ABSTRACT

Congenital cystic adenomatoid malformation (CCAM) is a rare developmental anomaly of lung characterized by the presence of abnormal bronchial structures of varying sizes or distribution. We report a case of CCAM in a fifteen month old previously asymptomatic girl child who presented as persistent lobar pneumonia.

Keywords: Congenital cystic adenomatoid malformation, Persistent pneumonia.

INTRODUCTION

Congenital cystic adenomatoid malformation (CCAM) is a rare embryonic congenital disorder of the lung. The diagnosis of this condition may be missed in a developing country where infective conditions of lung are much more common. We present a 15 month old girl child who presented with persistent lower lobe pneumonia. She needed multiple hospital admissions and repeated courses of antibiotics before a diagnosis was reached.

CASE REPORT

A fifteen months old previously asymptomatic girl was referred to our hospital with history of high grade intermittent fever of 21 days. She also had cough and breathing difficulty of 1 week duration. She was diagnosed as a case of right lower lobe consolidation and received multiple courses of intravenous antibiotics from other hospitals. She was referred to our hospital as she persisted to be symptomatic. At admission to our hospital the child was febrile, toxic and tachypnoeic. Oxygen saturation was 98% in room air. There was no tracheal or mediastinal shift. Clinical examination also showed reduced air entry, increased vocal fremitus and impaired resonance over the right infraaxillary, inter scapular and infrascapular areas. A high pitched bronchial breath sound was audible over the right interscapular area.

A clinical diagnosis of right lower lobe consolidation was made. Chest Xray showed opacities in the right lower lobe with multiple thick walled pneumatocoeles (Fig1). Investigations showed neutrophilic leucocytosis, microcytic hypochromic anaemia with raised inflammatory markers. She was started on injection Ceftriaxone, Vancomycin, nebulised bronchodilators and other supportive measures after collecting samples for relevant cultures. As the child was still symptomatic after 3 weeks of treatment, a CT Chest with contrast was done which was reported as breaking down consolidation with cavitatory changes in the right lower lobe. Mantoux and gastric aspirate for AFB were negative. An ECHO was done and vegetations were ruled out. The child improved symptomatically with treatment and became afebrile within 72 hours of initiating treatment. Blood and urine cultures were sterile. IV antibiotics were continued for 21 days as the possibility of staphylococcal pneumonia was considered. Chest Xray repeated after 3 weeks of treatment showed clearance of opacities but a large septated lesion persisted in the right lower lobe. Man- toux and gastric aspirate for AFB were negative. An ECHO was done and vegetations were ruled out. The child improved symptomatically with treatment and became afebrile within 72 hours of initiating treatment. Blood and urine cultures were sterile. IV antibiotics were continued for 21 days as the possibility of staphylococcal pneumonia was considered. Chest Xray repeated after 3 weeks of treatment showed clearance of opacities but a large septated lesion persisted in the right lower lobe (Figure 2).

At thoracotomy, pleura over the right interscapular area was considered. The child was taken up for surgery. At thoracotomy, pleura over the involved part of the lung was thickened. Dense adhesions were seen between the lung, diaphragm and pleura. A large thick walled cystic area was seen in the posterior lateral part of the right lower lobe. Right lower lobectomy was done. Histopathology of the resected lung showed a thick wall cyst lined with focal flattened epithelium resting on loose mesenchymal tissue consistent with Type IV CCAM (Figure 4). The immediate post operative period was uneventful. The patient is on regular follow up for the past five months and has remained asymptomatic.

DISCUSSION

Congenital cystic adenomatoid malformation of the lung is a rare developmental anomaly caused by abnormal fetal development of terminal respiratory structures resulting in adenomatoid proliferation of bronchiolar elements and cyst formation. Its exact pathogenesis is still uncertain. Studies have investigated role of HoxB5 gene and protein expression as well as other growth factors such as platelet derived growth factor-BB. CCAM was first acknowledged as a separate entity and introduced into English medical literature by Chin and Tang in 1949. A classification system was proposed by Stocker et
al who classified CCAM into three types based on clinical, gross pathological and histopathological features. Recently Stocker has added two more types to the existing classification based on anatomic and microscopic properties of pulmonary airway and has used the term congenital pulmonary airway malformation (CPAM) for the anomaly.

The left lung is involved as often as the right lung with single lobe disease observed four times more often than multi lobe disease. The clinical spectrum varies depending on the extend of malformation in the lung and the presence of associated conditions. In neonatal period they present as acute respiratory distress secondary to air trapping or because of mass effect and pulmonary compression or hypoplasia. It may remain asymptomatic and be discovered later in life on routine chest Xrays or present beyond neonatal period as recurrent or persistent pneumonia or pneumothorax. Malignant changes have also been reported later on in life.

CCAM should also be differentiated from other cystic lesions in the lung in children like pulmonary sequestration, bronchogenic cyst, congenital lobar emphysema, diaphragmatic hernia and cystic bronchiectasis. Chest radiography is essential in the work up of a child with suspected CCAM. Computed tomography of thorax provides a safe and rapid means of defining the extent of CCAM in all age groups. The typical appearance of CCAM is of a multilocular cystic lesion with thin walls surrounded by normal lung parenchyma. In this patient, persistent secondary infection complicated the appearance of the lesion.

The definitive treatment of CCAM is surgery. In lobectomy the remaining lung grows and expands well enough so that total lung volume and pulmonary function tests return to normal. Histopathology of the resected lung in this patient showed a thick walled cyst lined with flattened epithelium resting on loose mesenchymal tissue consistent with Type IV CCAM. Prenatal diagnosis of CCAM by ultrasound has improved the management of fetus as well as helped to define the natural history and pathophysiology of this malformation.

It is important to be extra vigilant and actively seek alternative diagnosis in children who present with persistent chest infections or recurrent chest infections involving the same lobe. Although rare it is important to recognize CCAM early in life so that appropriate surgical intervention can be done early thereby preventing the consequences of recurrent infection.
Figure 3- HRCT chest showing thick walled multi-septated cystic lesion in the lower lobe of the right lung.

Figure 4- Histopathology of the resected lung in this patient showed a thick walled cyst lined with flattened epithelium resting on loose mesenchymal tissue consistent with Type IV CCAM.

REFERENCES


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