Congenital Cystic Adenomatoid Malformation of Lung in an Infant: A Case Report

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Abstract

This case report is of an infant born with a rare lung abnormality, Cystic Adenomatoid Malformation (CCAM). CCAM is a rare congenital lung malformation (incidence) representing 25% of congenital malformations and 95% of congenital lung lesions.(1,2) CCAM may be diagnosed in-utero by ultrasound examination, or after birth with the presentation of respiratory distress like symptoms. Differential diagnosis includes diaphragmatic hernia, pulmonary sequestration, bronchogenic cysts, and congenital lobar emphysema. Because most CCAM lesions are manageable with the proper assessment, diagnosis, and interventions. We report here a case of 45 day-old-male baby with features of respiratory distress since birth and X-ray chest suggestive of large cystic lesion of left lung CCAM.

Key Words: *CCAM, Cystic lesion, Respiratory distress*

Introduction

Congenital Cystic Adenomatoid Malformation (CCAM) is a rare abnormality of lung development. CCAM is cystic area within the lung that stems from abnormal embryogenesis.[1] 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 infection.[2] Three histologic patterns have been described. Type 1 (50%) is macrocystic and consists of a single or several large (>2 cm in diameter) cysts lined with ciliated pseudostratified epithelium. The wall of the cyst contains smooth muscle cell and elastic tissue. One third of cases have mucus secreting cells. Cartilage is rarely seen in wall of cyst. This type has usually good prognosis for survival. Type 2 (40%) is microcystic and consists of multiple small cysts with histology similar to that of the type 1 lesion. Type 2 is associated with other congenital anomalies and carries a poor prognosis. In Type 3 (40%) the lesion is solid with bronchiole like structures lined with cuboidal ciliated epithelium and separated by areas of non-ciliated cuboidal epithelium. This lesion carries poorest prognosis and can be fatal. Prenatal ultrasonographic findings are classified as macrocystic (single or multiple cysts >5mm) or microcystic (echogenic cysts <5mm). The pathophysiologic effects of CCAM may be divided into prenatal and postnatal effects. 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 the amniotic fluid.[4] CCAM may remain undiagnosed until it is discovered as an incidental finding later in life, however it is usually detected postnataly and presents as 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.[5] A risk of malignant transformation in later year is also noted.[6]

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Case Report

A 45 days old male baby with weight was 3.4 kg, height 56 cm and head circumference 36.5 cm, presented with

chief complaints of fast breathing after 12 days of birth, which was gradual in onset and more during feeding and non-progressive in nature. There was no history of cyanosis, vomiting, decreased feeding or a suck rest suck cycle. There was no history of perinatal hypoxic insult, prolonged labour, meconium aspiration. Baby was admitted for three days to hospital and discharged on oral medications at that time and readmitted after one month for same complain. History of the mother was uneventful. On examination, baby had respiratory distress (Respiratory rate>70/minute) and air entry was significantly decreased on left side of chest. Percussion note was hyperresonant. X-ray chest (fig.1) was suggestive of preliminary impression of collapse lung like picture on right side with mediastinal shift to right side. Computerised tomography (CT) (fig. 2) chest was suggestive of cystic lesion in right lobe and was described by radiologist as congenital adenomatoid malformation Type 1. Patient was referred to Pediatric surgical care for further management.

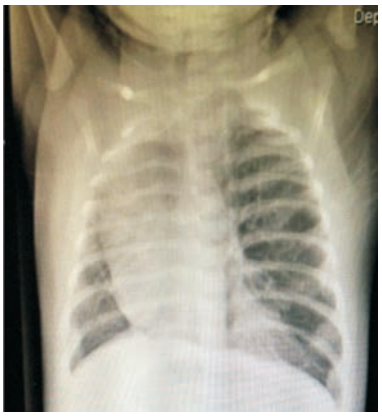

Fig. 1:

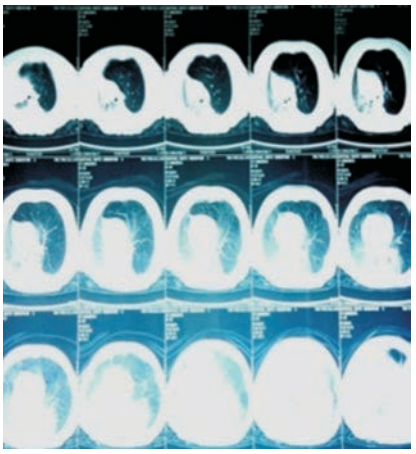

Fig. 2:

Discussion

Congenital cystic adenomatoid malformation is an uncommon congenital malformation of the lung that arises from excessive disorganised proliferation of tubular bronchial structures excluding the alveoli. The left lung is involved as often as the right lung with single lobe disease observed four times more often than multilobe disease. Cases are typically indentified prenatally by routine ultrasonography screening.[7] Most postnatally identified cases present in the newborn period. The most common mode of presentation is acute respiratory distress secondary to the cyst expanding and compressing its surrounding structures. The distress occurs through a ball-valve managed mechanism leading to air trapping. This mode of presentation is common during the neonatal period. Child may also present with recurrent infection, hemoptysis, dyspnea, chest pain, cough, fever, failure to thrive and on examination tachypnea, pneumothorax, cyanosis, accessory muscle use, grunting may be present. It may remain asymptomatic and be discovered later in life on routine chest films or present after the neonatal period as recurrent pneumonia.[8] CCAM may present in the older child and adults as an incidental finding or secondary to repeated infection.[9,10] Complications like fetal death, premature delivery, recurrent pneumonia, hemothorax, malignant change can occur. In imaging studies chest radiography, CT scanning, Magnetic Resonance Imaging (MRI), prenatal ultrasonography and renal, cerebral ultrasonography and echocardiography in newborns may be done as indicated. Pulmonary resection during infancy is associated with low morbidity. In lobectomy the remaining lung grows and expands well enough so that the total lung volume and pulmonary function tests return to normal. To conclude, early recognition and surgical treatment of CCAM is essential to prevent the consequences of recurrent pulmonary infections and the potential risk for malignant transformation. The treatment of CCAM is always surgical. 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.[11] There is agreement between surgeons regarding the treatment of symptomatic patients, but controversy exists about the management of asymptomatic neonates and infants with CCAM with respect to the decision and timing of surgery

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