

Congenital Cystic Adenomatous Malformation: A Case Report

Jaswir Singh

Department of Pediatrics, Government Medical College and Rajindra Hospital, Patiala, Punjab, India

Abstract: Congenital cystic adenomatoid malformation (CCAM) is a rare developmental abnormality of the lung. We present here a three year three month male child who was in perfect health, until he suffered from lung abscess followed by spontaneous pneumothorax as manifestations of CCAM of the lung. The characteristics of this case are discussed.

Key words: CCAM, Pneumothorax.

INTRODUCTION

Congenital cystic adenomatoid malformation (CCAM) is a rare abnormality of lung development. CCAM is cystic area within the lung that stems from abnormal embryogenesis¹. The fundamental pathological feature of the lesion is adenomatoid proliferation of bronchioles that form cysts at the expense of normal alveoli. CCAM is usually discovered in neonates because of respiratory distress and may occasionally be discovered in older children or adults who have recurrent infection².

Pathophysiologically large lesions may be associated with the development of hydrops fetalis in as many as 40% cases and is a poor prognostic sign. Hydrops is thought to arise from compression of the inferior vena cava.³ Polyhydramnios has also been associated with CCAM. This develops as a result of elevated intrathoracic pressure that leads to esophageal compression and the inability to swallow.

CCAM may remain undiagnosed until it is discovered as an incidental finding later in life; however, its usual postnatal presentation is respiratory distress in the newborn period. This may be due to pulmonary hypoplasia, mediastinal shift, spontaneous pneumothorax, and pleural effusions secondary to hydrops. Recurrent chest infections may be a feature later in life. A risk of malignant transformation in later years is noted.

CASE REPORT

A 3 year 3 months male child presented with fever, cough and nasal discharge for 10 days, height 84 cm and head circumference 45 cm. His heart rate was 110/min, respiratory rate 38 per minute and Blood pressure was 90/64 mm hg at the time of admission. On examination, pallor was present, breath sounds were decreased and percussion note was dull in right inframammary region. Hb 7g%, TLC 20400/mm³; DLC P80 L18M2E0; PBF neutrophilic leukocytosis. At the time of admission, X-ray chest showed right lower zone lung abscess with adjacent consolidation (Fig.1). Further X-rays showed right pneumothorax with collapse consolidation tight lung with right lung abscess. CT Chest showed pneumothorax with collapse consolidation right lower lobe with multiple thin walled cavities in right lower lobe suggestive of CCAM (Fig. 2). Pleural tap showed growth of staphylococcus with TLC 22500/mm³ P70% L30%.

Injection ceftriaxone and Vancomycin were given initially followed by Injection Ampicillin & injection Amikacin after pleural tap c/s report. Intercostals chest tube drainage was done and patient was referred to higher center for further management/pneumonectomy.

DISCUSSION

CCAM is a congenital condition. Cases are typically identified by routine ultrasonography screening. Most postnatally identified cases present in the newborn period. CCAM may present in the older child and adult as an incidental finding or secondary to repeated infection.

Child may present with respiratory distress, recurrent infection, hemoptysis, dyspnea, chest pain, cough, fever, failure to thrive and

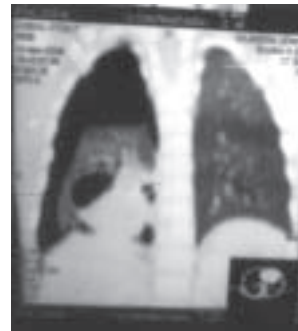


Fig. 1



Fig. 2

on examination tachypnea, pneumothorax, cyanosis, accessory muscle use, grunting may be present.

Complications like fetal death, premature delivery, pneumothorax, pulmonary hypertension, recurrent pneumonia, hemothorax, malignant change can occur.

Three types have been described. Type-I consists of a single cyst or multiple large cysts, lined by pseudo stratified ciliated columnar epithelium. The cyst walls are thick and contain smooth muscle and elastic tissue. Cartilage is rarely present. Normal-appearing alveoli are found between these cysts. Type-II contains numerous smaller cysts, usually less than 1 cm in diameter. Large alveolus-like structures are present between the cysts. The types-III forms a solid and bulky mass, composed of regularly spaced bronchiole-like structures separated by masses of alveolus-like structures lined by cuboidal epithelium.

Pulmonary resection during infancy is associated with low morbidity and mortality rates and may prevent the late complications of infection and occult malignant transformation⁵.

The treatment of CCAM is always surgical as soon as the diagnosis is made. Thoracotomy and delivery of the hyperinflated lobe into the wound brings immediate relief of the ventilatory and circulatory problems. Lobectomy is usually necessary, but segmental resection is occasionally feasible.

There is agreement amongst surgeons regarding the treatment of symptomatic patients; however, controversy exists about the management of asymptomatic neonates and infants with CCAM with respect to the decision and timing of excision.

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Correspondence: Dr. Jaswir Singh, Associate Professor, 5084, Urban Estate-I, 26 Acve Area ITBP wall, Patiala, Punjab-147001, India