

Case Report

Congenital Cystic Adenomatoid Malformation in an Adolescent: An Unusual Presentation with Pleural Effusion and Pneumatocele

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ABSTRACT

Congenital cystic adenomatoid malformation (CCAM) encompasses a continuum of hamartomatous cystic lung lesions characterised by the presence of abnormal bronchiolar structures of varying sizes or distribution. The CCAM is a disorder of infancy with majority of the cases being diagnosed within the first two years of life. We describe CCAM in a 13-year-old girl complaining of recurrent lower respiratory tract infections since infancy who presented with post-infectious pneumatocele with loculated pleural effusion, and suspected abscess formation and had undergone resection.

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Key words: Congenital cystic adenomatoid malformation, Pneumatocele, Pleural effusion.

INTRODUCTION

Congenital cystic adenomatoid malformations (CCAM) are rare congenital developmental anomalies of the lung.¹ They are considered bronchopulmonary foregut malformations caused probably by an arrest in lung development between 4th and 7th week of fetal life.² The CCAM is most commonly found in the neonatal period and upto 90% of diagnoses are made within the first two years of life.³ We present a case of a 13-year-old girl with a history of chronic infectious respiratory disease. The case is unusual due to the late age of onset and its uncommon presentation as post-infectious pneumatocele with pleural effusion.

CASE REPORT

A 13-year-old girl was referred to our hospital with the complains of high grade fever, productive cough, dyspnoea and left-sided pleuritic chest pain of one week duration. She gave history of recurrent episodes of lower respiratory tract infections since infancy and had received treatment intermittently with bronchodilators and antibiotics.

On examination, she was febrile and weighed 22Kg. Minimal digital clubbing and pallor were observed. Examination of the respiratory system revealed sinking in of the left hemithorax with

decreased chest wall movement on the left side. There was decreased air entry over the left lower lobe of the lung with occasional end-inspiratory crackles. Rest of the systemic examination was normal. Laboratory investigations revealed microcytic hypochromic anaemia with neutrophilic leukocytosis. Chest radiograph showed evidence of pleural effusion along with left lower lobe consolidation (Figure 1). Computed tomography (CT) of the thorax revealed left lower lobe

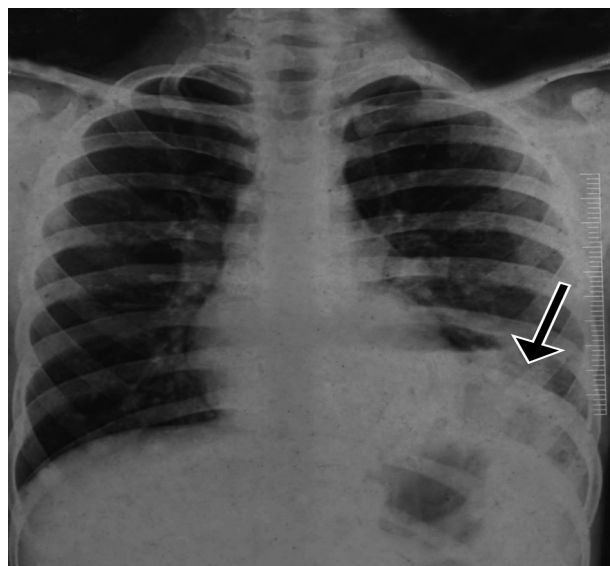


Figure 1. Chest radiograph (postero-anterior view) showing left lower lobe consolidation (arrow).

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consolidation with multiple septae with loculated pleural effusion and pneumatocele formation (Figure 2). The patient was taken up for surgery.



Figure 2. Contrast enhanced axial computed tomography of the chest demonstrating pleural effusion and pneumatocele of the left lower lobe of the lung (arrow).

At thoracotomy, the left lower lobe of the lung showed a large cyst (8cm × 5cm) lined by necrotic debris, and multiple small cysts (Figure 3). Histopathological examination showed a lesion comprised of cystic and malformed bronchi and bronchiolar structures. The larger cysts were lined by pseudostratified ciliated columnar epithelium and were filled with mucoid material and mixed inflammatory infiltrate. The intervening stroma was composed of smooth muscle bundles, proliferating blood vessels and chronic inflammatory infiltrate (Figure 4).

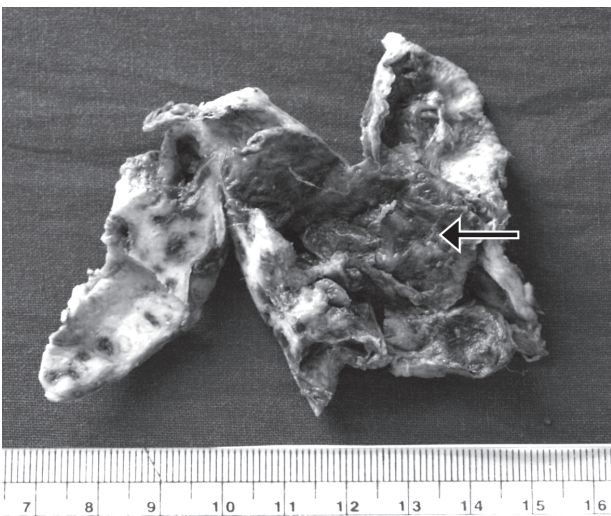


Figure 3. Gross photograph of the resected specimen showing a large cyst wall lined by necrotic debris (arrow), along with multiple small cysts.

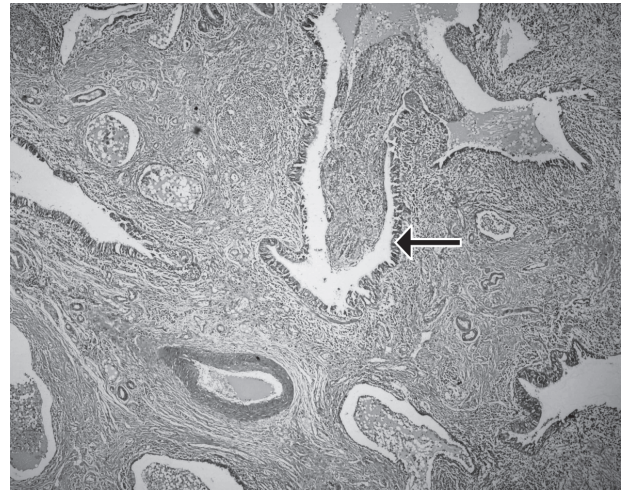


Figure 4. Photomicrograph showing cysts lined by pseudostratified ciliated columnar epithelium (arrow), along with cystic and malformed bronchi and bronchioles. The intervening stroma is comprised of smooth muscle bundles (Hematoxylin - Eosin × 100).

Immunohistochemistry revealed positive staining of the muscle fibres with desmin, and non-reactivity for myogenin. Based on these features, a diagnosis of congenital cystic adenomatoid malformation, type I (congenital pulmonary airway malformation, type I) was made.

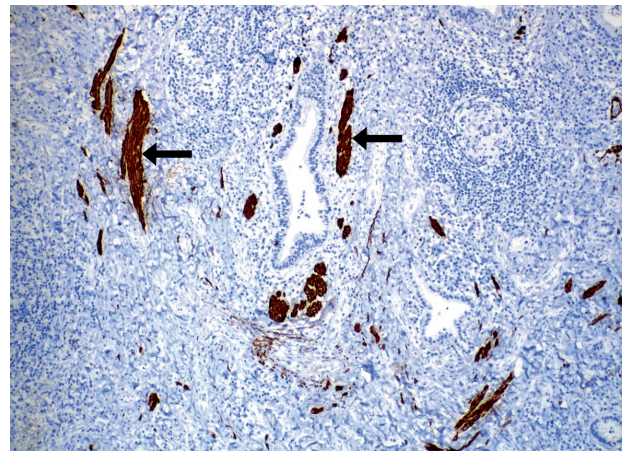


Figure 5. Photomicrograph of the immunohistochemistry revealing positive staining of the muscle fibres with desmin (IHC × 100).

The post-operative course was uneventful and the patient was discharged on the 7th post-operative day. The patient is being followed up for the past three months. She has had no respiratory infections during this tenure and her physical health is improving with weight gain and resolution of digital clubbing.

DISCUSSION

Congenital cystic adenomatoid malformation is an uncommon congenital malformation of the lung that

arises from excessive disorganised proliferation of tubular bronchial structures excluding the alveoli. These are believed to represent focal pulmonary dysplasia because skeletal muscle may be identified from within the cyst wall.⁴ Male subjects are affected as frequently as female subjects. The left lung is involved as often as the right lung with single lobe disease observed four times more often than multilobe disease.⁵

The most common mode of presentation is acute respiratory distress secondary to the cyst expanding and compressing its surrounding structures. The distress occurs through a ball-valve mechanism leading to air trapping. This mode of presentation is common during the neonatal period. It may remain asymptomatic and be discovered later in life on routine chest films or present after the neonatal period as recurrent pneumonia.⁶ Similar presentation was seen in our case as well wherein the patient gave history of recurrent episodes of lower respiratory tract infections since infancy. Late presentation of such cases has also been documented in the literature by other authors.^{3,7} The CCAM may be complicated by secondary bacterial, mycotic or tubercular infection. An occasional case of CCAM in an adult has been reported presenting as lung abscess.⁷ The CCAM presenting as pneumatocele is very rare, and to our knowledge only one such case has been documented in the literature.⁵ Our case is highly unusual because the patient presented with pneumatocele with localized pleural effusion.

Stocker *et al*⁸ in 1977 described three different types of CCAM. Type I is the large cyst category and accounts for 50% of cases. The lesion consists of an irregular cystic mass in which the larger cysts (3cm to 10cm) dominate, though smaller cysts are frequently noted in the background. The cysts are lined by pseudostratified columnar ciliated epithelium overlying a relatively prominent fibromuscular layer. Type II constitutes 40% of the cases and is comprised of numerous small cysts (0.5cm to 2cm) throughout the lesion without a predominant large cyst component. The lesion is composed of thin-walled bronchiole like structures. Type III accounts for 10% of CCAM cases and consists of innumerable evenly distributed small cysts measuring less than 0.2cm. The lesion tends to be large and may occupy an entire lobe or most of one lung. The microscopic appearance is suggestive of immature lung with a predominant background of alveoli-like structures lined by cuboidal cells.⁹

Type II CCAM is usually associated with other congenital anomalies like renal agenesis, pulmonary hypoplasia, pectus excavatum and various cardiac anomalies.⁵

Recently, Stocker¹⁰ has added two more types of CCAM to the existing classification according to the

anatomic and microscopic properties of pulmonary airway, and has used the term congenital pulmonary airway malformation (CPAM) for this anomaly. Briefly, type 0 CPAM (acinar dysplasia, 1% to 3%): tracheobronchiolar origin, solid appearance with small and firm lungs. Microscopy shows bronchial-type airways with cartilage, smooth muscle and glands separated by abundant mesenchymal tissue; type I CPAM (60% to 70%): bronchi/bronchiole origin, one or more large cysts measuring 2cm to 10cm in diameter, accompanied by smaller cysts. Cysts are lined by pseudostratified columnar epithelium and the walls are comprised of muscle, elastic or fibrous tissue; type II CPAM (10% to 15%): bronchiole origin, multiple small cysts ranging in size from 0.5cm to 2cm, cysts are lined by cuboidal to columnar epithelium and have a thin fibromuscular wall; and type III CPAM (5%): bronchiolar/alveolar origin, multiple small cystic or solid type; and type IV CPAM (28%): distal acinary originated peripheral cystic type. Large cysts (upto 10cm) lined by flattened epithelium and resting on loose mesenchymal tissue.¹¹

An important differential diagnosis to be considered is pleuropulmonary blastoma (PPB). Radiographically, PPB shows either intracystic mass or septal thickening, both of which militate against a diagnosis of CCAM. Morphologically there are three types of PPB. Type I PPB shows multiple cysts lined by benign respiratory type epithelium underlaid by malignant cells forming a cambium layer. The latter consist of small cells that may show rhabdomyoblastic differentiation. There may be small nodules of mature fibroblasts, fetal cartilage or hyalinised stroma. In transition to type II, a sarcomatous component begins to overgrow the septa. Type III PPB is predominantly solid variant with mixed blastematos and sarcomatous appearance.

On immunohistochemistry, the rhabdomyoblastic component may show positive staining for myogenin, MyoD1 and desmin. Ki-67 staining may show upto 90% proliferation index within the cambium layer.¹²

The CPAM in contrast to PPB shows only cysts lined by respiratory epithelium, with muscle and fibrous tissue within the cyst wall. There is absence of malignant cells, cambium layer and any blastemal or sarcomatous component. The muscle fibres and myofibroblasts may show positive staining for desmin, but will be non-reactive for myogenin and myoD1. Type IV CPAM can be associated with PPB.¹³

CCAM should also be differentiated from other cystic lesions of the lung, like pulmonary sequestration, bronchogenic cyst, congenital lobar emphysema, diaphragmatic hernia and cystic bronchiectasis.¹⁴

Various diagnostic modalities for CCAM include chest radiograph and computed tomography. Prenatal diagnosis in the fetus can be done through ultrasound.¹³ Definitive diagnosis can be made only following histopathological examination.

Rarely, CCAM may be the harbinger of a malignancy. Development of bronchoalveolar carcinoma¹⁴ and a case of embryonal rhabdomyosarcoma¹⁵ within CCAM have been described, in the literature.

The definitive treatment of CCAM is surgery. In lobectomy the remaining lung grows and expands well enough so that the total lung volume and pulmonary function tests return to normal.¹⁶

To conclude, early recognition and surgical treatment of CCAM is essential to prevent the consequences of recurrent pulmonary infections and the potential risk for malignant transformation.

REFERENCES

1. Chi'n KY, Tang MY. Congenital cystic adenomatoid malformation of one lobe of a lung with general anasarca. *Arch Pathol Lab Med* 1949;48:221-9.
2. Morotti RA, Cangiarella J, Gutierrez MC, Jagirdar J, Askin F, Singh G, *et al*. Congenital cystic adenomatoid malformation of the lung (CCAM): evaluation of the cellular components. *Hum Pathol* 1999;30:618-25.
3. Morelli L, Pisciole I, Licci S, Donato S, Catalucci A, Nonno FD. Pulmonary congenital cystic adenomatoid malformation, type I, presenting as a single cyst of the middle lobe in an adult: case report. *Diagn Pathol* 2007;2:17.
4. Mohta A, Kanojia RP, Bathla S, Khurana N. Cystic adenomatoid malformation of the lung: a diagnostic dilemma. *Afr J Paediatr Surg* 2009;6:112-3.
5. Cloutier MM, Schaeffer DA, Hight D. Congenital cystic adenomatoid malformation. *Chest* 1993;103:761-4.
6. Sindhwani G, Whig J, Gupta A, Rawat J. Tuberculosis in congenital adenomatoid malformation: a rare presentation. *Lung India* 2007;24:148-9.
7. Dahabreh J, Zisis C, Vassiliou M, Arnoyiannaki N. Congenital cystic adenomatoid malformation in an adult presenting as lung abscess. *Eur J Cardiothorac Surg* 2000;18:720-3.
8. Stocker JT, Madewell JE, Drake RM. Congenital cystic adenomatoid malformation of the lung: classification and morphologic spectrum. *Hum Pathol* 1977;8:155-71.
9. Kuhn C, West WW, Craighead, Gibbs AR. Lungs. In: Damjanov I, Linder J, editors *Anderson's Pathology*. St. Louis: Mosby & Co.;1996:pp951-84.
10. Stocker JT. Congenital pulmonary airway malformation: a new name and an expanded classification of congenital cystic adenomatoid malformations of the lung. *Histopathology* 2002;41:424-31.
11. Hill DA, Jarzembowski JA, Priest JR, Williams G, Schoettler P, Dehner LP. Type I pleuropulmonary blastoma; pathology and biology study of 51 cases from the international pleuropulmonary blastoma registry. *Am J Surg Pathol* 2008;32:282-95.
12. MacSweeney F, Papagiannopoulos K, Goldstraw P, Sheppard MN, Corrin B, Nicholson AG. An assessment of the expanded classification of congenital cystic adenomatoid malformations and their relationship to malignant transformation. *Am J Surg Pathol* 2003;27:1139-46.
13. Singh M, Mitra S, Kumar L, Narang A, Rao KLN, Kakkar N. Congenital cystadenomatoid malformation of lung. *Indian Pediatr* 2000;37:1269-74.
14. Kaslovsky RA, Purdy S, Dangman BC, McKenna BJ, Brien T, *et al*. Bronchoalveolar carcinoma in a child with congenital cystic adenomatoid malformation. *Chest* 1997;112:548-51.
15. d'Agostino S, Bonoldi E, Dante S, Meli S, Cappellari S, Musi L. Embryonal rhabdomyosarcoma of the lung arising in cystic adenomatoid malformation: a case report and review of literature. *J Pediatr Surg* 1997;32:1381-3.
16. Filler J. Effects upon pulmonary function of lobectomy performed during childhood. *Am Rev Respir Dis* 1964;89:801-10.