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CASE REPORT

CONGENITAL PULMONARY AIRWAY MALFORMATION (CPAM) TYPE II – A RARE CASE

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ABSTRACT

Congenital cystic adenomatoid malformation of the lung is a rare lesion which manifests as neonatal respiratory distress secondary to progressive expansion of the affected lung. Three distinct types have been described based on the size of the cysts and the microscopic appearance. The treatment of choice is excision of the affected lobe. The prognosis is favourable in the absence of pulmonary hypoplasia, foetal hydrops, or associated congenital anomalies.

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INTRODUCTION

Congenital pulmonary airway malformation of the lung is a rare disease that shows multiple cystic lesions in pulmonary tissues in the development process. It was first described by Chin *et al,* in 1949 and its incidence is known to be 1:25,000 to 1:35,000. It is detected within 2 years because of such symptoms as respiratory distress by compression of surrounding lung tissues immediately after birth and repeated lower respiratory tract infections in infancy (Ho Sung Lee *et al.,* 2012). The lesion affects male subjects slightly more frequently than female subjects and has no racial predilection. The vast majority of patients present in the immediate neonatal period, but antenatal diagnosis is possible and late presentation in olden children and adults occurs (Melissa, 1991).

CASE REPORT

A 1year old, female child presented at our hospital with dyspnoea and history of repeated lowerrespiratory tract infections. Chest exam revealed decreased airway entry in right lung. CT Thorax (HRCT + Post Contrast) revealed Multiple large thin walled cystic lesions involving the right middle lobe likely suggestive of Congenital Cystic Adenomatoid Malformation. Blood Investigations were within normal limits. Open thoracotomy with Right Middle Lobectomy was performed. Specimen (DYP/S/2575/2017) of Right middle lobe of lung received measuring 7 x 5 x 1 cm.

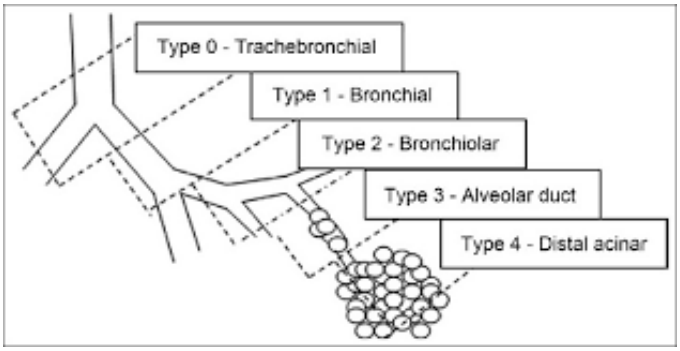
On gross examination, external surface was reddish brown, congested. Pleura was thickened. Cut surface revealed multiple inter-communicating cysts of varying sizes ranging from 0.5 to 2.0 cm. Cysts were merging with each other with intervening septations. On microscopy sections from the submitted lung tissue show multiple cysts arising from bronchiole. Cyst walls are thin and are lined by ciliated pseudo-stratified cuboidal to columnar epithelium surrounded by fibro-collage nous tissue. Cyst macrophages were seen. Speckled areas of calcification and necrosis was also noted. Focal areas of haemorrhage was seen. Surrounding lung tissue showed alveoli filled with blood. Intervening septa was thickened and showed fibrosis. No evidence of atypia or malignancy was seen.

Follow up

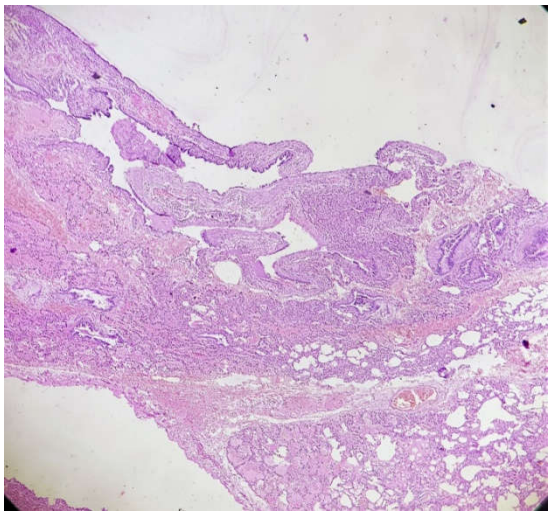
The surgery was uneventful and patient does not have any complications. Patient is doing well with normal respiratory functions and is not on any treatment.

DISCUSSION

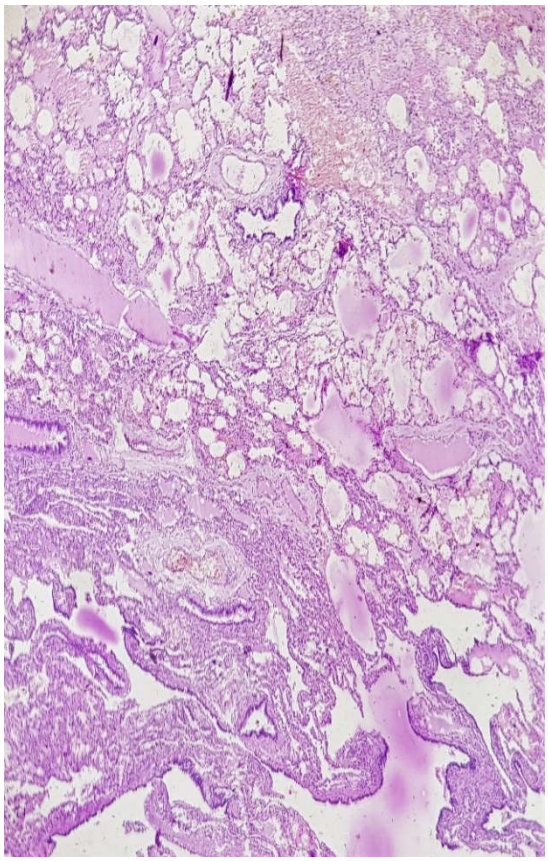
The CCAM is part of the group of structural abnormalities collectively referred to as bronchopulmonary foregut malformations due to altered development of the respiratory system at 3 weeks (Gianluigi Sergiacomi *et al.,* 2014). It results from regional deregulation of involved genes at a specific time point during organogenesis and differentiation and not by chromosomal imbalance (Elvira Stacher *et al.,* 2004).



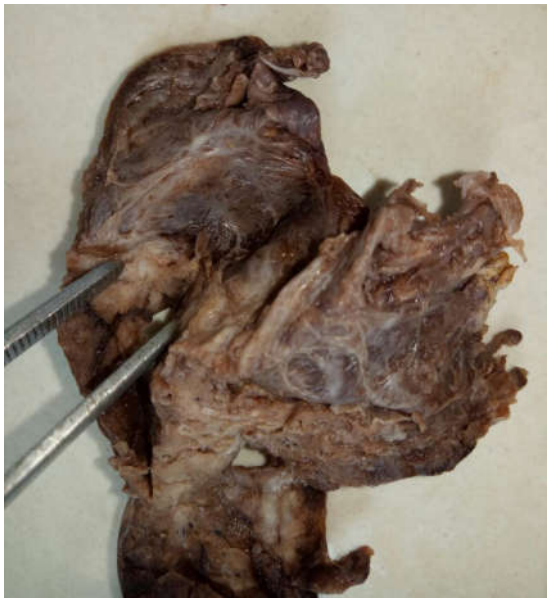
Pic 1. Reference No. 5



Pic 2. DYP/S/2575/2017. Section studied shows cyst walls lined by ciliated pseudo-stratified cuboidal to columnar epithelium surrounded by fibro-collagenous tissue. H&E; 4X



Pic 3. DYP/S/2575/2017. Section studied show columnar to cuboidal lining of the cyst wall along with air spaces filled withoedema fluid. H&E;10x



Pic 4. DYP/S/2575/2017. Multiple inter-communicating cysts of varying size ranging from 0.5 to 2.0 cm are seen merging with each other with intervening septations

Classification schemes for CCAM have evolved, and there are currently five main types, which differ based on the embryologic level of origin and the histologic features (Figure 1) (Anna *et al.,* 2012):

Type 0 (bronchial) It is the rarest form. and arises from the trachea or bronchus. The presentation is severe and usually lethal.10 Cysts are small (Anna *et al.,* 2012).

Type 1 (bronchial/ bronchiolar) It is the most common form, representing 50% to 70% of cases, and it arises from the distal bronchus or proximal bronchiole (Anna *et al.,* 2012).

Type 2(bronchiolar) It accounts for 15% to 30% of cases and arise from terminal bronchioles (Anna *et al.,* 2012). Which was seen in our case.

Type 3 (bronchiolar/ alveolar) It account for 5% to 10% of cases and are thought to arise from acinar-like tissue (Anna *et al.,* 2012).

Type 4 (peripheral) it account for 5% to 15% of cases and are alveolar in origin (Anna *et al.,* 2012).

Type 2 lesion: The intermediate cyst type, contains cysts that are simpler and rarely exceed 2 cm in diameter. The cysts lined by cuboidal to columnar epithelium resemble dilated terminal and respiratory bronchioles organized in a back to back configuration (Anna *et al.,* 2012). Cartilage plates are rarely present except for those that are part of normal bronchi near the edge of the lesion. Mucogenic cells are not seen in type 2 CCAM (Melissa *et al.,* 1991). It may be associated with other severe malformations like renal agenesis or dysgenesis, Cyst walls are thin and are lined by ciliated pseudo-stratified columnar to cuboidal to columnar epithelium surrounded by fibro-collagenous tissue (Melissa *et al.,* 1991). Present case did not show any of these malformations on investigations.

Conclusion

CPAM may cause severe respiratory distress at birth and may require prompt respiratory support and immediate surgical

intervention. Therefore, understanding the morphologic characteristics of this lesion may head to the recognition of its varied radiologic appearances, allowing early diagnosis and optimal patient management.

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