

Case Report

Spontaneous resolution of CCAM: a rare presentation

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ABSTRACT

Congenital cystic adenomatoid malformation (CCAM) is a rare developmental abnormality of lung occurring in 1-4/100000 live births. The mainstay of treatment is usually surgical excision as it can lead to recurrent pneumonias, abscess or malignancy. We here report a rare case of CCAM who presented at one year of age with right sided lung cysts and had favorable outcome as it resolved spontaneously.

Keywords: Congenital cystic adenomatoid malformations, Lung abscess, Respiratory distress

INTRODUCTION

CCAM is a rare hamartomatous cystic lesion of the lung.¹ It accounts for 25% of all congenital lung abnormalities and 95% of congenital cystic lung disease.² The incidence of prenatally diagnosed CCAM is 1: 25,000 – 35,000.²⁻⁵ CCAM may present in older children and adults as incidental finding secondary to repeated infections.^{3,5} Prenatally diagnosed cases mortality rate ranges from 9 to 49%.^{4,6} The diagnosis of CCAM can be made prenatally by ultrasonography and postnatally by radiography and MRI.

CCAM is subdivided into 3 major types:

- Type I lesions, the most common, are composed of one or more cysts measuring 2-10 cm in diameter. They are frequently lined by pseudostratified columnar epithelial cells, which occasionally produce mucin. Mucinogenic differentiation is unique to this subtype of CCAM.
- Type II lesions are characterized by small, relatively uniform cysts resembling bronchioles. They are lined by cuboid-to-columnar epithelium and have a thin fibromuscular wall. The cysts generally measure 0.5-2 cm in diameter.

- Type III lesions consist of microscopic, adenomatoid cysts and are grossly a solid mass without obvious cyst formation. Microscopic adenomatoid cysts are present.⁷

The mainstay of CCAM treatment is surgical excision (lobectomy)

that prevents complications such as recurrent infections, pneumothorax and malignancy.^{6,8} Cases complicated by pneumonia are treated by antibiotics and oxygen supplementation and sometimes mechanical ventilation.

CASE REPORT

Our patient 1-year old female child presented to us with fever for 7 days, cough and cold since 2 days, rapid breathing since 1 day prior to the admission. No significant or similar history in past. She was born of non-consanguineous marriage, an outcome of normal delivery to primi mother with no postnatal complications. She was immunized for age.

On examination, she was conscious, cooperative and alert with respiratory distress. She had tachypnea, sub costal retraction, with saturation 92% off oxygen. On

percussion dull note was heard on right side in subscapular region with corresponding air entry reduced. Vocal resonance was increased in right subscapular region. Thus, a working diagnosis of lobar pneumonia was made.



Figure 1: Chest X-ray showing cyst.

Blood investigations revealed neutrophilic leukocytosis with crp-238, X-ray chest showed consolidation in right lower zone. Ultrasonography was suggestive of right sided loculated pleural effusion with underlying consolidation. She was supported with oxygen and empirical treatment with antibiotics was started. However, she did not respond, hence an X-ray chest was repeated which showed multiple cystic lucencies in lower lobe of right lung (Figure 1).



Figure 2: HRCT showing cystic lucency in the right side.

At this stage, differential was kept as staphylococcal infection with bullae formation or congenital lung disorder. HRCT chest was done which revealed multilocular, thin walled, cystic lesions, surrounded by normal lung parenchyma. Few cysts showed air fluid level. Finding were suggestive of type 1 CCAM (Figure 2). Patient was advised lobectomy, but parents did not consent. The child was managed conservatively with 6 weeks therapy with higher antibiotics. Symptoms resolved, and repeat X-ray appeared normal. HRCT was done to confirm the X-ray findings, which in comparison with previous CT showed near complete resolution of cavitating/cystic lesions in the right lower lobe (Figure 3).



Figure 3: HRCT showing complete resolution of the right side.

DISCUSSION

W Congenital cystic adenomatoid malformation arises from excessive disorganized 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. In our case report only, left lung was involved. Cases are typically identified prenatally by routine ultrasonography screening. If missed prenatally, the commonest mode of presentation is acute respiratory distress due to expansion of the cyst which compresses its surrounding structures. Our patient also presented with similar complaints. The commonest mode of presentation is during the neonatal period, but our patient presented during infancy which a rare presentation. 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.⁷ It may remain asymptomatic and can be discovered as an incidental finding on routine X-ray chest. Complications like fetal death, premature delivery, recurrent pneumonia, hemothorax, malignant change can occur. In imaging studies chest radiography, CT scanning, MRI and prenatal ultrasonography may be done as indicated.⁴

Surgical intervention during infancy is associated with low morbidity and mortality rates and may prevent the late complications of infection or malignant transformation.⁶ In lobectomy the remaining lung grows and expands well enough so that the total lung volume and pulmonary function tests return to normal.⁸ Early recognition and surgical treatment of CCAM is essential to prevent the consequences of recurrent pulmonary infections. CCAM rarely resolve spontaneously, with only 4% not requiring any surgical intervention.

Our child is unique as she presented rather late and had a favorable outcome without surgery.

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