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Original Article

Clinicopathological Study of Congential Pulomonary Airway Malformations

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Abstract

Introduction: Congenital pulmonary airway malformation (CPAM) is a rare developmental abnormality of the lung. The underlying feature of a CPAM is an excessive over growth of terminal respiratory bronchioles and forming cysts of various sizes. This abnormal lung tissue is of defective epithelial-mesenchymal architecture.

Aim: The aim of the study is to study the various modes of presentation of CPAM, the effectiveness of antenatal ultrasonogram and pathology of these lesions.

Materials and Methods: All patients with radiologically proven CPAM were included in this study. The patients were subjected to detailed clinical examination, and relevant investigations were performed, namely, chest X-ray, and computed tomography (CT) scan chest. CPAM diagnosed cases were undergone thoracotomy and excision of affected lobe.

Results: Of these 20 patients, seven were antenatally diagnosed. Six patients presented with fever and cough, CT scan detected 16 cases of CPAM, lower lobe was affected in 11 patients, in our series, 94% of cases belong to stocker Type I lesion.

Conclusion: An early intervention of CPAM patients causes less morbidity.

Key words: Antenatal ultrasound, Congenital pulmonary airway malformation, Stocker's pathological types

INTRODUCTION

Congenital pulmonary airway malformation (CPAM) is a rare developmental abnormality of the lung. They are similar to benign lung tumors.[1] Congenital lesions of lung are rare, with overall incidence of 1/10000 to 1/25000 births.[2] The underlying feature of a CPAM is an excessive over growth of terminal respiratory bronchioles and forming cysts of various sizes. There are different types of lesions (types 0-3), some associated with cystic areas and adenomatous overgrowth of the terminal bronchioles.[3] This abnormal lung tissue is of defective epithelial-mesenchymal architecture. The congenital lesions are detectable of prenatal ultrasound. They appear as solid or cystic masses. Mode of presentation of CPAM varies widely in antenatal and neonatal period.

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Aim

The aim of the study is to study the various modes of presentation of CPAM, the effectiveness of antenatal ultrasonogram (USG) and pathology of these lesions.

MATERIALS AND METHODS

This prospective study was conducted in Institute of Child Health and Hospital for Children, Madras Medical College, Chennai, Tamil Nadu, India. All patients with radiologically proven CPAM were included in this study. Other cystic lesions of chest were excluded from the study. The patients were subjected to detailed clinical examination, and relevant investigations were performed, namely, chest X-ray, and computed tomography (CT) scan chest. CPAM diagnosed cases were undergone thoracotomy and excision of affected lobe was done 17 patients. The treatment modalities were studied and patients were followed up to

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assess the effectiveness after 6 months of surgery with relevant investigations and extended to the available period. The results were tabulated and analyzed.

RESULTS

Of these 20 patients, seven were antenatally diagnosed. The majority five of antenatally detected patients were asymptomatic at the time of presentation. Remaining two patients were admitted with complaints of having respiratory distress since birth. In this study, 11 patients were male, and the remaining nine were female. There is no sex predilection for patients with CPAM.

Among our study group of 20 patients, seven patients presented in the neonatal period, 11 presented between 1 month and 1 year, two patients presented between 1 and 5 years. No patient was seen above 5 years of age. This means that 90% of CPAM patients in our study are below 1 year of age. High degree of suspicion is needed to detect CPAM in infants if they are not antenatally diagnosed (Table 1).

In our study, two patients had congenital heart disease, and one patient had glycogen storage disease Type III. In our study, there was no association with bronchopulmonary sequestration (BPS).

In the patients who were enrolled for study, 18 underwent at least one antenatal USG. Two patients did not have any antenatal USG. Among this group of 18, only seven were suspected to have cystic lesion in lung. This means that only 39% patients were detected antenatally. Of the 18 patients who went antenatal USG, only one patient had polyhydramnios. However, this infant was not detected to have CPAM antenatally.

CPAM presents in various forms. In our study, five asymptomatic patients, all these five were antenatally detected. Three patients presented with complaints of respiratory distress since birth. Among these three patients, two patients presented on the 1st day of life. One patient with respiratory distress since birth presented on 34th day of life.

Six of our patients presented with fever and cough. Among this group, three were initially treated outside as bronchopneumonia, two patients as empyema and one as loculated pyopneumothorax. Four CPAM patients presented with difficulty in breathing and two presented with wheeze (Table 2).

Patients with empyema and loculated pyopneumothorax were initially treated with intercostal chest tube drainage. Only later they were found to have CPAM.

All of our patients had chest radiography at the time of admission. Radiologists were able to suggest the diagnosis of CPAM in only ten patients. For two patients they gave the diagnosis of lung cyst, and for five patients they gave the diagnosis of bronchopneumonia. Two patients were diagnosed to have empyema and one was diagnosed to have loculated pyopneumothorax (Table 3).

CT scan was done in 18 of our CPAM patients. Among these children, 16 were diagnosed to have CPAM, one was found to have BPS, and one was diagnosed as loculated pyopneumothorax. This shows postnatal CT scan has a higher diagnostic value for detecting lung lesions (Table 4).

In our study group, nine patients had right sided lesions, ten patients had left-sided lesions and one had bilateral CPAM (Table 5).

Lower lobe was affected in 11 patients, upper lobe two patients, middle lobe two patients, and three patients had

Table 1: Distribution study patients in age group

Age distribution	Number of cases (%)
Up to 1 month	7 (35)
1 month to 1 year	11 (55)
1-3 years	1 (5)
3-5 years	1 (5)
5-12 years	0 (0)

Table 2: Distribution of presenting complaints

Presenting complaints	Number of cases
Asymptomatic	5
Respiratory distress since birth	3
Fever with cough	6
Dyspnea with cough	4
Cough with wheeze	2

Table 3: Distribution of X-ray findings

X-ray findings	Number of patients
CPAM	10
Bronchopneumonia	5
Lung cyst	2
Empyema	2
Pyopneumothorax	1

CPAM: Congenital pulmonary airway malformation

Table 4: Distribution of CT findings

CT findings	Number of patients
CT scan picking up CPAM	16
BPS	1
Loculated pyopneumothorax	1

BPS: Bronchopulmonary sequestration, CPAM: Congenital pulmonary airway malformation, CT: Computed tomography

both upper and middle lobe involvement (Table 6). Lower lobectomy was done in ten patients (right - 3; left - 7), upper lobectomy was done in 5 patients (right - 4, left 1), and middle lobectomy was done in five patients. Among the above patients combined upper and middle lobectomy was done in three patients. One patient who presented with empyema also underwent decortication.

Post-operative period was uneventful in eight patients, 4 patients had wound infection and 2 patients need post operative ventilatory support. Antenatal detected patients were operated after 1 month of age.

Average duration of hospital stay for operated patients was 14.4 days. However, the average hospital stay duration of antenatal detected patients was significantly less. It is about 10 days for antenatal detected patients. Hence, it is clear that infected CPAM patients had longer hospital stay.

In our series, 94% of cases belong to Stocker Type I lesion and 6% of cases belong to Stocker Type III lesion (Table 7).

In our series, there is no mortality among operated patients. Among the 17 patients who we have operated, 15 patients turned up for follow-up. Out of this, two patients were treated for respiratory tract infection. Their chests X-ray are normal.

DISCUSSION

In our study, among antenatally detected seven patients, only two were symptomatic at birth. These two patients were admitted with complaints of having respiratory distress since birth. In study done by Stanton *et al*., among antenatally detected 505 neonates, 16 (3.2%) became symptomatic in infancy.[4]

Table 5: Distribution of laterality of lesions

Side of lesions	Number of patients
Right side lesions	9
Left side lesions	10
Bilateral lesions	1

Table 6: Distribution of lobes involved

Lobes involved	Number of patients
Upper lobe	2
Upper and middle lobe	3
Middle lobe	2
Lower lobe	11

Table 7: Distribution of pathological types

Pathological types	Number of cases
Stocker Type I	16
Stocker Type III	1

Most of the children operated in our study were in <5 years age group, early resection strategy to avoid the onset of symptoms. Truitt *et al*., argued that the risks associated with congenital lung lesions (infection and malignancy) justify intervention in the asymptomatic patient.[5]

In our study, two patients had congenital heart disease, and one patient had glycogen storage disease Type III congenital cystic adenomatoid malformations are usually isolated and sporadic although they have been associated with other anomalies (most commonly cardiac and renal) in 15-20% of cases.[6]

In our study, fever with cough was the most common presenting symptom in <7 years children. In study done by Giubergia *et al*., the most common presenting symptoms were respiratory distress in children under 6 months of age (40%) and recurrent pneumonia in older ones.[7]

Screening X-ray confirmed the diagnosis of CPAM in 50% of the patients only, whereas CT scan had higher chance of diagnosing CPAM 89%. In study done by Patz *et al*., approximately 25% of lesions diagnosed as CAM may be either pulmonary sequestration or bronchogenic cysts. Overlapping CT features can also exist among other foregut malformations.[8]

In our study, lower lobe was affected in 11 patients and single lobe was affected commonly. Puri in endorses this by saying involvement is usually unilobar which is slight predilection for the lower lobes.[1]

In our study, infected CPAM patients had longer hospital stay than patients who underwent elective surgery. This was confirmed Stanton *et al*., who said for all ages, elective surgery was associated with significantly less complications than emergency surgery.[4]

Stocker *et al*., found pathological Type I lesions were the most common CPAM variety, accounts for 50-70% of diagnosed cases. In our study, we found maximum number of cases were of Type I cases, accounts for 96% of the patients.[3]

CONCLUSION

Most of the patients presented with symptoms of fever with cough and dyspnea. Antenatally detected patients, reported to the hospital earlier and they were mostly asymptomatic. Ability of X-ray to detect CPAM was

only 50%. CT scan has a greater success rate in picking up CPAM cases. Lower lobe involvement is significantly higher. Stockers Type I is the predominant type (94%) in our series.

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