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Case Report

Congenital cystic disorders of lung masquerading as complicated pneumonia and pneumothorax: A case series

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Abstract. Congenital cystic adenomatoid malformation and congenital lobar emphysema are rare embryonic congenital disorders of lung. We report seven such cases of congenital cystic pulmonary disorders out of which, four cases were of congenital cystic adenomatoid malformation and three cases of congenital lobar emphysema. All cases presented with recurrent or persistent pulmonary infection and pneumothorax. These cases had been masquerading as pneumonia and/ or its complication and were treated for it for a variable period of time before being diagnosed as congenital lung malformation after appropriate investigations. We discuss here these rare cases.

Key words: Congenital cystic adenomatoid malformation, congenital lobar emphysema, pneumonia, pneumothorax, congenital cystic disorders of lung

1. Introduction

Congenital cystic disorders of the lung are rare. The diagnosis of these conditions may be missed if not specifically thought of and looked for, especially in the setting of a developing country where infectious conditions of lungs are far more commoner. We present here a series of seven cases where congenital disorders of lungs were masquerading as various infectious conditions before a final definitive diagnosis was reached.

2. Case reports

Respiratory infections are the most common causes of morbidity and mortality in a developing country. We got seven cases over one year where the presenting features were either persistent or recurrent respiratory infections, but investigations revealed a congenital malformation of lungs. Out of these seven, four were diagnosed to be having congenital cystic adenomatoid malformation.

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There were three female and four male patients. All the cases were clinically suspected of having congenital cystic adenomatoid malformation (CCAM) and three having congenital lobar emphysema (CLE). The age of presentation varied from 6 days to 10 months. congenital lung malformation when they did not respond to adequate doses of antibiotics and/or chest tube drainage both. The details of each patient are enumerated in the following table (Table 1). After diagnosis, all the cases were referred to Department of Cardio-thoracic Surgery for definitive operative management.

3. Discussion

We have reported 7 cases of congenital cystic disorders of lungs, of which 4 were CCAM and 3 were diagnosed to be having CLE.The review by Schwartz and Ramachandran (1) showed seventy cases of congenital disorders of lung occurring in a period of 25 years. Among them, ten cases were of CLE and five were of CCAM.

In this series, most of the cases presented with features of persistent/ recurrent or unresolving pneumonia. Even the chest X-rays were also indistinguishable from pyo-pneumothorax or pneumonia.

Table 1. (Description of the Cases)

Age/ Sex Onset of Symptoms	Symptomatology on admission	Clinical course before admission	Investigations Final diagnosis
Case 1			
6 mo/F 4 mo	Respiratory distress, fever off and on. Breath sound was diminished over left hemithorax, left infra-clavicular part was dull to percussion. Chest tube was in situ on left side.	Chest X-ray showed left sided pneumothorax with pneumonia	HRCT features suggestive of CCAM causing shifting of mediastinum.
Case 2			
10 mo/M 5 mo	High fever, respiratory distress, moderate growth failure. Tachypnea, breath sound diminished bilaterally in lower zones. Crackles in both lungs.	Chest X-ray showed ill-defined opacity in both lower zones.	CECT scan of thorax showed irregular cystic lesion mixed with alveolitis in whole left lobe, lateral segment of left lower lobe, posterior segment of right upper lobes and bilateral lower lobar collapse consolidation suggestive of CCAM with infection
Case 3			
I mo/M since birth	Respiratory distress, difficulty in feeding, irregular fever. Air entry was reduced in both lung fields, crackles were heard bilaterally (R>L).	Chest X-ray consistent with pneumothorax. Closed thoracotomy with water seal drainage was done.	HRCT scan of thorax showed, the lung parenchyma had been replaced by cystic cavities. Some showed fluid within them suggestive of CCAM
Case 4			
8 mo/M 4 mo	Respiratory distress for the last four months with intermittent fever. Tachypnea and tachycardia. Crackles were found in both lungs.	Chest X-ray revealed cystic areas in both lung fields. Recurrent pneumonia was treated with several courses of parenteral antibiotics	CECT scan of thorax showed irregular cystic lesion mixed with alveolitis in both lungs, with predominant involvement of bilateral lower lobes suggestive of CCAM
Case 5			
8 mo/F/ 6 days	Respiratory distress and fever for the last three months. Retarded growth. Air entry was reduced in eft lung and crackles were heard over left lung.	Chest X-ray revealed radiolucent area over whole left hemithorax and shift of mediastinum.Tube thoracostomy with water seal drainage had been done five times in previous two months	HRCT of thorax showed hyperinflated left upper lobe with herniation of lung to opposite side anteriorly. Mediastinum shifted towards right. There was also evidence of focal area of consolidation in both lower lobes posteriorly. Ground glass opacity was seen in both lower lobes. Vessels were attenuated in upper lobe. The features were suggestive of CLE of left upper lobe.
Case 6			
6 mo/F 4 mo	Respiratory distress Tachypnea, fever. Air entry was reduced over left lung and crackles were heard.	Chest X-ray showed that radiolucent area over whole left hemithorax and shift of mediastinum. The girl had been admitted elsewhere two months back and was treated with antibiotics for pneumonia.	HRCT of thorax showed hyperinflated left upper lobe with herniation of lung to opposite side anteriorly. Mediastinum shifted towards right. Ground glass opacity was seen in both lower lobes. The features were suggestive of CLE of left upper lobe.
Case 7			
4 mo/M since birth	Cough since birth. tachypneic, intermittent fever. Air entry was reduced in right lung.	Admitted in this institute two months back as LRTI. Treated with various antibiotics. Chest X-ray showed that radiolucent area over whole right hemithorax and shift of mediastinum.	HRCT of thorax showed hyperinflated right upper lobe with herniation of lung to opposite side anteriorly. Mediastinum shifted towards left. The features were suggestive of CLE of right lung.

CECT: Contrast enhanced computed tomography
HRCT: High resolution computed tomography
LRTI: Lower respiratory tract infection

So there is a high possibility that the diagnosis of congenital lung malformations would be missed, unless specifically kept in mind and sought for by advanced imaging studies as pneumonia is the most common cause of respiratory morbidity in children, especially in developing countries (2).

CAM was first acknowledged into English medical literature by Ch'in and Tang in 1949 (3). A classification system was proposed by Stocker et al. who classified CCAM into three types based on histopathology (4).Congenital cystic adenomatoid malformation (CCAM) constitutes approximately 25% of congenital lung diseases (5). CCAM is caused by consequence of embryogenic insults before the 35th day of gestation and abnormal development occurs in terminal bronchiolar structure. Clinical presentations of CCAM are in neonatal period respiratory distress (80%) secondary to mass effect and pulmonary compression or hypoplasia and in severe case due to air trapping. Beyond neonatal period, CCAM presents with recurrent or persistent pneumonia, pneumothorax, and rarely haemopneumothorax (5-7).

Gross and Lewis first reported CLE in 1954 (7). It can affect any lobe but being more common in the left upper lobe (41%) followed by the right middle (34%) and right upper lobes (21%) Rarely more than one lobe are affected. Involvement of lower lobes is extremely rare. Out of the 3 cases of CLE in our series, 2 had involvement of left upper lobe and one was having CLE of right upper lobe.

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 (8-12)

So the purpose of this communication 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. They should undergo appropriate imaging studies to diagnose congenital lung malformations. Once diagnosed, these malformations are well treatable by surgical interventions. There is really a dearth of studies regarding the incidence and epidemiology of this condition in Indian and world literature barring a a few case series (12-14). CT scan of the chest is the diagnostic method of choice for these disorders (14).

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