



## Persistent Respiratory Distress in Adolescent Girl: Unusual Cause

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### Abstract:

Congenital cystic adenomatoid malformations (CCAM) encompass rare developmental anomalies of lung that commonly present in infancy and early childhood, very rarely in adolescence with respiratory distress or recurrent pneumonia. We report a case of CCAM with pneumonia presenting first time at an age of 11 years.

**Key words:** Adolescent, Congenital Cystic Adenomatoid Malformation, Dyspnea, Pneumonia.

### Introduction

CCAM (congenital cystic adenomatoid malformation) or congenital pulmonary airway malformations represent rare spectrum of congenital lung abnormality. These result from aberrant lung development at 7<sup>th</sup> to 17<sup>th</sup> week of gestation resulting in cystic or solid overgrowth of tubular bronchiolar structure excluding alveoli. The incidence of CCAM (congenital cystic adenomatoid malformations) range between 1:8000 to 1:35,000 live births [1,2]. These anomalies are usually detected antenatally or postnatally in infancy and early childhood [2].

### Case Report

A 11 year female presented with complaints of intermittent fever, dry cough, chest pain for one month and fast breathing for 5 days. On examination there was no pallor, cyanosis, clubbing

or lymphadenopathy. Her chest examination revealed resonant note bilaterally with decrease air entry on right side, rhonchi and crepitations predominantly on right side. Her cardiovascular and abdominal examination revealed no abnormality. There was no history of similar illness, tuberculosis or hospitalisation in past. Her hemogram revealed haemoglobin: 14 gm%, total leukocyte count: 6300 mm<sup>3</sup>, and chest X-ray revealed thin walled lesion in right upper zone [Fig.1]. She received oxygen, 3<sup>rd</sup> generation cephalosporin and vancomycin. Her blood culture came sterile. She became afebrile on day 4 of hospitalisation but continued to have decreased breath sound on right side, so she was evaluated for other possible cause including tuberculosis, congenital lung malformation, and cavitary lung lesion. Her Mantoux was not reactive, sputum was AFB negative and sterile. Her CECT chest [Fig.2] revealed 4.8x2.9 cm thin walled cystic lesion

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in right upper anterior segment and subcentric cyst in posterior segment with peribronchial thickening. Subtle ground glass opacity were seen in right middle, posterior and left pericardiac and lingular regions. Cystic lesion suggesting possibility of CCAM type 1 with evidence of superadded infection was the final impression. She received antibiotics for 14 days and later referred for surgical consideration.

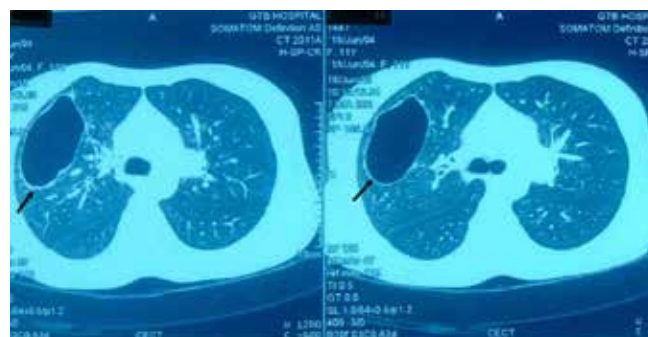
## Discussion

Congenital cystic adenomatoid malformation, a rare type of congenital lung malformation is result of overgrowth of lung tissue occurring at different stages of lung development. CCAM occurs due to developmental arrest at terminal bronchiolar level (pseudoglandular stage 7-17 week of gestation) followed by proliferation of bronchiole excluding mature alveoli and transformation into heterogeneous cystic or non-cystic areas which are supplied by pulmonary artery, rarely from systemic artery as in hybrid lesions. Based on embryological stage of insult, histology and dominant cyst size, these malformations have been classified into five types (type 0,1, 2, 3 and 4) [3]. Adzick *et al.* classified them as macrocystic type (cyst size 5 mm or more, type 1,2) and microcystic type (cyst size < 5 mm, type 3) on the basis of gross anatomy and ultrasound findings [4].

Type 1 lesions (cyst size more than 2 cm) are most common CCAM (60%-65%) and often occur unilaterally with equal incidence in male and female. Histologically type 1 cysts are lined by pseudo-stratified columnar epithelium and wall is made of fibro-elastic, muscular tissue or rarely has cartilage. Type 2 CCAM (0.5-2 cm) is associated with extrapulmonary malformation. Type 3 CCAM is usually solid and is microcystic lesion with bronchioalveolar morphology. Diagnosis can be made in antenatal period by ultrasound, Doppler or MRI and postnatally by chest X-ray or CT chest



**Fig.1:** X ray chest showing thin walled lesion in right upper lobe.



**Fig.2:** CECT chest: Thin walled cystic lesion in right upper anterior segment and subcentric cyst in posterior segment with peribronchial thickening suggestive of type I CCAM.

[3,4]. CECT of our case revealed type 1 CCAM with maximum width of dominant cyst 4.8 cm.

Complications of CCAM include polyhydramnios, hydrops, fetal death, recurrent pneumonia, haemoptysis, pneumothorax, and malignant transformation [3,4]. Commonly CCAMs present in infancy or in childhood. Small cystic malformation can regress in the third trimester [1]. Wong *et al.* [5] found that one patient (n=35) showed antenatal regression, 40% (n=35) of neonate were symptomatic at birth and 86% became symptomatic

at median age of two year (range 1 month-13 years). Our index case presented at 11 year of age. Rarely, they present first time in adolescence and adulthood as case of persistent pneumonia. Surgery is treatment of choice in these lesions.

## Conclusion

CCAM represent a rare spectrum of congenital lung malformation. Rarely they can have later age of presentation. Diagnosis is made by chest X-ray and HRCT chest and definitive management is surgery.

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