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Case Report Open Access

# Congenital Pulmonary Airway Malformation - A Case Report with Review of Literature

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**Abstract**

Congenital Pulmonary Airway Malformation (CPAM) is a rare hamartomatous lesion. There are five major clinicopathologic types, most of which are incompatible with life. We report a case of 75 year old man, who was relatively asymptomatic throughout his life till he was 74, and was diagnosed to have CPAM on investigations for chronic cough by open lung biopsy. A brief review of clinicopathologic features of various types of CPAM also done.

**Keywords:** Cystic lung disease; Congenital pulmonary airway malformation, Hamartomatous lesion

## Introduction

CPAM is a rare developmental anomaly of lower respiratory tract due to abnormal lung branching morphogenesis. The lesions are classified into five types according to the stages and levels of lung development affected. Most of these cases present in the new born period with respiratory distress and many are incompatible with life. A few of these cases may remain asymptomatic until later in life. We report a case of 75 year old man who presented with chronic cough and on lung biopsy was diagnosed to have CPAM. An analysis of histomorphogenesis of different types of CPAM along with literature review of adult presentation of CPAM is also done.

## Case Report

A 75 year old man presented with dyspnea on exertion for 1 year and associated cough. Cough was mostly dry without any positional variation. There was no significant past history of any illness or drug use. On examination, the patient had clubbing. There were coarse crackles on both sides, with predominance on the right side. Other systems were normal on examination. A clinical diagnosis of bilateral bronchiectasis was made and he was evaluated as per the standard protocol for bronchiectasis. Basic blood investigations were normal. Chest X-Ray PA view showed cystic lesions on both lungs involving mainly the upper zones. CT chest Plain showed multiple large air containing cysts in the right lung with intervening areas of sub segmental collapse/ground glassing/interstitial thickening in the right upper lobe (Figure 1).

With a revised clinical diagnosis of Langerhans cell histiocytosis, wedge biopsy was done from Right middle and lower lobes. The biopsy tissue measured 1.5 × 1.2 × 1 cm and 5 × 1.8 × 0.5 cm respectively. Pleura were seen on some aspects. Cut surface showed lung tissue with multiple cystic spaces of varying sizes, largest measuring 1.5 cm in largest dimension. Pale brown lung tissue was seen in between.

On microscopy, the sections showed lung with many closely packed cystic spaces of varying sizes mostly lined by ulcerated ciliated pseudostratified columnar epithelium. Some of the cysts were lined by flattened alveolar cells and multinucleated giant cells (Figure 2). Small bronchioles noted in some areas. Intervening areas shown fibrosis, smooth muscle bundles and mild patchy lymphocytic infiltration. There was no evidence of Langerhans Cell Histiocytosis. Special stains did not show any parasite. Immunohistochemistry with TTF1 was done which confirmed alveolar lining cells in a few of the cysts. The features were of Congenital Pulmonary airway malformation Type 1.

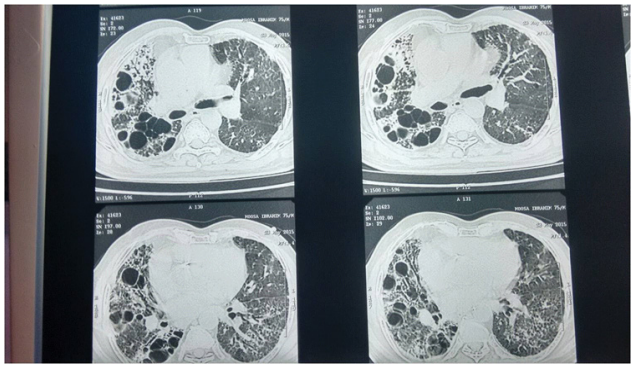

**Figure 1:** Chest X-Ray PA view showing cystic lesions on both lungs involving mainly the upper zones.

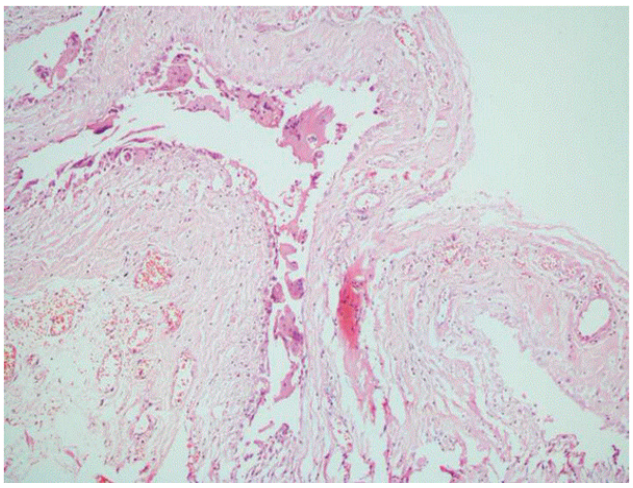

**Figure 2:** Microscopic sections showing lung with many closely packed cystic spaces of varying sizes.

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## Discussion

Congenital pulmonary airway malformation is a rare developmental anomaly of the lower respiratory tract [1]. The lesions are due to abnormal lung branching morphogenesis at different stages of lung development and these lesions are classified accordingly into five types from Type 0 to Type 4. These anomalies were previously called Congenital Cystic Adenomatoid Malformation(CCAM) [2]. Now, this term is not prefered as cystic change is seen only in 3 of 5 types of CPAM and adenomatoid change is seen in only one type. Five stages of fetal lung development are described -embryonic phase, pseudoglandular phase, canalicular phase, saccular phase, and alveolar phase [3]. CPAM types 0-3 originate during the pseudoglandular stage of lung development and type 4 originates during the saccular stage of lung development. CPAM is classified into 5 types, depending on the site of origin of the pathology. As the pathology moves from the bronchus, to bronchiole, then to alveolar tissue, the disease is classified from type 0 to type 4. Type 0 shows proximal bronchus like structures and is a malformation of the proximal tracheobronchial tree. Many mucous cells and extensive cartilaginous areas are features of Type 0 CPAM. Type 0 is rare and is incompatible with life. Infants are cyanotic at birth and do not survive more than a few hours. They usually have cardiovascular anomalies and adrenal hypoplasia [4].

Type 1 shows bronchus like and proximal bronchiole like structures. A few mucous cells can be seen. Compressed lung parenchyma may be seen in some areas. The wall shows thick fibro muscular tissue .The commonest type is Type I with 60-70% incidence. Type 1 patients usually present in the first month of life, but can be seen in older population including young adults. The outcome of Type 1 depends on the size of the cysts and if resectable, they have a good prognosis [5].

Type 2 is composed of bronchiole like structures and resembles bronchiolar segment of the acinus. Type 2 is usually associated with other severe developmental anomalies. They present in the first year of life and has a poor outcome [5].

Type 3 is composed of bronchiole like structures, alveolar ducts and saccules corresponding to midacinar region. Type 3 has high mortality rate as the lesion is of large size and is associated with maternal polyhydramnios and fetal anasarca. Type 3 is seen exclusively in the first days to months of life and shows male predominance [5].

Type 4 with thin walled structures lined by alveolar cells suggesting distal acinar Malformations. The epithelium varies from pseudostratified to cuboidal to low-cuboidal and simple squamous with an overlap between the different types. Type 4 accounts for 10-15% of cases and is seen from new born period to 4 years. The lesion affects single lobe in 80% cases and has an excellent survival after resection [5].

Type 1 and type 4 CPAM, which have larger cysts, and are difficult to differentiate from cystic pleuropulmonary blastoma (Type I) which is a cystic or solid sarcoma seen in children. Pleuropulmonary blastoma(Type I) also shows presence of rhabdomyoblasts and cartilage nodules. Most of the cases of CPAM are diagnosed during prenatal or during immediate postnatal period [6].

Very few cases (about 50) of adult CPAM are reported. Only five cases of patients above 50 years are published and to the best of our knowledge no case reports on patients above 70 years of age in the literature. Our case showed dilated cysts with pseudostratified columnar epithelial lining and fibro muscular wall, consistent with bronchial lining. A few mucinous cells also noted. These features were of Type -1 CPAM.

## Conclusion

The patient was started on symptomatic management with mucolytics, steam inhalation and was also administered pneumococcal and influenza vaccines. He became asymptomatic and at the time of review after a year, he continues to be free of symptoms. This case is being presented to highlight a rare disease and even rarer presentation at a very late phase in life. For more than six decades, the patient was asymptomtic, despite having a congenital disease with extensive lung parenchymal involvement with giant cysts, and only having minimal normal lung tissue for sustaining his physiological requisites of respiration.

### References

1. Hussain Aliya N (2012) Congenital pulmonary airway malformation (CPAM): Thoracic Pathology (1st edn), Elsevier Saunders:15-17.
2. Dipti B, Bindu A, Daniel Z, Raj MD, Birendra S (2015) Congenital pulmonary airway malformation in an adult male: A case report with literature review. Case Reports in Pulmonology.
3. Stocker JT, Mani H, Husain AN (2013) The Respiratory Tract, In: Stocker JT, Dehner LP, Husain AN. Paediatric Pathology (3rd edtn), Lippincott Williams & Wilkins, Wolters Kluwers, Netherlands, 201: 441-515.
4. Hyun JK, Ji-Yong M, Sa-Il K, Kim TH, Sohn JW, et al. (2012) Congenital cystic adenomatoid malformation with bronchial atresia in elderly patients. Tuberc Respir Dis (Seoul) 72: 501-506.
5. Taştekın E1, Usta U, Kaynar A, Ozdemır C, Yalçin O, et al. (2014) Congenital pulmonary airway malformation type 2: a case report with review of the literature. Turk Patoloji Derg 44: 278–281.
6. Noguchi M, Schimosato Y (2010) Pulmonary Neoplasms, In: Mills SE, Carter D, Greenson JK, Reuter VE, Stoler MH (editors) Diagnostic Surgical Pathology (4th edn) Wolters Kluwer/Lippincott Williams & Wilkins, Netherlands pp: 1053-1093.