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# Congenital Cystic Adenomatoid Malformation involving right lower lobe of lung: A case report

Surgery

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## ABSTRACT

Congenital cystic adenomatoid malformation (CCAM) is a rare congenital hamartomatous malformation of lung with incidence 1 in 2500 to 1 in 3500 live births. In this case report, we discuss type 1 Congenital Cystic Adenomatoid Malformation in 5 year old male child who presented with complains of recurrent attack of cough and respiratory distress since 2 months after birth. Radiological examination confirmed the diagnosis. Patient was treated with lobectomy.Histopathological examination reported the diagnosis of type1 Congenital Cystic Adenomatoid Malformation.

## KEYWORDS:

Congenital cystic adenomatoid malformation, cystic appearance Microcystic lesions, Macrocystic lesions, lobectomy.

### INTRODUCTION

Congenital cystic adenomatoid malformation (CCAM) is a rare abnormality of lung development characterized by a cessation of normal bronchiolar maturation, resulting in cystic overgrowth of the terminal bronchioles that resemble fetal lung at 20 weeks gestation. It is a hamartomatous lesion in which there are multiple cystic masses within the lung that shoots from abnormal embryogenesis1

### CASE REPORT

We report a case of 5 year old male child presented with complains of recurrent attack of cough and respiratory distress which started 2 months after birth. Routine blood investigations were within normal range. X-ray chest showed multiple cystic lucencies in right lower lung field (Fig. 1). CT Scan chest revealed, multilocular, thin walled, cystic lesions, involving right lower lobe of lung largest measuring approximately 7×6.2cm ( fig. 2). Few cysts showed air fluid level with superadded infection. Findings were suggestive of type 1 Congenital Cystic Adenomatoid Malformation (CCAM). Patient was treated with right lower lobectomy as shown in fig 3. Postoperative X-ray shows expansion of the right lung ( fig.4) Histopathology report of resected lobe of lung showed the dagnosis of type 1 CCAM.

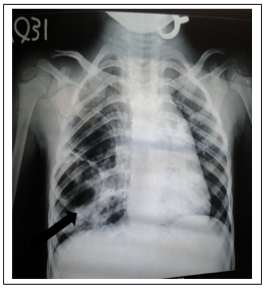


Fig 1

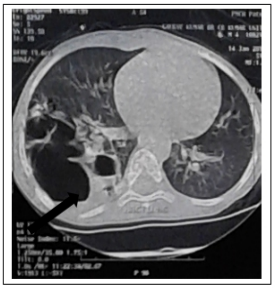

Fig 2

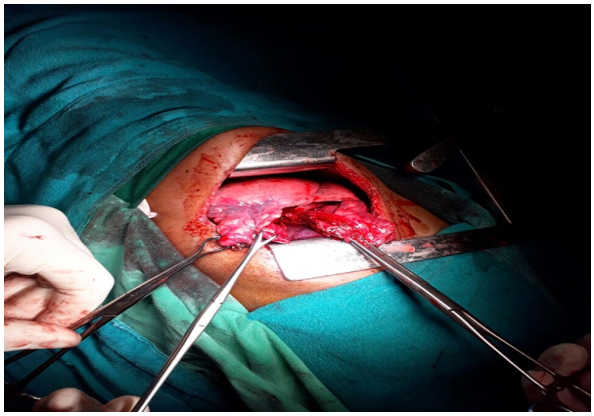

Fig 3

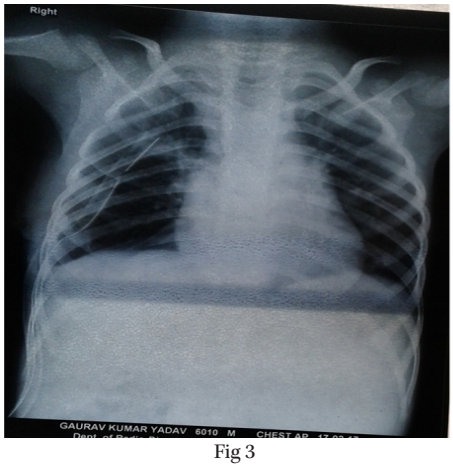


Fig 3

## DISCUSSION

CCAM is a rare condition characterized by immature, malformed lung tissue with cystic appearance, which results from an adenomatous overgrowth of terminal bronchioles and alveoli which leads ultimately to large masses, communicating with the tracheobronchial tree. These got blood supply from pulmonary vessels (1). First acknowledged as a separate entity and introduced into English literature by Chin and Tang in 1949(2). CCAM was first described and classified by Stocker et al. in 1977 (2), who classified it into 3 types. CCAM is best diagnosed with computed tomography (3), and classified as "Type I" - This is the most common type constituting 50-70%and composed of multiple cysts of size 2-10cm with no adenomatoid areas. These are lined by pseudostratified ciliated columnar epithelium. Prognosis is excellent. Type II -Composed of multiple individual cysts up to 2 cm with adenomatoid areas. This form is commonly associated with anomalies (especially renal, cardiac, intestinal and skeletal). Type III - Least common and is collection of multiple microcysts which are entirely adenomatoid.These appear solid on gross examination. Prognosis is poor due to respiratory compromise and associated congenital abnormalities (4). These lesions are almost always seen in males. CCAM can be detected in prenatal period and some cases present with maternal polyhydramnios and fetal hydrops. At birth patients usually present with life threatening respiratory distress. Some patients have history of recurrent chest infections during childhood and few are asymptomatic and are discovered as an incidental finding on radiography. CCAM is usually unilateral and more common in boys.It may be confused with congenital lobar emphysema which present in similar way but CT scan confirms diagnosis. CT scan is the investigation of choice for diagnosis (5).

Prenatal ultrasound classified the lesion as: - " Microcystic lesions (< 5 mm): usually associated with fetal hydrops and has poor prognosis." Macrocystic lesions (> 5 mm): has favorable prognosis (6).Chest radiography almost invariably identifies CCAM of sufficient size to cause clinical problems.Treatment is lobectomy. After lobectomy the remaining lung grows and expands well enough so that total lung volume and pulmonary function tests return to normal (7). This response is most dyanamic in very young because new acini and alveoli form upto 5 years age (8). Post-lobectomy results are excellent especially in younger groups who have lower ratio of residual volume to total lung capacity and higher maximum breathing capacity. This suggests that hyperplasia rather than over-distension occurs in the remaining lung (9).

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