Original Research Paper

Volume - 14 | Issue - 12 | December - 2024 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Histopathology

# CONGENITAL PULMONARY AIRWAY MALFORMATION :A CASE REPORT

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**ABSTRACT** Congenital pulmonary airway malformation (CPAM), formely known as Congenital cystic adenomatoid malformation (CCAM) is a rare congenital disorder of the fetal tracheobronchial tree.It accounts for 25% of all congenital lung malformations.Majority of cases are found prenatally or within first 2 years of life.We report here a case of CPAM in in a newborn discovered prenatally. At birth,the baby was having respiratory distress and was thus intubated. Left pneumectomy was performed on the 3 day old neonate.We present herewith gross and microscopic findings of the case.

**KEYWORDS :**

## INTRODUCTION

- Congenital pulmonary airway malformation (CPAM )was previously called as Congenital cystic adenomatoid malformation (CCAM) [1]
- It encloses a spectrum of Cystic and Non cystic malformations in lung which develops in uter
- Type 1 and 3 CPAMs are associated with KRAS mutations and have an increased risk of metastatic mucinous adenocarcinoma if incompletely resesected. Whereas ,Type2 CPAMs are associated with bronchial atresia and do not have KRAS mutations[2][3]
- The lesions are commonly detected on prenatal USG.
- But the definitive diagnosis requires pathological examination following resection

## CASE

A 35 year old, G3P1+1 presented to opd for routine antenatal checkup. The antenatal USG revealed a presence of mutiloculated cystic lesion measuring 8x4.9cm in the left hemithorax.The largest cyst measuring 5x4cm in size.Mediastinal deviation to the right was noted .The right lung parenchyma appeared normal.No e/o pleural or pericardial effusion was noted. Abdominal and visceral situs was normal.No other structural anomalies were noted and amniotic fluid was adequate for gestation .The imaging features were suggestive of CPAM (Macrocystic).

LSCS was performed at 37 weeks ,and a female newborn was delivered.The baby was found to have respiratory distress at birth and not maintaining the target oxygen saturation. Thus,baby was intubated and no further desaturation was noted.

On 3rd neonatal day, left complete pneumectomy was performed.

On gross pathological examination,the cut section revealed multiple cysts ranging in size from 4cm to 0.5cm with interspersed normal lung parenchyma in between.

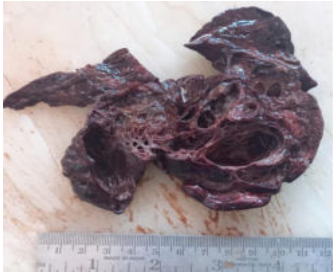

**Figure 1:** Cut section of the left lung shows multiple cysts and the intervening normal lung parenchyma

On microscopic examination,variable sized cysts noted lined by ciliated cuboidal to columnar epithelium with interspersed alveolar type spaces.Some of them having papillary projections.Few areas of mucinous cell clusters are seen.Thus a diagnosis of Type1 CPAM was made.

On follow up examination at 2 months,the baby is well with normal development

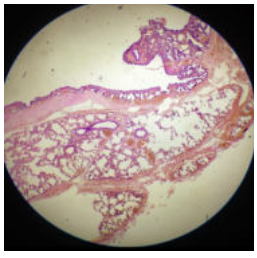

**Figure 2** H&E (low power) :Shows multiple irregularly shaped cystic spaces and intervening normal lung parenchyma

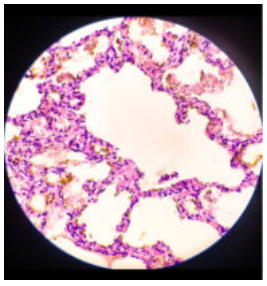

**Figure 3** H &E (high power):Shows the cysts lined by cuboidal to columnar epithelium

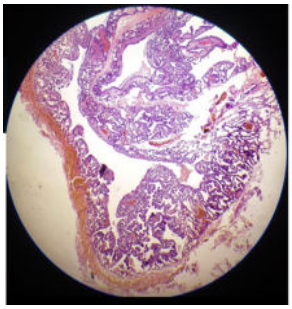

**Figure 4** (low power):Shows features of epithelial complexity

including frquent transition from columnar epithelium to alveolar type spaces,branching papaillae and irregularly shaped smaller cysts

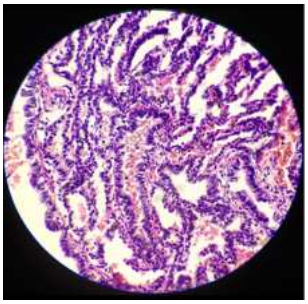

**Figure 5**(High power): Shows cysts lined by ciliated columnar epithelium

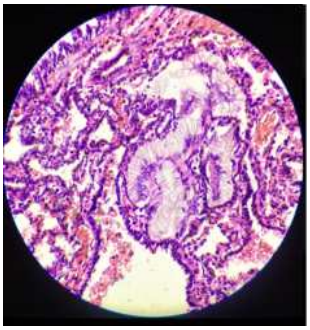

**Figure 6** H&E (high power): Shows foci of mucinous cell clusters

## DISCUSSION

Congenital pulmonary airway malformation (CPAM) is a congenital anomaly which includes a spectrum of cystic and non-cystic lung malformations that develop in utero.Earlier it was known as Congenital cystic adenomatoid malformations (CCAM).[1]Most commonly used classifications are Stocker and Langston classification systems[4].Stocker type 0 and type 4 CPAMs are no longer used,they represent other lung abnormalities[5].Type 2 CPAMs are most common ;usually asymptomatic;and are associated with bronchial atresia and do not have KRAS mutations.Type 1 and 3 CPAMs are usually symptomatic ;associated with KRAS mutations and have an increased risk of metastatic mucinous carcinoma if incompletely resected.CPAMs[6][3] ;most commonly involve a single lobe of lung.Rarely, 1 lesion may involve multiple lobes or a patient may have 2 distinct lesions.The clinical course is dependent upon the size of the lesion and timing of resection.Type 2 CPAMs are rarely associated with other congenital anomalies including congenital diaphragmatic hernias[7].Lesions are typically detected on prenatal USG.Multiple hypoechoic lung cysts are noted on USG.[8] But the definitive diagnosis requires histopathological examination.Complete surgical resection is the treatment.

Grossly,in Type1 (Large cyst )CPAM ;the largest cyst sixe is variable ranging in size from,0.5cm to >7cm with interspersed normal appearing lung parenchyma.[6]

In Type 2 (Small cyst)CPAM ,the cysts can measure upto 2.5cm with intervening normal appearing parenchyma.It may also have a mucocele or grossly evident bronchial atresia

Type 3 CPAMs consists of solid or dense appearing lesion,often with a lobulated appaearnce.It may have very small interspersed cysts[6]

Microscopically,Type1 CPAM consists of cysts lined by ciliated cuboidal to stratified columnar epithelium with interspersed alveolar type spaces[6].It often shows epithelial complexity such as papillary projections.in 75% of patients ,mucinous cell clusters are seen[9]

In type 2 CPAMs,cysts are typically round and lined by ciliated columnar epithelium[6].Epithelial complexity is rare.It often has evidence of mucostasis(including pools of mucin and foamy intra alveolar macrophages)

Type3 CPAMs consists predominatly irregular airway spaces lined by ciliated cuboidal to columnar epithelium.The surrounding septa often appears thickened with prominent mesenchyme.In 45% of patients ,mucinous cell clusters are seen multifocally with papillary or acinar architecture.[9]

PAS stain and MUC5 can be used to highlight the mucinous cells.The presence of mucinous cell clusters is highly specific for KRAS mutation but not entirely sensitive.The differential diagnosis to be considered for CPAM are type 1 pleuropulmonary blastoma,intrapulmonary sequestration and intrapulmonary bronchogenic cysts

## CONCLUSION

Congenital pulmonary airway malformation (CPAM)is a rare congenital entity.Majority of the cases are detected prenatally The clinical course depends upon the size of the lesion and timing of resection highlighting the importance of early diagnosis and treatment.

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