

CASE REPORT

CONGENITAL CYSTIC ADENOMATOID MALFORMATION OF LUNG: A CASE REPORT

Niranjan J¹, K. V. Santosh², Shashidhar S³, Shefali H. K⁴

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INTRODUCTION: Congenital cystic adenomatoid malformation (CCAM) is a rare developmental abnormality of the lung previously termed as 'benign hamartoma' or 'dysplastic lung tumor'. It is characterized by abnormal proliferation of tissue resembling terminal bronchioles with cessation of bronchial maturation at the 5th and 7th week of development.⁽¹⁾ The lesion is predominantly unilateral, mostly in the left lung and usually involve one lobe. CCAM has a mild male predominance, with an incidence of 1 in 11000⁽²⁾ to 1 in 35000⁽³⁾ live births. The exact pathogenesis is unknown; some authors suggest a possible role of over expression of fibroblastic growth factor in the pulmonary mesenchymal cells⁽²⁾ and others suggest Fatty Acid Binding Protein (FABP 7) under expression in the fetal lung.⁽⁴⁾

CASE REPORT: A 2 year old female child presented to our OPD with history of recurrent episodes of cough, fever and breathlessness since one month after birth. There was no significant antenatal, natal or post natal history.

On examination, the trachea was shifted towards right; with reduced expansion and hyper resonance on the left side. Breath sounds were reduced bilaterally, left side more than right. No crepitation's, ronchi or transmitted sounds were heard. Biochemical and hematological work up were within normal limits. Chest X ray showed large areas radio lucency in the left lung with displacement of mediastinum to the right side (Fig. 1). CT findings were suggestive of multiloculated cystic lesion in the lung. (Fig. 2). Clinically, a diagnosis of "Congenital lobar emphysema" was made.



Fig. 1: Shows radio lucent left lung with displacement of mediastinum



Fig. 2: shows multi cystic lesions in the left lung

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Left posteriolateral thoracotomy and left upper lobectomy was done under general anesthesia. The post-operative period was uneventful. Macroscopically, the lobectomy specimen measured 7x7x1.5 cm (Fig. 3). Cut section showed many cystically dilated areas with hemorrhage surrounded by normal appearing lung tissue. Microscopy showed multiple variably sized intercommunicating cysts lined by cuboidal to ciliated Pseudostratified columnar epithelium (Fig. 4); at places interspersed with mucin cells (Fig. 5). The subepithelium shows thickened fibromuscular connective tissue with areas of calcification and necrosis.

A final diagnosis of "Congenital cystic adenomatoid malformation - Type I" was made.



Fig. 3: Shows cystically dilated areas surrounded by normal appearing lung tissue

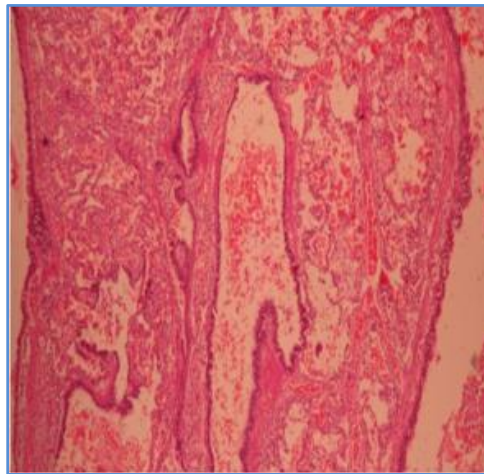


Fig. 4

Fig. 4: Shows multiple variably sized intercommunicating cysts lined by cuboidal to ciliated Pseudostratified columnar epithelium, subepithelium shows thickened fibromuscular connective tissue. (10X, H&E).

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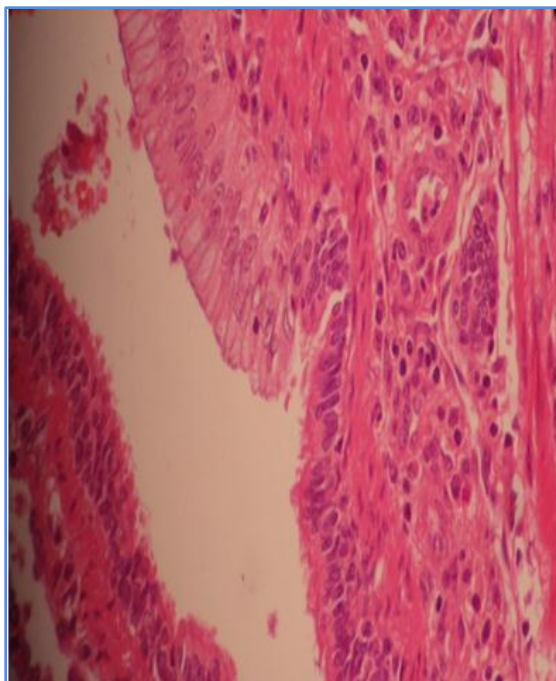


Fig. 5

Fig. 5: High power showing ciliated Pseudostratified columnar epithelium with mucin cells (40X, H&E).

DISCUSSION: CCAM was described initially by Stocker in 1897 and later by Ch In and Tang in 1949 as a developmental abnormality.⁽¹⁾ CCAM accounts for 95% of congenital cystic lung diseases⁽⁵⁾, usually presenting in infancy with progressive respiratory distress, recurrent infections and compression of normal surrounding lung tissue.^(6,7) Stocker classified CCAM into 3 subtypes based on proportion of cysts, adenomatous tissue and on the dominant cell type.⁽⁸⁾ Type I (macrocytic adenomatoid malformation) has large single to multiple cysts more than 2 cm in diameter lined by columnar epithelium with smooth muscle and elastic tissue in the walls. Mucin cells and cartilage are seen in this type. This type accounts up to 50% - 70% of cases and has a better prognosis. Stocker type II (microcytic adenomatoid malformation) consists of less than 1 cm diameter cysts with replacement of lung parenchyma by microcytic mal development. The solid part of the lung is filled with distended respiratory bronchiolar and alveolar material. This type accounts up to 20%- 40% of cases. Stocker III (solid adenomatoid malformation) is made up of solid airless lesions lined by ciliated cuboidal epithelium of bronchiolar epithelium and some alveolar elements. Accounting for 10% of cases, it has the worst prognosis.⁽⁹⁾ Later on, two variants were added: type 0 consisting of bronchial like histology and type 4 with acinar like lesions.⁽⁸⁾

The natural history of CCAM is variable and spontaneous regression during the course of gestation is not uncommon.⁽¹⁰⁾ Fetal sonography helps in detection of in-utero CCAM lesions, but appropriate post natal diagnostic investigations like CT scan are necessary to confirm diagnosis and for surgical decision making. CCAM is reported to harbor foci of mucous cell hyperplasia, foci

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of bronchoalveolar carcinoma; and cases of development of bronchioloalveolar carcinoma⁽⁹⁾ and mucoepidermoid carcinoma⁽¹¹⁾ have been reported.

To conclude, the treatment of choice is surgical intervention like segmental resection, or lobectomy. Radiological examination provides a morphological assessment of lung cavitary lesions but is inadequate to differentiate CCAM from other cystic lung diseases. The differentiation is essential since CCAM can be associated with malignancy.

REFERENCES:

1. Ch'in K Y, Tang M Y. congenital adenomatoid malformation of one lobe of a lung with general anasarca. Arch Pathol 1949; 48: 221-229.
2. Laberge J M, Flaeole H, Pugash D et al. Outcome of the prenatally diagnosed congenital cystic adenomatoid malformation: A Canadian experience. Fetal Diagn Ther 2001; 16: 178-186.
3. Gornall AS, Budd JL, Draper ES, et al. Congenital cystic adenomatoid malformation: accuracy in a prenatal diagnosis, prevalence and outcome in general population. Prenatal Diagn 2003; 23: 997-1002.
4. Amy J. Wagner, Amber S, Zachary T, Jess Edmondson et al. Genetic analysis of congenital cystic adenomatoid malformation reveals a novel pulmonary gene: Fatty acid Binding Protein-7 (Brain type). Pediatric Research 2008; Vol64, No1: 11-16.
5. Cloutier MM, Schaeffer DA, Hight D: Congenital cystic adenomatoid malformation. Chest: 1993; 103: 761-764.
6. Haller JA, Golladay ES, Pickard LR. Surgical management of lung bud anomalies: Lobar emphysema, bronchogenic cyst, cystic adenomatoid malformation and intralobar pulmonary sequestration. Ann Thorac Surg 1979; 28: 33-43.
7. Adzick NS, Harrison MR, Glick PL, et al. Fetal cystic adenomatoid malformation: Perinatal diagnosis and natural history. J Pediatr Surg; 1985; 20: 483-488.
8. Stocker JT. Congenital pulmonary air way malformation: A new name and an expanded classification of congenital cystic adenomatoid malformation of the lung, Histopathology 2002; 41: 424-58.
9. Stocker JT. Congenital and Developmental diseases: Dail and Hammar's Pulmonary Pathology. Vol I, 3rd ed: 2008; 132-175.
10. Sauat F, Michel J. I, benachi A, Emond S, and Revillonet Y. Management of asymptomatic neonatal cystic ademomatoid malformations, Journal of Peadiatric Surg, 2003; 38(4):548-552.
11. Harini N, Chaktavarthy R, Archana L. Congenital pulmonary air way malformation with meucoepidermoid carcinoma: A case report and review of literature, Indian Journal of Path and Micro 2012; 55 (4): 540-542.

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AUTHORS:

1. Niranjan J.
2. K. V. Santosh
3. Shashidhar S.
4. Shefali H. K.

PARTICULARS OF CONTRIBUTORS:

1. Assistant Professor, Department of Pathology, Vydehi Institute of Medical Sciences and Research Center, Bangalore.
2. Professor, Department of Pathology, Vydehi Institute of Medical Sciences and Research Center, Bangalore.
3. Tutor, Department of Pathology, Vydehi Institute of Medical Sciences and Research Center, Bangalore.

4. Tutor, Department of Pathology, Vydehi Institute of Medical Sciences and Research Center, Bangalore.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Niranjan J,
No. ½, 16th D Main,
HAL IInd Stage, Bangalore – 560008.
E-mail: drniranjanjayaram@gmail.com
drniranjanjayaram@hotmail.com

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