

Congenital Cystic Adenomatoid Malformation of Lung: A Case Report

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Abstract: Congenital cystic adenomatoid malformation is a developmental, non-hereditary, hamartomatous abnormality of lung with unknown etiology. It is rare disease with an incidence of 1 in 25000 to 1 in 35000. It is disease of infancy with most of the cases diagnosed within the first two years of life. We report a case of type 1 CCAM in a 2 year old male with brief review of literature.

Key Words: Congenital cystic adenomatoid malformation, lung.

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I. Introduction

Congenital cystic adenomatoid malformations of lung are rare congenital cystic lung lesions that arise from excessive proliferation of tubular bronchial structures.¹ It is a rare lesion with incidence of 1 in 25000 to 1 in 35000 pregnancies and represents 25% of congenital lung malformations and 95% of congenital lung lesions.² It was classified into 3 subtypes in 19773. It was classified again by Stocker in 2002 into 5 types and renamed as congenital pulmonary airway malformation.

II. Case Report

A two years old boy complained of cough and fever on and off since 15 days for which the patient was admitted in our paediatric ward. Patient had multiple similar episodes in the past. Mother attended regular ANC checkup and antenatal USG was taken at 35 weeks of gestation which showed multiple cystic areas in right upper lobe of lung and large one measured 2.41cm (Fig 1). Patient was delivered by normal vaginal delivery and there was no any history of birth asphyxia, icterus, cyanosis and feeding difficulties. Patient had history of mild respiratory distress soon after birth. On examination, there was no pallor, no cyanosis and no clubbing seen, chest expansion was reduced on right side and on percussion hyper resonant sound was heard over right side. On auscultation, air entry was decreased on right side.

On further investigation, chest X- ray (PA view) showed multiple cystic lucencies in right lung field (Fig. 2). CT thorax revealed, multilocular, thin walled, variable sized (ranging from 3mm to 22mm) cystic lesions, surrounded by normal lung parenchyma (Fig 3, 4). All findings were suggestive of type 1 Congenital Cystic Adenomatoid Malformation (CCAM). Patient was treated conservative and referred to paediatric surgery and right upper lobectomy was done.

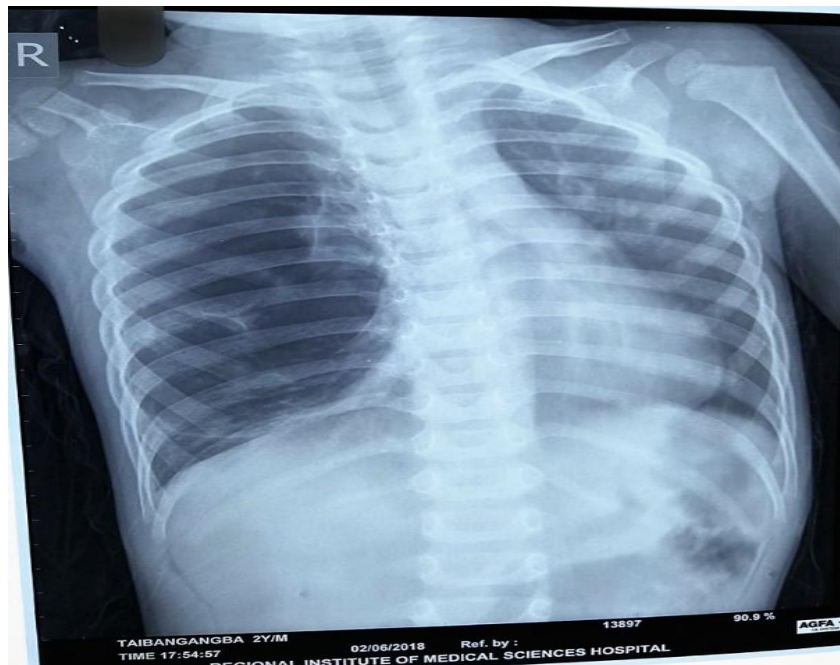
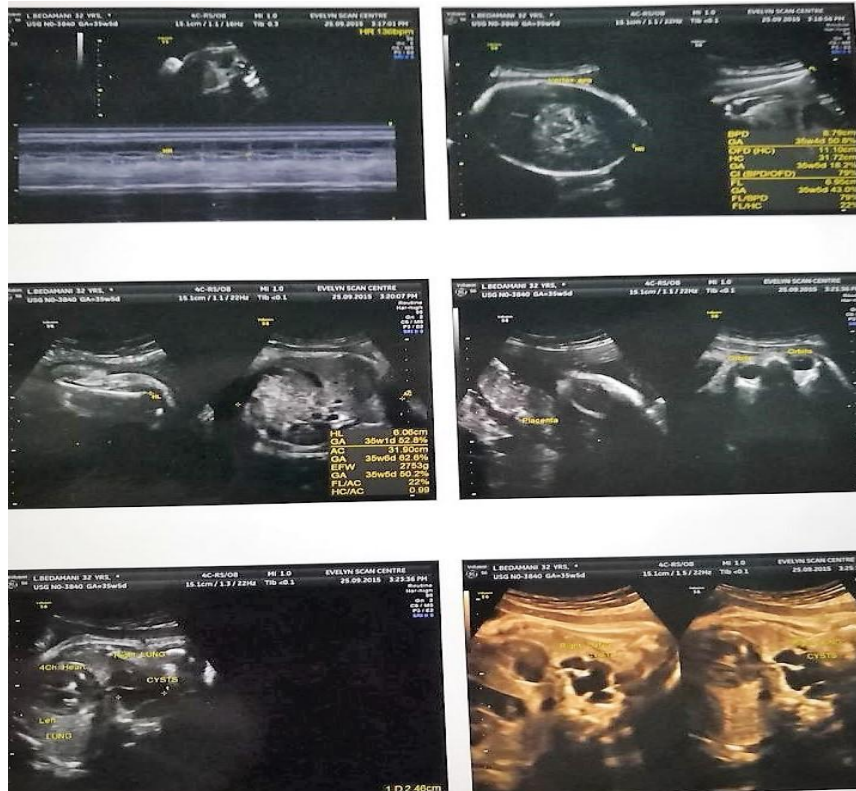




Figure3: CT thorax showing multilocular cyst

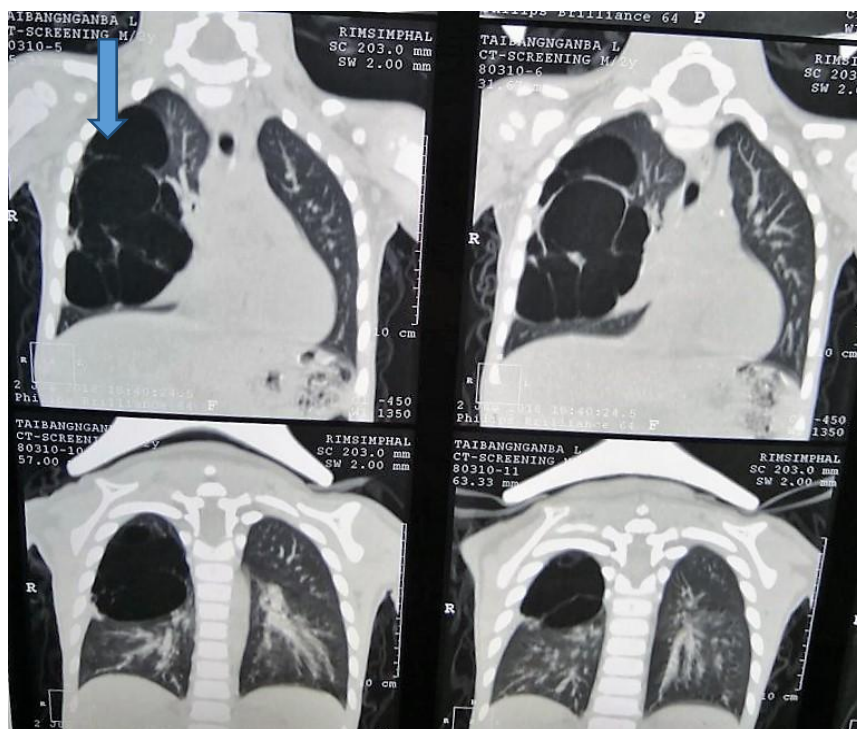


Figure 4: Multilocular cyst of different sizes

III. Discussion

CCAM is an unusual condition characterised by immature, malformed lung tissues with cystic appearance, which results from an abnormality of branching morphogenesis of the lungs. An adenomatous overgrowth of terminal bronchioles and alveoli leads ultimately to large masses, which communicate with the tracheobronchial tree and have feeding vascularisation from the pulmonary circulation.⁶

Stocker et al³ subdivided CCAM into three subtypes. Type 1 lesion constitutes 50-70% and is composed of single or large multiple cysts (>2cm) lined by flattened, cuboidal cells frequently producing mediastinal herniation. The walls of cysts contain prominent smooth muscle and elastic tissue. Mucus producing cells seen occasionally but presence of cartilage is extremely rare. Mucin production is unique to type 1 lesion. Type 2 lesions constitute 15-30% and are composed of multiple small cysts (<2cm), lined by ciliated cuboidal to columnar epithelium. Mucus cells and cartilage are not seen. This type is usually associated with systemic anomalies. Type 3 lesions constitute 5-10% and are usually large, bulky, non-cystic lesion producing mediastinal shift. Bronchial like structures are lined by ciliated cuboidal epithelium and separated by masses of alveolus sized structure lined by non-ciliated cuboidal epithelium.⁷ Systemic anomalies reported in type 2 are renal anomalies, abdominal wall defects, central nervous system defects, spinal deformities, gastrointestinal defects, cardiac anomalies, sirenomelia and anomalies of reproductive tract.⁸

In 2002, Stocker et al⁹ modified this classification by adding two more types (type 0 and 4) and renamed the lesion as CPAM, type 0 of tracheobronchial origin has solid appearance with small and firm lungs and microscopically shows bronchiolar type airway with cartilage, smooth muscle and glands separated by abundant mesenchymal tissue. Type 4 of distal acinar origin has peripheral cystic type, large cysts (>10 cm) lined by flattened epithelium and resting on loose mesenchymal tissue.¹⁰ The new classification is not much used as type 0 lesions are very difficult to differentiate from bronchogenic cyst and the similarities between type 4 and pleura pulmonary blastoma may cause problem in diagnosis.¹¹

CCAM is rare and usually presents before the age of 3 years. The exact incidence is not known. It is more common in boys than girls and it is usually unilateral. Patients can present with life threatening respiratory distress or a history of recurrent chest infections. Some cases are asymptomatic and are discovered as an incidental finding on radiography. Congenital lobar emphysema can present in a similar way and should be considered in a differential diagnosis. CCAM is best diagnosed with computed tomography.¹²

Classification based on prenatal ultrasound findings:

Microcystic lesions (<5mm): usually associated with fetal hydrops and has poor prognosis.

Macrocystic lesions (>5mm): not usually associated with hydrops and has favourable prognosis.¹³

The differential diagnosis includes lobar sequestration, bronchogenic cyst, congenital lobar emphysema, or congenital diaphragmatic hernia. Antenatally, it can lead to fetal hydrops and maternal polyhydramnios compromise. In adult patient, CCAM is a nidus for pneumonia, intralobar sequestration and development of bronchogenic carcinoma.^{14,15,16}

Serial antenatal sonographic evaluation, good obstetric care and delivery at a tertiary centre are preferred plan of treatment for antenatally detected cases. Postnatal and in adults patients, lobectomy is the treatment of choice for symptomatic cases.^{2,5,14,15}

Prognosis also depends on Stocker type, type 1 lesions carry overall good prognosis. In type 2 lesion, it is the associated anomalies that determine the prognosis. Type 3 lesions carry bad prognosis as they are usually large and present with cardiovascular compromise. Overall bilateral involvement, associated with hydrops and congenital anomalies carry poor prognosis.⁸

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