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Congenital Cystic Adenomatoid Malformation of Lung - Case Series

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ABSTRACT

Congenital cystic adenomatoid malformation (CCAM) is a rare entity with unexplained cause that effects the distal bronchi. It accounts for 25% of congenital pulmonary malformations with surgical excision as the treatment of choice. The mortality rate of patients with CCAM ranges from 9 to 49% if diagnosed prenatally. The outcome of a patient mainly depends on the presence of hydrops foetalis, microscopic type of CCAM, and the size of the lesions. The mainstay treatment of CCAM is surgical excision of the lesion which prevents complications of repeated infections, pneumothorax, and malignancy. We present 2 cases one is 5-month-old male child and other 8-month female child admitted to hospital with shortness of breath and poor suckling who was evaluated for respiratory distress. Clinically diagnosed as pneumonia, CT chest showed a cystic malformation in right lower lobe of lung and excision was done. Specimens were sent for histopathological examination and was diagnosed as congenital cystic adenomatoid malformation.

Keywords: congenital cystic adenomatoid malformation, infant, lobectomy

INTRODUCTION

Congenital Cystic Adenomatoid Malformation (CCAM) is a rare abnormality of lung development with idiopathic cause, that affects distal bronchi [(1).] It accounts for 25% of congenital pulmonary malformations and most of the cases are found in neonates and infants. There are single or multiple cysts in different locations, with ipsilateral or even mediastinal pulmonary compression [(2)]. The incidence of prenatally diagnosed CCAM is 1: 25,000–35,000. CCAM may present in old children and adults as an incidental finding due to repeated infections. [(3)] The mortality rate of prenatally diagnosed cases ranges from 9 to 49%. The risk factors for poor prognosis include hydrops foetalis [(4,5)] microcystic CCAM [(4,6)] and the total size of the lesions [(4,7,8).] Currently, with advances in radiology prenatal imaging, pulmonary tract defects can be detected during pregnancy or at birth [(9).] The diagnosis is based primarily on CT scans, but remains difficult because of its rare presentation [(10).] The mainstay of CCAM treatment is surgical removal of lesion whereby complications such as recurrent infections, pneumothorax, and malignancy can be prevented. [(3,10)]

CASE REPORT 1:

A 5-month-old boy was born by normal vaginal delivery without any complications. The mother was on regular antenatal care and it was normal. He had several episodes of pulmonary infections with history of shortness of breath and had a history of pneumonia 3 months back. He presented with shortness of breath and poor suckling. He was admitted to intensive care in view of respiratory distress. Physical examination

showed a sick baby with tachypnea. A working diagnosis of pneumonia was given and treated accordingly. CT chest showed a large cystic lesion with a well-defined wall in the right lower lung lobe consisting of multiple cysts of varying sizes suggestive of cystic malformation. Due to continuing recurrent infections and the risk of progressive respiratory distress with subsequent pneumothorax, right lower lobectomy was done and the specimen was sent for histopathological examination.

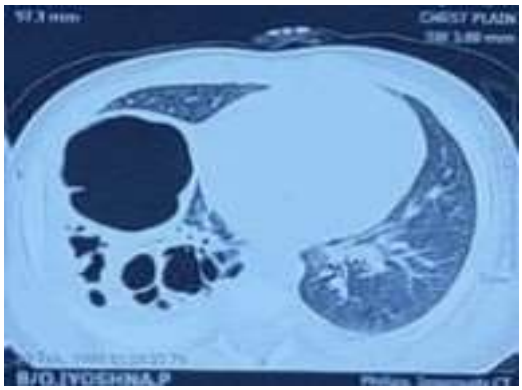

Figure 1: Thoracic CT showed a cystic malformation in the right lower lung lobe.

GROSS:_ In our department, we received lobectomy specimen in two bits. Largest measuring 7x5x2.5cm, smallest bit measures 4x2.5x1.5cm. External surface of both the bits was grey-brown with multiple bronchial openings noted and cut surface showed dilated cystic spaces measuring 3x3cm with multiple grey-white areas.

Figure 2: Cut surface of right lobectomy specimen showing dilated cystic spaces

MICROSCOPY: Multiple sections studied show lung tissue with irregularly dilated alveolar spaces with focal sequestration of lung tissue in between. A large cyst was observed which is lined by single layer to multiple layers of pneumocytes with hyalinization underneath. Foci of cartilage is seen within the wall of the large cyst. Multiple small-sized cysts and focal area of atelectasis were noted. Based on above histomorphology, a diagnosis of congenital cystic adenomatoid malformation was given.

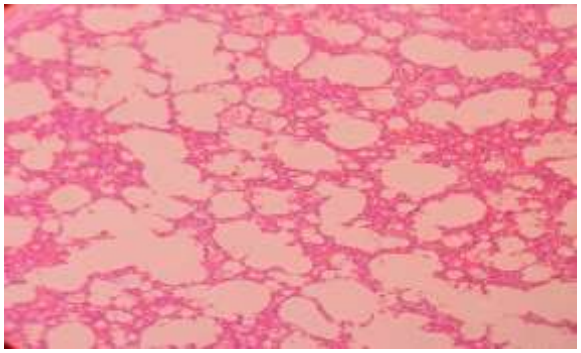

Figure 3: 100xH&E showing multiple irregularly shaped cystic spaces within normal lung parenchyma

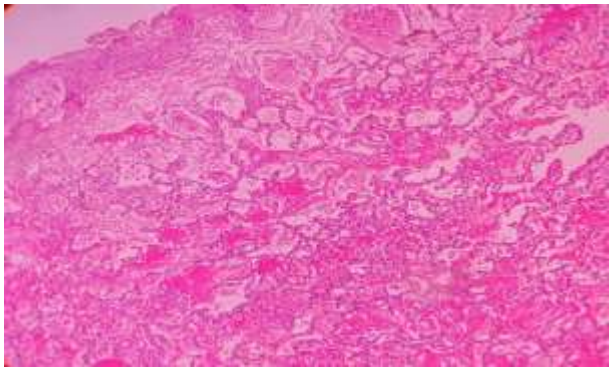

Figure 4: 400XH&E Showing cysts are lined by single to multiple layers of pneumocytes with hyalinization

CASE REPORT 2:

An 8-month-old girl was born by normal vaginal delivery. The mother was on regular antenatal care and her antenatal ultrasound examination was given as suggestive of congenital cystic adenomatoid malformation (CCAM) / Congenital pulmonary airway malformation (CPAM) of right lung and child got delivered at term. She had several episodes of pulmonary infections with history of shortness of breath which were successfully treated with antibiotics. She had a history of severe pneumonia recently and got treated conservatively. CT chest showed a large cystic lesion with a well-defined wall in the right upper lung lobe consisting of multiple cysts of varying sizes suggestive of cystic malformation. Due to continuing recurrent infections and the risk of progressive respiratory distress with subsequent pneumothorax, right upper lobectomy was done and the specimen was sent for histopathological examination.

GROSS: In our department, we received lobectomy specimen and lymphnode. Lobectomy specimen measuring 6x5x1.5cm, lymphnode measures 0.3x0.3 cm. External surface lung was grey-brown with multiple bronchial openings noted and cut surface showed dilated cystic spaces measuring 3x2.5cm with multiple grey-white areas. Lymphnode external and cut suface is grey tan in colour.

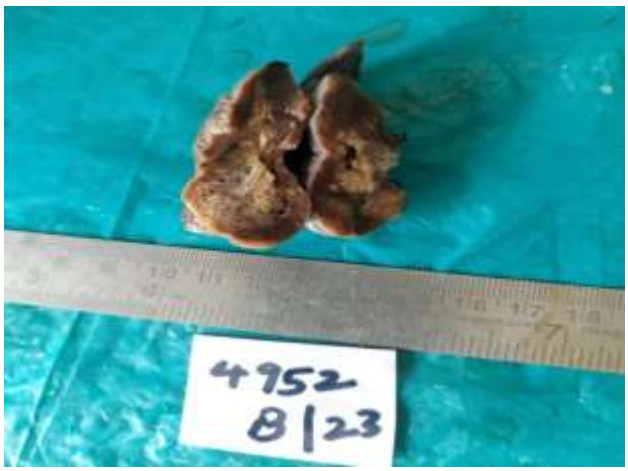

Figure 5: Cut surface of right lobectomy specimen showing dilated cystic spaces

MICROSCOPY: Multiple sections studied show cysts of varying sizes lined by ciliated columnar epithelium with focal stratification and are surrounded by alveolar spaces. Few of the cysts are filled with alveolar macrophages and eosinophilic material. In lymphnode no abnormality detected Based on above histomorphology, a diagnosis of congenital cystic adenomatoid malformation was given.

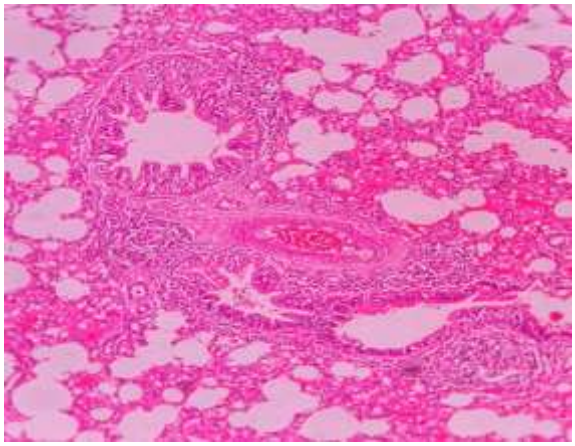

Figure 6: X100 H&E showing multiple irregularly shaped cystic spaces lined by ciliated columnar epithelium within normal lung parenchyma

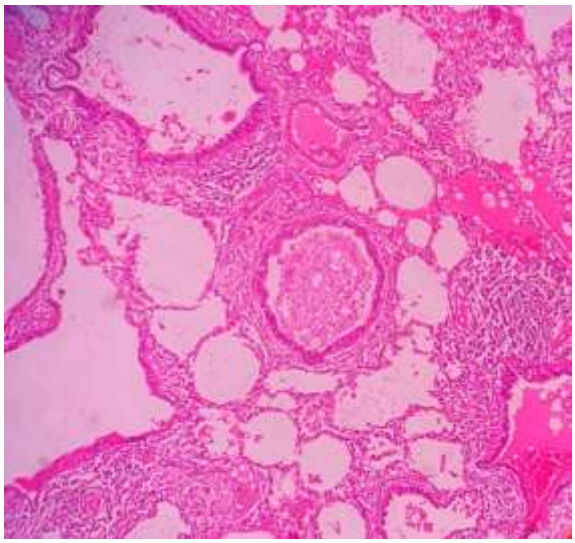

Figure 7: X100 H&E showing multiple irregularly shaped cystic spaces filled with alveolar macrophages

DISCUSSION

Stocker's original classification included types 1, 2, and 3, while types 0 and 4 were added later. [(11)] They should be different in the recent updates, Stoker's classification has been replaced by the term, Congenital Pulmonary Airway Malformation (CPAM). CPAM classified into 0 to 4 on histopathology. Type 0, where small lungs are seen with nodular surface. Microscopically type 0 shows disorganized proximal airways with absence of distal components. Type 1, shows medium to large interconnecting cysts with 1-2cm limited to one lobe, histologically cyst wall is composed of bronchial epithelium with clusters of mucus cells, smooth muscle cells and vascular connective tissues. Type 2, shows back-to-back dilated bronchoalveolar cysts (0.5 to 2cm). Type 3, presence as solid mass involving one lobe or entire lung which histologically shows randomly arranged gland like structures. In the last Type 4, cysts are distributed peripherally, which may be multiple. The cysts may be 0.1 to 0.3 cm.

Differential Diagnosis

They should be differentiated from pulmonary sequestration where there is systemic blood supply and there is no communication with tracheobronchial tree.

CONCLUSION

CCAM is a rare developmental malformation of the lung, usually diagnosed in fetal or neonatal life. Serial ultrasonographic evaluation, fetal lung mass size, and fetal echocardiography are needed for the management of antenatally detected cases. We report this case for its rarity.

Declaration by Authors

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Conflict of Interest: The authors declare no conflict of interest.

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