

Case Report

The telltale Cinderella story of Congenital cystic adenomatoid malformation of the lungs in a child: A rare case report from Dakshin Kannada

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Abstract

Congenital cystic adenomatoid malformation (CCAM) is a developmental anomaly of the lung tissue occurring in paediatric age group. They are conglomerates of bronchopulmonary foregut malformations. Synonymously also denoted as congenital pulmonary airway malformation (CPAM). In this anomaly an entire lobe of lung is replaced by a non-working cystic piece of abnormal lung tissue. This abnormal tissue will never function as normal lung tissue. This is a case of a child who remained asymptomatic for long and presented complaints at the age of two.

Keywords: Congenital Cystic Adenomatoid Malformation Lung Paediatric Age Group

1. Introduction

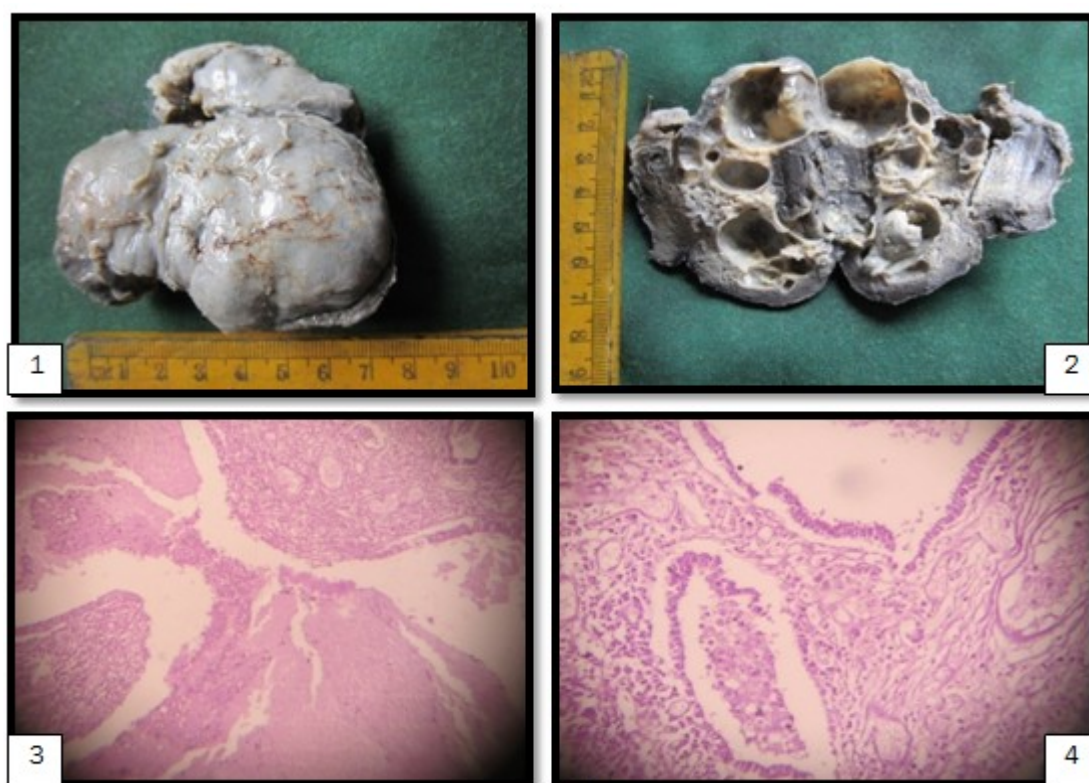
Congenital cystic adenomatoid malformation (CCAM) is a developmental anomaly, first illustrated as a distinct disease or entity by Ch'in and Tang in 1949.^{1,2} It was classified into 3 subtypes in 1977³, and expanded into 5 types with a new name as congenital pulmonary airway malformation (CPAM) by Stocker in 2002.⁴ 80% to 85% of cases are recognized in the first 2 years of life, adult presentation is uncommon⁵. The first ultra sonographic detection of CCAM in a fetus was done in 1975^{6,7} when fetal chest masses were considered rare entities.^{8,9,10,11} After 1990's the incidences of CCAMS Increased. It occurs in approximately 1 in every 35000 to 1 in 10,000 pregnancies.^{6,12,13,16} Recent studies have shown that antenatally detected CCAMS offer a better prognosis and favourable outcome for the patients.^{12,13,14,15,16}

2. Case Report

A two year old male child antenatally diagnosed with left sided cystic mass involving the left lower lobe of the lung. It was confirmed postnatally by CT scan and X ray findings (Fig:5,6). Two months back, succumbed to Respiratory tract infections and breathlessness. He had developed high grade fever and dyspnoea. The child had history of recurrent episodes of respiratory infections in the past and was intermittently treated with antibiotics and bronchodilators. He had no similar complaints or condition in any member of his family. On examining the child he appeared to be febrile and weighed 10 kgs. On examining the respiratory system it was noticed that left side of chest wall showed decreased movements with

decreased air entry in the left lower lobe. Further hematological investigations revealed that child had leucocytosis. Chest X-ray revealed multiple cystic spaces in the lower left lobe (Fig:5). CT scan showed multiple septated cystic spaces in the left lower lobe(Fig: 6). Left lower lobectomy was planned. Histopathology grossly of the lung mass measured 9 x 6.6 x 3 cms in dimensions whose lining consisting of necrotic areas and multiple cysts (Fig:1) .Cut surface showed multiloculated thin walled cystic spaces of varying sizes ranging from 3cms to 0.5 cms (Fig:2). Microscopically the lung tissue showed the cysts were lined by cuboidal to columnar epithelium. Some of the larger cystic spaces were lined by pseudostratified ciliated columnar epithelium and some cystic portions showed mucoid material in a background of mixed inflammatory cells. In between these cysts the stroma was comprised of smooth muscles, regenerating blood vessels and chronic inflammatory cell infiltrates (Fig: 3,4). Post operatively condition of the patient was uneventful, and he was discharged within 10 days. The patient has been followed up since then, and at present is absolutely healthy without any respiratory infections and all parameters are normal.

Fig (1 to 4): 1) Gross of the left lobe. 2) cut surface of the lobe showing multiple cysts ranging from 3 cms to 0.5cms. 3) light microscopy picture (Scanner view) showing multiple cystic spaces.4) (10X) showing cyst wall lined by columnar epithelium.



3. Discussion

Congenital cystic adenomatoid malformation is a developmental anomaly of the lung tissue.¹ They constitute the category of bronchopulmonary foregut malformations .The disorder is often referred to by another name, congenital pulmonary airway malformation (CPAM). In this anomaly an entire lobe of lung is replaced by a non-working cystic piece of abnormal lung tissue. This abnormal tissue will never function as a normal lung tissue. The blood supply is derived from the normal lung circulation.^{15,16} It may affect any lobe of the lungs .The cause is unknown. CCAM presents as a respiratory distress syndrome in a newborn, accompaniments are pulmonary hypoplasia, mediastinal shift, pleural effusion, recurrent infections, and if left neglected potent grounds for malignancies. All types are intrinsically the same lesion, which differ on the radiological presentation. Type I CCAM is characterized by single or multiple cysts more than 3 cm in diameter lined by pseudo stratified ciliated columnar epithelium, along with mucous cells which are considered to be potentially mutant to adenocarcinoma^{1,19}. Type II lesion is consisted of multiple terminal bronchiolar-like uniform cysts less than 2 cm in

diameter, lined by cuboidal to columnar epithelium.¹⁹ Type III CCAM usually involves an entire lobe of lung and has a sponge-like appearance, constructed by bulk gland-like structures. Another 2 types were proposed in 2002.²⁰ The new classification system with added type 0 and IV was not widely applied because type 0 is hard to differentiate from bronchogenic cyst and similarities between type IV and cystic pluropulmonary blastoma may result in misdiagnosis.²¹ Late-onset CCAM in adults may be more complicated on radiographic images due to recurrent infections. CCAMS are categorized into four types based on size of the cysts in mass¹⁷.

Table 1: Morphology of Cysts

Types	Morphology of Cysts
Type I	(70% of cases) is characterized by multiple large cysts surrounded by small cysts (>3 cm in diameter). These CCAMs seem to have the highest potential for becoming cancerous .
Type II	(20% of cases) has smaller cysts (< 2 cm in diameter). Associated with congenital lesions.
Type III	very small cysts < 0.5 cms (but often appears solid on ultrasound or X-ray).
Type IV	combination of small and large cysts.

(macrocyts : good prognosis, microcyts: bad prognosis)

Cystic adenomatoid transformation is characterized by the presence of variously sized intercommunicating cysts lined by an 'adenomatoid' cuboidal-to-ciliated pseudostratified columnar epithelium.¹⁸ It may be seen in association with bronchial atresia. It usually presents with respiratory distress in neonates, never the less found in older children and in adults. Solitary lesions usually involve a lower lobe. Some of the patients have associated pulmonary or extrapulmonary anomalies. Lobectomy is the treatment of choice. An increased number of cases of mucinous bronchiole-alveolar carcinoma have been reported in young patients with type 1 cystic adenomatoid transformation.¹⁸ Congenital cystic airway malformation is a rare developmental anomaly that presents within two years of life for which there is no absolute etiological rulebook ,only certain genetic , hereditary or environmental . However amongst so many reports published till date some of the CCAMS present in childhood and some present in adulthood. Some mention about the changes in the cytoplasm and nuclear details by intervening the use of immunohistochemistry markers like Ki-67,p53 mutations and K-ras mutations.^{22,23} Careful vigilance and a thorough follow up remains the icing on the cake as ever.Written informed consent was obtained from the patients for publication of this Case Report and any accompanying images.

Fig 5:X-Ray Showing Left Sided Mass

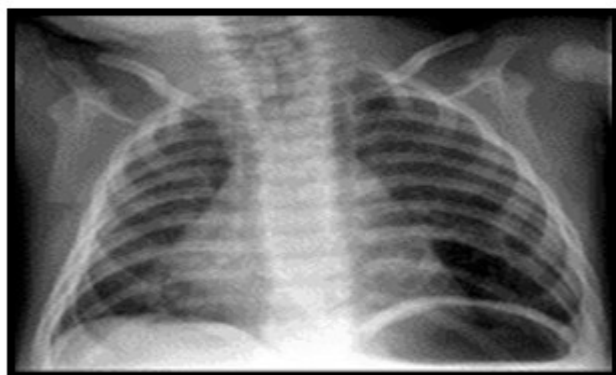
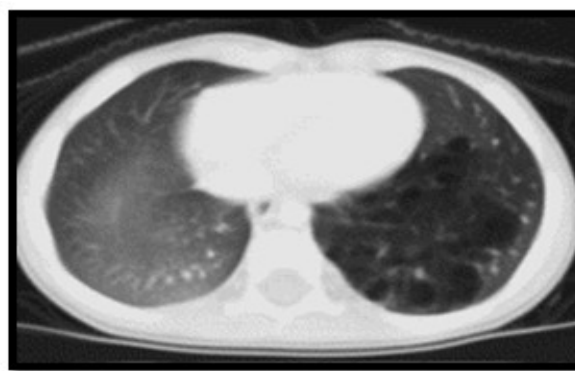


Fig 6 :CT of the Left Lung



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