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Case Report

Paediatrics Section

Type 1 Congenital Pulmonary Airway Malformation (CPAM): A Case Report

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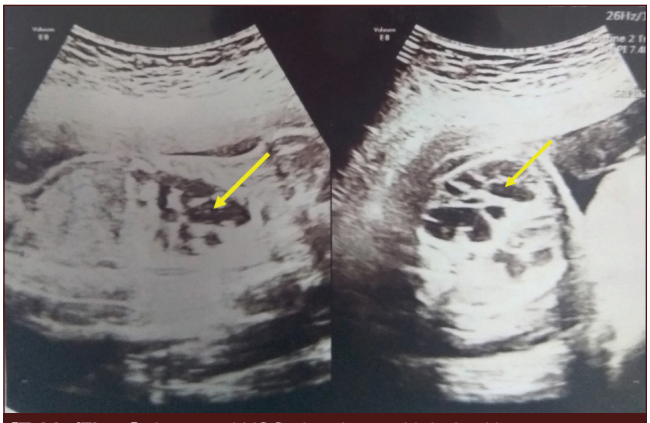
ABSTRACT

Congenital Pulmonary Airway Malformation (CPAM) is a lower respiratory tract developmental malformation. It is due to overgrowth of terminal bronchiole causing a dysplastic lesion. Affected areas consist of mass of cysts lined by bronchial or cuboidal epithelium, which may contain cystic and adenomatoid portions with intervening normal lung tissue. We present a rare case of full term female baby with type 1 CPAM, antenatally diagnosed and successfully managed with left lung lower lobe complete lobectomy with maximum cyst size measuring about 5.3x2.3 cm. Histologically cyst resembling bronchioles.

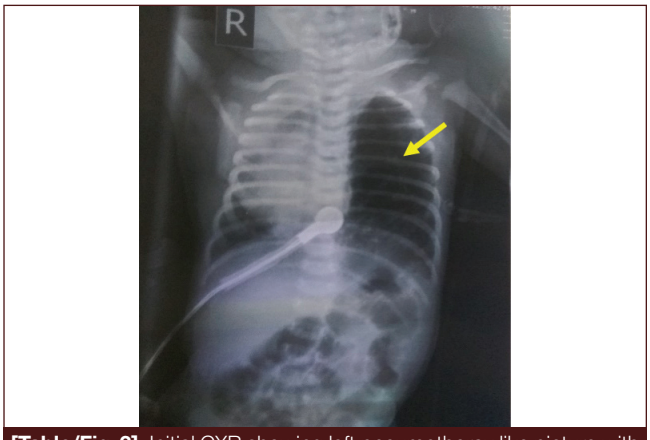
Keywords: Congenital cystic adenomatous malformation, Hamartomatous lesion, Lobectomy, Lung malformation

CASE REPORT

A full term five day old female baby with birth weight of 3.2 kg delivered vaginally was brought to NICU emergency with gasping and history of progressive hurried breathing since one day and one episode of convulsion at home with aspiration of milk. On admission, baby was found to be gasping with cyanosis and SpO_2 of 55% at room air. She was intubated and ventilated. Mother conceived at the age of 24 years, she was Gravida 3 Living 1 spontaneous abortion 1 at 3 months with no history of gestational diabetes,drug intake and consanguineous marriage. Family history was insignificant. On probing antenatal history, 3rd trimester antenatal scan showed enlarged fetal left lung, hyperechoic with multiple enlarged cysts with maximum cyst measuring about 5.3x2.3 cm suggestive of CPAM [Table/Fig-1]. Doctor explained about the cystic lesion and availability of surgical treatment postnatally. Chest x-ray was taken which was showed left pneumothorax like picture with mediastinal shift to right and dextrocardia [Table/Fig-2]. Baby improved with mechanical ventilation and other supportive measures and extubated after four days. Pediatric surgeon opinion was taken and planned for CT thorax. CT thorax was done which showed large multiple thin walled cysts which was suggestive of Type 1 congenital pulmonary airway malformation [Table/Fig-3]. Baby was prepared and taken up for left thoracotomy after taking consent from parents. Peroperatively, a cystic large air filled lesions without any obvious lung parenchyma was seen involving the entire left lower lobe. The upper lobe appeared normal, though compressed by the mass lesion. Left lower lobe complete lobectomy was done and drain placed

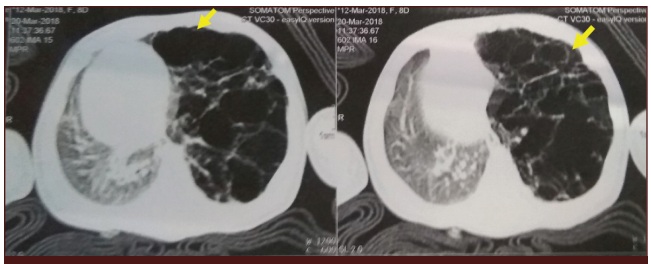


[Table/Fig-1]: Antenatal USG showing multiple fetal lung cysts.

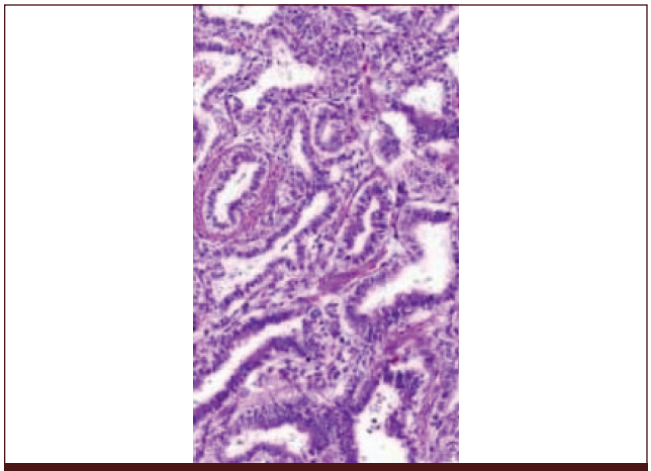


[Table/Fig-2]: Initial CXR showing left pneumothorax like picture with mediastinal shift.

in position. Macroscopically resected lung tissue showed multiple cysts. Histologically showing multiple sized cysts lined by pseudostratified cuboidal and columnar cells similar to type 1 CPAM [Table/Fig-4]. Baby improved clinically, extubated after 24 hours and showed good lung expansion by day four of operation. Baby was active and lactating, was discharged on 6th post-operative day, in near normal condition. Periodic follow-up has been carried since and the baby's clinical condition was normal.



[Table/Fig-3]: CT thorax showing left lung parenchyma replaced by multiple pleomorphic cysts with large cyst measures about 5.3 x2.3 cm. Cardia and mediastinum noted on right side with right parenchymal compression.



[Table/Fig-4]: Histopathology showing multiple varying sized cysts with pseudostratified cuboidal and columnar epithelial cells. (H&E 200X)

DISCUSSION

Lung malformations constitutes a spectrum of diseases that originates from embryonic period [1]. The rate of diagnosis of malformations in prenatal and neonatal period has increased in recent times. CPAM is a hamartomatous lesion of the lung [2]. It is due to overgrowth of terminal bronchiole causing a dysplastic lesion in the lungs [3]. The accepted pathogenesis is an abnormal airway patterning and branching during morphogenesis resulting in lung cyst appearance [4]. This condition was first reported by Ch'in and Tang where the term CCAM (Congenital Cystic Adenomatoid Malformation) was used [5]. CCAM is a rare condition, predominantly affecting the lower lobes of the lungs [3]. These lesions have also been referred to as CPAM and also, along with other variants, congenital lung and airway cysts and pulmonary sequestration, congenital thoracic malformations [5]. Unilateral CCAM is more common. This malformation can spontaneously regress, increase in size or cause non immune hydrops fetalis [5].

The incidence of CCAM has been estimated at 1 in 25,000-30,000 pregnancies [3]. It is not related to factors such as race, age or exposure, but occurs more in males with a ratio of 1.8:1 [6]. There are no recognised patterns of inheritance. The aetiology of CCAM remains obscure. CPAM is a multicystic lung mass due to terminal bronchiolar proliferation and suppressed alveolar growth. Its usual presentations are respiratory distress, cyanosis and grunting. On physical examination it will be similar to the features of pneumothorax like decreased breath sounds, hyper resonance and mediastinal shift to opposite side. Polyhydraminos and fetal hydrops can be seen in severe lesions. It is due to venacaval compression and decreased cardiac contractility. Some patients will present later with failure to thrive, recurrent chest infections, vomiting [7].

CPAM is classified into three types radiologically and into 5 types histopathological based on modified Stocker classification [8].

Type 1 (large cyst type) is the most common of these, accounting for approximately 50% of cases. It consists of multiple, thin-walled cysts, 2-10 cm, lined with ciliated pseudostratified columnar epithelium, which may contain mucus-secreting glands. Type 2 CCAM (intermediate cyst type) accounts for approximately 40% of cases and consists of multiple, smaller cysts (less than 1-2 cm diameter), which are lined by ciliated cuboidal or columnar epithelium without glandular tissue. Type 3 (small cyst type) lesions are rarer (less than 10% of CCAMs) and relatively solid lesions, which have very small cysts lined with ciliated cuboidal epithelium. In our case, its type 1 CPAM.

According to newer expanded stocker classification, there is addition of two more types ie, type 0 and 4. Type 0 occurs in <3% cases, arises from trachea and either do not have cysts or cysts <0.5cm, acinary dysplasia. It involves all lobes and is incompatible with life. Type 4 is seen in 10-15% of cases and has alveolar or distal acinar origin, peripheral cystic type [9].Type 0,1,2,3,4 constitutes the five histological based classification.

A simpler classification describing macrocystic (cysts >5 mm), tend to grow slowly have a favourable diagnosis and microcystic (cysts <5 mm) lesions when it becomes large and grow rapidly, they are associated with mediastinal shift, pulmonary hypoplasia and hydrops [10].

In about 25% cases of CPAM, an associated systemic/pulmonary arterial connection is noted. The majority of cases of CCAM are diagnosed by antenatal USG, usually at around 20 weeks' gestation.It appears as a hyperechogenic mass in microcystic lesions. Most have a good prognosis, if not associated with the

development of hydrops. The lesion may grow initially, reaching a maximum size at around 25 weeks gestation [11]. CCAM can be an incidental finding in older children and adults or as a result of repeated infection. The complications which can occur are fetal death, premature delivery, recurrent pneumonia, hemothorax, malignant change [12]. As investigation, chest radiography, CT scanning, Magnetic Resonance Imaging (MRI), prenatal ultrasonography, renal, cerebral ultrasonography and echocardiography in newborns are indicated [13].

The appropriate management of congenital cystic lung disease remains a matter for debate; however, surgical excision seems to be the treatment of choice to remove CCAM. Lobectomy is preferred over segmentectomy, because of the difficulty in distinguishing the extent of CCAM from normal parenchyma [14]. In the treatment of symptomatic CCAM patients, there is no controversy, but for the management of asymptomatic neonates and infants there is no clear cut management protocol regarding the decision and timing of an excision.

Termination of pregnancy may be considered with poor prognostic features such as bilateral CCAM, early hydrops or associated severe congenital anomalies [3].

Overall prognosis depends greatly upon the size of the lung mass and the pathophysiologic involvement [6]. Hydrops, polyhydraminos, mediastinal shift, type 3 disease, presence of other congenital malformations and preterm birth has a worse outcome [3]. In follow-up of our patient, she had no post surgical symptoms and her general conditions were great. It should be noted that timely clinical suspicion to the disease and appropriate imaging followed by prompt surgical management are the keys to prevent further complications and even death in affected cases.

CONCLUSION

CCAM is a potentially life threatening and reversible, rare congenital disorder which frequently presents with respiratory distress. In this case, even though antenatally diagnosed baby did not have distress at birth which misled and later presented with gasping, and timely intervention has saved the life.

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