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RESEARCH ARTICLE

CONGENITAL CYSTIC ADENOMATOID MALFORMATION – A RARE CASE REPORT

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ABSTRACT

Congenital cystic adenomatoid malformation (CCAM) is a hamartomatous lesion of the lung, with an incidence of about 1 in 5,000 live births. It is an uncommon anomaly characterized by multicystic lesions due to the proliferation of the respiratory bronchioles. It is seen in term and premature infants. The etiology remains unknown. Eighty percent of the lesions are present in the neonatal period. Here, we have described a case of CCAM in a newborn child.

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INTRODUCTION

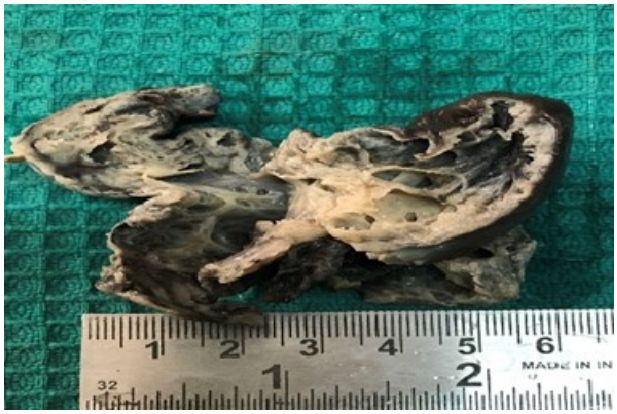
Lung development begins by the third week of gestation and involves five stages[6].Congenital cystic adenomatoid malformation (CCAM) is an uncommon development abnormality of the lung. It accounts for 25% of all congenital lung malformations and 95% of congenital lung lesions [3]. It is a hamartomatous lesion of the lung, with an incidence of about 1 in 5,000 live births[1].Congenital cystic adenomatoid malformation is characterized by multicystic lesions due to the proliferation of the respiratory bronchioles[2]. It is seen in term and premature infants who are cyanotic at birth and they survive only a few hours and are associated with cardiovascular abnormalities and dermal hypoplasia. The etiology of Congenital cystic adenomatoid malformation remains unknown, but it is suggested that there is a disturbing interaction between mesodermal and ectodermal components of the lung during embryonic development[2]. Eighty percent of the lesions are present in the neonatal period, however, it is rarely seen in adults[2,7].

CCAM in the adult is mostly unilateral and it is complicated by pulmonary bacterial infections[5] . Here we have discussed a case of CCAM in a newborn.

Clinical history: One-day-old newborn infant, male, was born to a G1P1 mother. The mother had spontaneous normal labor at 39 weeks of gestation. The patient's mother had a history of pregnancy-induced hypertension. A routine ultrasound examination done at 36weeks revealed polyhydramnios and cystic lesions within the right lung. Based on the findings, Congenital pulmonary airway malformation (CPAM) is felt to be the most likely diagnosis. At birth, the baby had mild respiratory distress, cyanosis, and air entry to the right-side lung was very much diminished. The APGAR score at 1min and 5min were 7/10,9/10 respectively. The birth weight was 3000 grams. The patient was admitted to NICU with respiratory distress and a chest x-ray was performed. A postnatal chest x-ray showed features of CPAM in the right upper lobe of the lung. The patient was shifted to the pediatric surgery unit where they planned for an emergency thoracotomy.

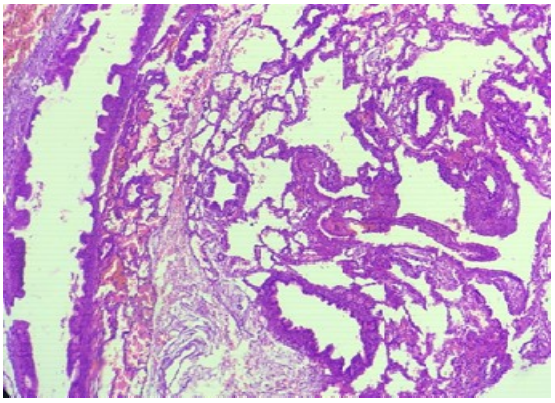
The patient underwent surgery, thoracotomy was performed and a large cystic lesion in the right upper lobe was decompressed by needle aspiration. Right upper pulmonary lobectomy was done. The specimen was sent for histopathological examination.

Macroscopy: A lobectomy specimen measuring 6.5x4.5x2cm is received. The macroscopic examination of the surgical specimen revealed a cavity measuring 3x2cm surrounded by hemorrhagic pulmonary tissue. Small cystic spaces are identified adjacent to this large cavity. The cavity contained 5 cc of clear fluid. The outer surface of the cyst was smooth and the inner surface was characterized by small papillary protrusions.

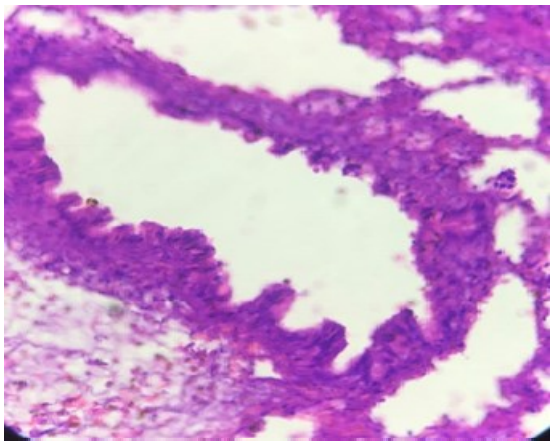


Macroscopic view of lobectomy specimen showing cystic spaces in CPAM

Microscopy: Multiple bits were taken and the slides were studied extensively and the following observations were made.



A. Microscopic appearance of Congenital Pulmonary airway malformation showing cystic spaces(Hematoxylin & Eosin x10)



B. Microscopic appearance of Congenital Pulmonary airway malformation (Hematoxylin & Eosin x 40)

Sections studied reveal, several dilated irregular bronchiole-like structures of variable size lined by flattened cuboidal to columnar epithelium. Also seen is a large cystic space lined by respiratory epithelium. Mucosal glands, cartilage, and inflammatory cells are absent. Areas of hemorrhage noted. There is a rim of normal pulmonary parenchyma seen at the periphery. With the above features, the diagnosis of a Congenital Pulmonary airway malformation of lung – Stocker type II was made out.

DISCUSSION

In 1949 Congenital cystic adenomatoid malformation (CCAM) was described by Ch'in and Tang and later on, in 1977 it is classified into 3 subtypes by Stocker *et al*.In 2002 Stocker named CCAM as Congenital pulmonary airway malformation (CPAM) and subdivided it into 5 typesbased on clinical and pathological features[5]. Among the 5 types, three are cystic and one is adenomatoid[1]. Congenital cystic adenomatoid malformation is generally a unilateral lesion. It encompasses 25% of all congenital lung lesions. CCAM consists of solid and cystic airless tissue with the absence of cartilage in the wall. The pulmonary lobe may be partially or completely involved[10]. The mechanism behind the development of CCAM is not exactly known. Few reports suggest the spontaneous resolution of CCAM (microcystic type) usually after 28weeks of gestation[11]. Malignancies like rhabdomyosarcoma and bronchioloalveolar carcinomamay be associated with large cystic type CCAM[12] The cause of Congenital cystic adenomatoid malformation is anomalous development of terminal respiratory structures, which results in adenomatoid proliferation of bronchiolar elements. There will be cyst formation which leads to enlargement of the affected lobe[2]. The terminal bronchiole proliferates abnormally and inhibits alveolar development and generates intercommunicating cysts[3]. Histopathologically, the cysts are lined by respiratory-type epithelium, and mucigenic epithelium [4]. Classification of Congenital pulmonary airway malformation(CPAM) by Stocker *et al* into five types[1,8].

CPAM type 0 - It is a rare type. There is acinar dysplasia and this type is incompatible with life. Type 0 is mostly seen in term and preterm infants. These newborns with type 0 are cyanotic at birth and they survive only for a few hours. Type 0 is associated with cardiovascular and dermal abnormalities. Grossly the lungs are small and firm and involve all lobes.

CPAM type 1: It is the most common type and it accounts for 65% of cases with a good prognosis for surgery. Grossly it consists of single or multiple cysts more than 2cm in size. Cysts are lined by ciliated columnar to pesudostratified tall columnar epithelium. Mucus-producing cells are seen in between these epithelial linings.

CPAM type 2: It consists of single or multiple medium-sized cysts. These cysts surround the normal parenchyma. Type 2 has a poorer outcome and is frequently associated with other anomalies. The characteristic feature is the back-to-back arrangement of bronchiole-like structures which are lined by cuboidal to columnar cells. Mucus cells and cartilage are absent except in the normal area. Rhabdomyomatous dysplasia is a variant of this lesion.

CPAM type 3: It consists of small cystic or solid areas. It consists of bulky mass-producing mediastinal shift and hypoplasia of the uninvolved lung. The size of the cyst is approximately 0.2cm. There are few scattered bronchiole-like structures seen. This type is associated with a bad prognosis.

CPAM type 4: Type 4 is hamartomatous malformations involving the distal acinus. There are large air-filled cysts lined by flattened alveolar cells. There is a good prognosis followed by resection.

CONCLUSION

The clinical symptoms of the patient depend on the extent of the lesion in the lung and other associated conditions. Bilateral lung

involvement is rare[2]. These malformations cause pulmonary hypoplasia that results in respiratory distress, pneumothorax, mediastinal shift, and pleural effusion.Congenital cystic adenomatoid malformation is associated with other anomalies such as cardiac, skeletal, renal, and gastrointestinal atresia[3]. With the help of an ultrasonogram, all cases of Congenital cystic adenomatoid can be detected by the 20th week of gestation[4]. The treatment is either by wedge resection or lobectomy[9]. We report this case for its infrequent occurrence and availability of only limited reports.

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