysema, which includes patent ductus arteriosus (PDA), atrial septal defect and ventricular septal defect. The presence of concomitant CHD should be kept in mind in infants having unusual respiratory distress symptom.[6]

Our patient (Case 1) presented with all the symptoms characteristic of congenital lobar emphysema, which was initially diagnosed wrongly as secondary pneumothorax. With the availability of appropriate imaging facilities diagnosis of congenital lobar emphysema was made. Controversy exists regarding surgical and conservative management of this malformation. There is no consensus opinion. However, management of congenital lobar emphysema has traditionally been surgical.[7] With the increased use of imaging, this lesion is frequently found in asymptomatic and mildly symptomatic children, prompting us to adopt a more conservative approach to these children.[8] Our patient was mildly symptomatic with the episodic cough, fever and breathlessness and there was resolution of symptoms on administration of short course of antibiotics. In view of his stable condition without any associated complications he was treated conservatively and kept on follow up. However, if the child is frequently symptomatic even after the introduction of antibiotics and vaccination, he/she should be advised to undergo surgical resection as was in Case 2.

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