Congenital Pulmonary Airway Malformations in a Tertiary Health Care Facility in North Kerala - A Retrospective Study

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Abstract

Congenital pulmonary airway malformation (CPAM) is a rare congenital disorder presenting in infancy. Most of the cases are diagnosed during antenatal period or during infancy. However, few cases live up to adulthood and detected during an infectious complication. Primary objective of the study is to find the prevalence of CPAM among hospitalized patients. Secondary objective is to study the clinical and radiological profile of these patients. In this retrospective study hospital data base is searched for the diagnosis of this condition for a period of 5 years. Four cases were diagnosed during this period, one in infancy and other three in adulthood. The conclusion is that all cases of CPAM are not diagnosed in infancy and it may be due to lack of awareness among clinicians about this condition.

Keywords: Congenital Cystic Adenomatoid Malformation; Congenital Pulmonary Airway Malformation; Cysts

Introduction

Congenital pulmonary airway malformation (CPAM) is a rare condition characterized by immature, malformed lung tissue with cystic appearance. CPAM is seen mainly in newborns and is rarely encountered in older children and adults. However, few adult cases of CPAM were reported in literature. It is seen that number of CPAM diagnosed in this region are less when the overall prevalence is considered. These cases are either missed in infancy or alternate diagnosis is considered. Minimally symptomatic patients live up to adulthood and experience frequent pneumonia, lung abscess formation and life threatening haemoptysis. This study is an attempt to look at the diagnosed cases of CPAM in a tertiary care facility in North Kerala.

Aim of the Study

1. Primary objective of the study is to find out the prevalence of CPAM among hospitalized patients in a tertiary health care facility in North Kerala.
2. Secondary objective is to study the clinical and radiological profile of adult CPAM patients.

Study Methodology

This is a retrospective data base analysis of hospital records for 5 years from 2014 - 2018. Electronic data of a tertiary care facility in North Kerala was searched for words “congenital cystic adenomatoid malformation” and “congenital pulmonary airway malformation”. Matched records were analyzed for demographic data, clinical presentations, investigations and management. These data were reviewed by 2 independent consultants, and cases in which both agreed with the clinic diagnosis were included in the study. Permission from the hospital administration and institutional ethics committee was obtained prior to the study.

Results

During the 5-year period, 4 matches were retrieved from the hospital database, one for congenital cystic adenomatoid malformation and 3 for congenital pulmonary airway malformation. The two observers agreed with the clinic diagnosis in all 4 cases. All the cases were females. One case was an infant aged four months where the diagnosis was achieved by histopathology. All other cases were adults ranging from 26 years to 58 years. These cases were diagnosed by the clinical data and CT appearance of cysts conforming to CPAM.

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<td>Multiple large cysts in right upper lobe and left lower lobe</td>
<td>Based on CT findings</td>
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*Table 1: Demographic and clinical data of patients with CPAM.*

Case-1

A 4-month-old infant was having respiratory symptoms since birth and presented to the emergency department with respiratory distress. Term baby born to otherwise healthy parents was having cough and feeding difficulty since birth. She had a short febrile illness followed by severe breathlessness. X-ray chest showed shift of mediastinum to left with large multi-cystic lesion in the right lung. CT Thorax confirmed multiple large cysts involving the right lung. Surgical removal of the cysts was done, and histopathology showed features of CPAM Type-1.

*Figure 1: 1A: X-Ray chest AP view showing large cystic lesion on the right side with shift of mediastinum to the left. 1B: CT Scan of the infant showing multiple cysts on the right side pushing the mediastinum to left causing compression on the left lung. (Courtesy- Ravindran Chetambath, Paediatric Respiratory Illnesses-2nd Edition (2014), Macmillan Medical Communications).*

Case-2

A 58-year-old female patient presented to the OPD with complaints of persistent cough and expectoration since past 3 months. There was a history of intermittent episodes of hemoptysis associated with low grade fever. She also had a history of dyspnoea on exertion. There was history of recurrent episodes of similar illness for the past 10 years. She used to get cough, scanty sputum, haemoptysis and fever, 2 - 3 times every year. CT Thorax done in 2014 reported it as bronchiectasis. She was under follow up treatment as a case of bronchiectasis for the past 4 years. She also gave history of anti-tuberculous treatment (ATT) twenty-five years back, details of which were not available. She is a known hypothyroid patient on thyroid replacement therapy. X-ray chest showed few cystic lesions in the right mid and upper zone (Figure 3). HRCT thorax showed a large cyst in the right upper lobe surrounded by a bunch of small cysts. Surgery was advised which the patient refused. She was put on conservative medical management and regular follow up.

Case-3

This 46-year-old female patient was admitted in a local hospital with a diagnosis of severe pneumonia and was ventilated for 7 days. Once improved, she was referred for persistence of shadows on the right side. Patient is symptomatic for the past 25 years with frequent cough, scanty sputum, progressive breathlessness and occasional haemoptysis. She had another episode of pneumonia 2 year back and was intubated for 3 days. Chest X Ray showed a cystic lesion in the right lower zone. CT Thorax taken at the time of pneumonia was reviewed which showed a large cyst surrounded by multiple small cysts in the right lower lobe. Evidence of resolving pneumonia on
the left side is also seen in the CT. A CT angiogram was taken to rule out sequestration and no systemic blood supply was detected. Since the patient already had two episodes of life threatening pneumonia she was advised to undergo vaccination against pneumococci and influenza. She was advised regular follow up.

Case-4

A 26-year-old female patient was referred with a history of fever and cough with purulent expectoration for 6 days. She was managed in a local health facility for 5 days and referred as the clinical response was not satisfactory. She also has exertional breathlessness. Patient was symptomatic since early childhood and was managed as a case of bronchiectasis. She used to get cough, scanty sputum, and fever, 2 - 3 times every year. CT Thorax done in 2011 reported it as bronchiectasis. X-ray Chest showed few cystic lesions in both the lungs (Figure 5). On review her HRCT thorax showed multiple cysts in the right upper lobe. Multiple cysts were also noted in the left lower lobe. On both sides one large cyst is seen surrounded by a bunch of small cysts (Figure 6). Lung parenchyma surrounding the cysts appeared normal. There is no evidence of fibrosis, mucus plugging or volume loss. Spirometry showed restrictive pattern. Since there is bilateral involvement surgical intervention is not preferred and put on conservative medical management.

Discussion

Ch’in and Tang in 1949 first described this developmental anomaly as congenital cystic adenomatoid malformation (CCAM) [1]. It was classified into 3 subtypes, and later in 2002 Stocker renamed it as congenital pulmonary airway malformation (CPAM) and classified into 5 types [2]. CPAM is an unusual condition characterized by immature, malformed lung tissue with cystic appearance. CPAM is seen mainly in newborns, still born infants and is an unusual condition in older children and adults. The patients with CPAM can present as neonates with severe, progressive respiratory distress due to cyst expansions. Exact etiology of CPAM is not known, it is to be considered as hamartomatous malformation and abnormal proliferation of the pulmonary tissue at different sites. The symptoms and prognosis depend on the degree of pulmonary involvement [3-5]. The clinical presentation spectrum varies from asymptomatic cases, recurrent pneumonia, or respiratory insufficiency [6]. In severe cases, patients develop respiratory failure even in the neonatal period, but others have a benign clinical course, with the manifestation of recurrent pneumonia [3-5].

In this series 4 cases were identified during a 5-year surveillance. Considering the overall prevalence of 1 in 30000 live births, this seems far from reality. 80% to 85% of cases are recognized in the first 2 years of life, adult presentation being uncommon [7]. This may be since many cases escape proper diagnosis. Only one case of neonatal CPAM was reported and it presented as rapidly expanding cysts with pressure effect on the opposite lungs. All other cases were adult cases suggesting that their diagnosis was not made at early life either due to minimal symptoms or due to a mistaken diagnosis of bronchiectasis [8]. Most CCAM in adults involve unilateral lobes of the lung and may be complicated with pulmonary bacterial infections and abscess [9,10]. In this series one patient had bilateral involvement whereas all the other three patients had single lobe involvement.

Lesion involving bilateral lobes of the lung is also reported. In CCAM associated literatures, a few bilateral CCAM cases in adult patients have been reported [11-13]. Bilateral CCAM may appear like interstitial pneumonia because of similar CT scan presentations showing grid-like opacity through the entire lung fields. The extensive involvement of the lesion increases the risk and complications of surgery. Therefore, most patients with such lesions are treated with conservative treatment after diagnosis is confirmed by lung biopsy.

Stocker type I and IV has good prognosis whereas mortality is very high in Type II and III. Microscopically, the large, thin walled cysts are lined by ciliated pseudostratified columnar epithelium with some mucin producing cells. The wall composed of fibromuscular and elastic tissue and occasional cartilage plate. The association between CPAM and malignancy has been well documented. Malformation and proliferation cause hamartomas over the tracheobronchial tree. Type I CPAM may involve malignant transformation of mucinous bronchioloalveolar carcinoma [14-16]. Type II CPAM may involve malignant transformation to rhabdomyosarcoma. Type III CPAM requires examination of the entire lesion to exclude pulmonary blastoma by confirming whether sarcomatous differentiation is present in the solid parts [17].
Management of CPAM is surgical resection in early life. Adult cases once diagnosed may be subjected to surgery as chances of malignant transformation is high. Bilateral lesion is a surgical concern due to risk of life-threatening complications. Those who do not undergo surgery may be conservatively managed with proper control of infections, vaccination and pulmonary rehabilitation.

Conclusion

During the five-year surveillance, only 4 cases of diagnosed CPAM is retrieved from hospital records. This appears to be too low considering the prevalence of this congenital anomaly. This may be due to failure to diagnose the condition in the infancy or making an alternative diagnosis such as cystic lung disease or bronchiectasis. Being a rare disorder classified under orphan diseases, awareness among medical community is to be improved to diagnose more cases among newborn. HRCT give a good diagnostic tool to point out a diagnosis with good sensitivity. This case series is an eye opener that there are cases of CPAM in the community and a proper evaluation in the neonatal period will improve case detection rate.

Bibliography
