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Case Report

Case series: unusual presentations of congenital lung malformations

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ABSTRACT

Congenital Lung Malformations (CLM) are a group of rare lung abnormalities affecting the airways, parenchyma, and vasculature. Congenital lung malformations encompass several conditions such as Congenital Cystic Adenomatoid Malformation (CCAM), Congenital Lobar Emphysema (CLE), Diaphragmatic hernia, sequestration of lung. A high index of suspicion is necessary to diagnose CLM in children irrespective of the age. Here we present a series of three cases of congenital lung malformations with unusual presentations.

Keywords: Congenital, Lung, Malformations, Infancy

INTRODUCTION

Congenital Lung Malformations (CLM) are a group of rare lung abnormalities affecting the airways, parenchyma, and vasculature. They represent a spectrum of abnormal development rather than discrete pathological entities.[1] They are caused by aberrant embryological lung development which occurs at different stages of intrauterine life. Congenital lung malformations encompass several conditions such as Congenital Cystic Adenomatoid Malformation (CCAM), Congenital Lobar Emphysema (CLE), Diaphragmatic hernia, sequestration of lung.

These lung congenital lung malformations usually present in the early neonatal period with respiratory distress due to a mass effect, while rarely others may be asymptomatic only to be detected in later life on CXRs performed for other reasons. Certain forms of CLM have the potential to undergo malignant change. Diagnosis of congenital lung malformations requires careful examination and high index of suspicion along with radiological modalities to confirm the diagnosis.

CASE REPORT

Case 1

A 2.5 year old female child, fifth born to a third degree consanguineous marriage was admitted with complaints of cough and fever since 20 days which responded poorly to antibiotics .Cough was non-productive and gradually progressive. Neonatal period was uneventful and there was no history of similar complaints in the past and no history of contact with tuberculosis. Right hemithorax was dull to percussion and had features of volume loss with crackles all over. Clinical impression was right sided collapse/fibrosis. Chest X-ray showed right sided heterogenous opacity with ipsilateral mediastinal shift. Ultrasound chest revealed right basal lung consolidation with air bronchogram. Flexible fiberoptic bronchoscopy did not show any intra bronchial obstruction. Hemogram showed anemia with polymorpho-nuclear leukocytosis. CT chest showed a large thin walled cavity in the right upper and middle lobe measuring 80 x 54 mm suggestive of congenital cystic adenomatoid malformation. Few septae were seen with in the cavity. Adjacent lung was

collapsed due compression by the large cavity within the right lung.

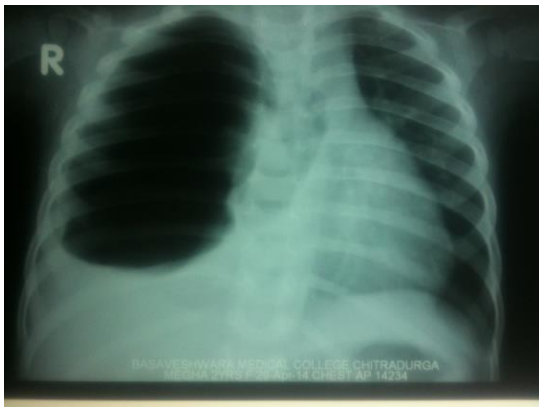

Figure 1: Chest X-ray CCAM.

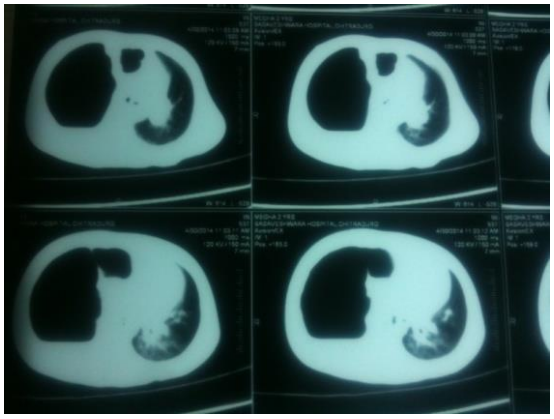

Figure 2: CT scan showing cystic lesion of right lung.

Case 2

A 5 month old female child was admitted in our PICU in view of fever, cough and respiratory distress which has been gradually increasing since 5 days. She gives history of recurrent bouts of similar episodes. There was no history of cyanosis or seizures. Development was appropriate for age and immunization status was upto date. The child was born to a non-consanguineous marriage. The antenatal period was uneventful. The child was being exclusively breast fed. On physical examination, the patient was febrile (T= 38°C, axillary) and tachypneic (RR = 70 BPM) with, subcostal and intercostals retractions, nasal flaring, and decreased breathing sounds in right hemithorax. A hyperresonant note was appreciated on the right side on percussion. The Trachea was shifted to the left. Other systems appeared normal.

Chest X-ray showed marked overdistension of the the right upper lobe with mediastinal shift to the left and collapse of the ipsilateral remaining lung field (Figure 4). CT chest showed features consistent with congenital lobar emphysema.

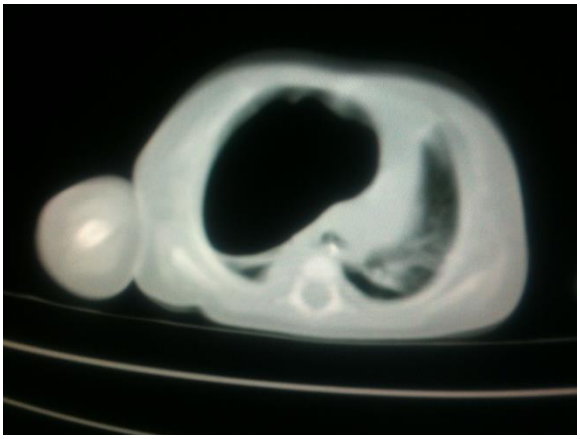

Figure 3: CT scan showing CLE.

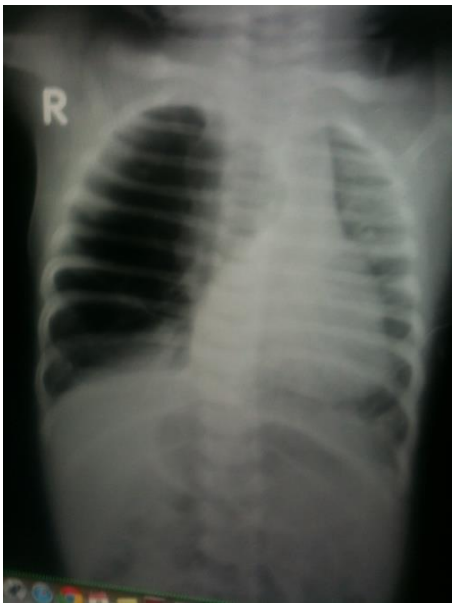

Figure 4: Chest X-ray showing CLE.

Case 3

A 3 month old child was admitted in our ward with complaints of recurrent episodes of respiratory tract infections since 2 weeks of life and complaints of poor feeding. Neonatal period was uneventful and no history of hospitalizations thereafter. Family history was noncontributory. Child was febrile, tachypnoeic (RR = 60/min) with subcostal retractions. Chest was bilaterally symmetrical and the trachea and mediastinum was pushed to the right side. Percussion note was equally resonant on both sides but air entry was decreased on the left side both anteriorly and posteriorly. On subsequent examination bowel sounds were heard over the left hemithorax which was initially missed.

Chest X-ray showed bowel loops herniating into the left thoracic cavity. Ultrasonography was suggestive of diaphragmatic hernia and the left diaphragm was not visualized.

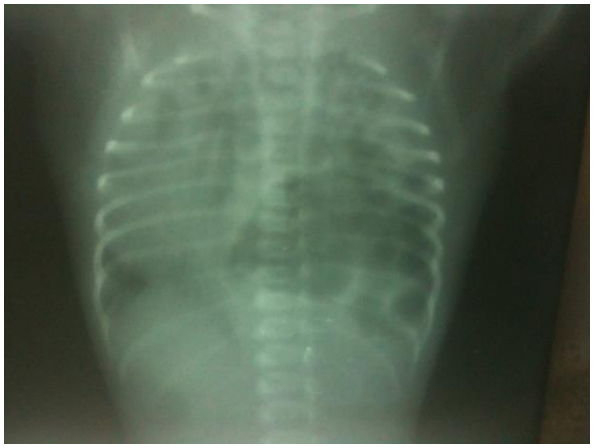

Figure 5: Chest X-ray showing congenital diaphragmatic hernia.

DISCUSSION

Congenital lung malformations are uncommon and depending on the size of the lesion and degree of functional impairment, these may lead to considerable morbidity and mortality. The incidence of congenital lung malformations is around 5000-1000 live births.[1] Surgery, either open thoracotomy or video-assisted thoracoscopy is the usual modality for management.[1,7,8]

CAM was first acknowledged into English medical literature by Ch'in and Tang in 1949.[3] Congenital Cystic Adenomatoid Malformation (CCAM) of the lung is caused by anomalous fetal development of terminal respiratory structures, resulting in adenomatoid proliferation of bronchiolar elements and cyst formation leading to enlargement of the affected lobe. There is a predilection of the right lung over the left for this anomaly.[4] Single lobe involvement is the most common. CCAM usually presents with respiratory distress in the neonatal period. 80% of the cases are diagnosed by one month of age.[5,6]

The features unique in our case is the late age of presentation and the involvement of both upper and middle lobes.

Gross and Lewis first reported CLE in 1954.[7] It can affect any lobe but being more common in the left upper lobe followed by the right middle and right upper lobes. It has been proposed that CLE is caused by air trapping in expiration due to bronchial collapse which is the result of aplasia or hypoplasia of major and branch bronchial cartilage rings and this theory is most accepted. It has two forms: Hypoalveolar in which alveoli are fewer in number than expected and Polyalveolar have greater number of alveoli than expected.[10,11]

Features unique in our case is the presentation at 5 months of age and the involvement of right upper lobe.

Congenital diaphragmatic Bochdaleck hernia is an anatomical defect of the diaphragm, which allows protrusion of abdominal viscera into the chest, causing serious pulmonary and cardiac complications in the neonate. Congenital Diaphragmatic Hernia (CDH) usually on the left hemidiafragm, caused by a lack of closure of the pleuroperitoneal canal between the eighth and tenth week of fetal life during the embryonic development.[12] It typically presents in the neonatal period with severe respiratory failure.Most cases of CDH are diagnosed antenataly or in the first few days of life.[12,13]

The presentation of CDH at 3 months of age is the unique feature in this particular case.

The purpose of this article is to highlight the importance of being extra-vigilant and actively seeking alternative diagnoses in children who present as infective chest conditions unresponsive to antibiotics irrespective of the age of the child. They should undergo appropriate imaging studies to diagnose congenital lung malformations. Once diagnosed, these malformations are treatable by surgical interventions and the long term prognosis is usually good depending upon the extent of involvement. Early intervention provides a better prognosis.

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