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Original Research Article

Congenital Lung Abnormalities: Differential Diagnostic Findings, Surgical Management and Radio-Pathologic Correlation

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Abstract:

Background and Objectives: A variety of developmental abnormalities of the tracheobronchial tree and pulmonary parenchyma are found in the newborn. There is limited data available on their presentation and clinical course from third world countries.

Methods: This was a retrospective data review, conducted at a tertiary care hospital in northern India. The medical records of those patients diagnosed with CLM from June, 2014 to June, 2018 were evaluated. This study was undertaken after due approval by the Institutional Review Board of Sher-I-Kashmir Institute of Medical Sciences (approval no: 80/2018, date: 18.06.2018). The study population included children diagnosed as having CLM based on their clinical, radiological, and pathological features. Surgical details which included the timings of their surgical intervention, the type of surgery, the duration of the operation, intraoperative findings, and their intraoperative and postoperative complications were recorded. Patients, who fulfilled inclusion criteria, gave consent for surgery were admitted and were subjected to surgery. A total of 30 cases were taken.

Results: Cystic lucencies were observed in all CPAM patients on chest radiograph; in 2 (n=3) patients of CLE; in all patients of BC; in 1 (n=2) patient of PS; in 1 (n=2) patient of BA whereas no cystic lucencies were observed in all PPB patients on chest radiographs. Mediastinal shift to opposite side was observed in 7 patients of CPAM; in 2 patients of CLE; in 1 patient of BC; and was not observed in any patient of PS, BA and PPB. USG chest indicated a significant finding in 73.3% of the patients. Cystic lesion was the most common finding present in 36.6% of the patients. most common procedure performed was left lower lobectomy in 23.3% of the patients followed by right upper lobectomy in 16.7% patients; right lower lobectomy in 16.7% patients; right middle lobectomy in 13.3% patients; left upper lobectomy in 10% patients;

Conclusion: Congenital pulmonary airway malformation was the most common type of lung abnormality in this investigation. The most frequent method of presentation was recurrent respiratory tract infection, either with or without respiratory distress. Prenatal ultrasound awareness and emphasis can be increased through health education, which can prevent unwarranted delays in diagnosis and treatment. Surgery has strong long-term results and is curative.

Keywords: Congenital pulmonary airway malformation, lobectomy, bronchogenic cyst, USG, congenital lobar emphysema.

Introduction

A variety of developmental abnormalities of the tracheobronchial tree and pulmonary parenchyma are found in the newborn. Congenital pulmonary airway malformations (CPAM), Congenital lobar emphysema (CLE), Bronchial atresia (BA), bronchogenic cysts(BC), pulmonary sequestrations (PS), and are examples of CLMs. [1]

Numerous instances share characteristics with CPAM, including as modifications to a sequestered lung region known as hybrid lesions. [2,3] The combined incidence of these uncommon congenital abnormalities is 30-42 occurrences per 100,000 people. [4] With the advent of near-routine prenatal ultrasonography, a great deal has been learned about the natural history and pathophysiology of Congenital Lung Malformations (CLM's). The traditional understanding is that the tracheobronchial tree and proximal gastrointestinal tract arise from common foregut anlagen. [5]

Ultrasonography (USG) is the primary imaging modality for fetal screening. It provides valuable information about the presence and size of a focal

lung lesion (lesion volume) [6], mass effect, associated hydrops fetalis, lung hypoplasia, and other organ malformations, all of which affect the prognosis and management. [7,8] Fetal magnetic resonance (MR) imaging is helpful—in select cases—for lung volume quantification, in addition to evaluation of the lesion.

This study was carried out to share our experiences from a tertiary care center, which is the only facility in the area treating such anomalies, because of the rarity of congenital defects and the dearth of literature from the developing world.

Materials and Methods

This was a retrospective data review, conducted at a tertiary care hospital in northern India. The medical records of those patients diagnosed with CLM from June, 2014 to June, 2018 were evaluated.

This study was undertaken after due approval by the Institutional Review Board of Sher-I-Kashmir Institute of Medical Sciences (approval no: 80/2018, date: 18.06.2018). The study population included children diagnosed as having CLM based on their clinical, radiological, and pathological features. We excluded syndromic children and those with major cardiac malformations.

Data on their demographic profiles such as their age and gender, clinical presentation, baseline investigations, chest imaging studies (X-ray and computed tomography), hospital admissions, surgical interventions, surgical outcomes and follow-ups were retrieved. Any other relevant investigations such as bronchoscopy or dye study findings were also noted. Surgical details which included the timings of their surgical intervention, the type of surgery, the duration of the operation, intraoperative findings, and their intraoperative and postoperative complications were recorded. Patients, who fulfilled inclusion criteria, gave consent for surgery were admitted and were subjected to surgery. A total of 30 cases were taken.

After establishing the diagnosis of congenital lung malformations, these patients were taken for surgery. Prior to surgery, cardiac and respiratory optimization was attempted, including treatment of infections, pneumonia, etc.

Surgical Methodology

Posterolateral thoracotomy was performed in all the cases optimized for surgery. Data of surgeries was recorded (operation, operative time, intraoperative findings, amount of blood loss, any intraoperative blood transfusion, any intraoperative complication). Postoperatively, patients were managed in paediatric surgery ward. Ventilatory support, was provided, and the time of ventilatory support was noted. Patients were observed for any immediate post-operative complications (bleeding, pneumothorax, atelectasis, lung collapse, infections, pneumonia, etc).

Patients were followed up initially, weekly for the first month, followed by, monthly for six months. During the follow up, patients were assessed for any residual disease; chest wall deformity like scoliosis, restrictive lung diseases; etc.

Statistical Analysis

The final data were recorded on a predesigned study proforma and was managed in Microsoft Excel. Data analysis was performed using SPSS software. The values of various parameters are presented as mean±SD, in absolute numbers and as percentage.

Results

Table 1: Presenting signs on admission of patients with congenital pulmonarymalformations

Signs	CPAM N=18	CLE N=3	BC N=3	PS N=2	BA N=2	PPB/?PPB N=2
RTI/Pneumonia	11	1	1	1	1	1
Fever	4	1	1	2	0	0
Difficulty Breathing	4	2	0	0	0	1
pneumothorax	4	0	0	0	0	0

As per table 1 most of the patients presented with respiratory tract infection/pneumonias (53.3%), fever (26.6%), difficulty breathing (23.3%), pneumothorax (13%).

Table 2: Chest Radiography Findings of patients with congenital lungmalformations

CLM	CPAM(N=18)	CLE(N=3)	BC (N=3)	PS (N=2)	BA (N=2)	PPB/?PPB(N=2)
Cystic Lucencies	18	2	3	1	1	0
Mediastinal shift opposite side	7	2	1	0	0	0
Presence of infiltrates	10	1	2	1	1	0
Pneumothorax	4	0	0	0	0	0
Atelectasis	4	1	2	0	1	0

As per table 2 Cystic lucencies were observed in all CPAM patients on chest radiograph; in 2 (n=3) patients of CLE; in all patients of BC; in 1 (n=2) patient of PS; in 1 (n=2) patient of BA whereas no cystic lucencies were observed in all PPB patients on chest radiographs. Mediastinal shift to opposite side was observed in 7 patients of CPAM; in 2 patients of CLE; in 1 patient of BC; and was not observed in any patient of PS, BA and PPB. Presence of infiltrates was observed in 10 patients of CPAM; 1 patients of CLE; 2 patients of BC; 1 patient each of PS and BA; and was not observed in any patient of PPB. Pneumothorax was observed in 4 patients of CPAM whereas no patient of other types had pneumothorax. Atelectasis was observed in 4 patients of CPAM; 1 patient of CLE; 2 patients of BC and 1 patient of BA; no atelectasis was observed in PS and PPB on chest radiographs.

Table 3: Ultrasonography findings of study patients

USG	Frequency	Percentage
Positive Findings	22	73.3
Consolidation	10	33.3
Cystic lesions	11	36.6
Pleural effusion	10	33.3
Pneumothorax	4	13.3
Atelectasis	3	10
Non-Significant	8	26.6

As per table 3 USG chests indicated a significant finding in 73.3% of the patients. Cystic lesion was the most common finding present in 36.6% of the patients. Consolidation was detected in 33.3% patients, pleural effusion was detected in 33.3% patients, pneumothorax was detected in 13.3% patients and atelectasis was detected in 10% patients.

Table 4: Distribution of study patients as per CT findings

CT findings	Frequency	Percentage
Cystic Lesion	22	73.3
Bronchiectasis	3	10.0
Emphysema	2	6.7
Bronchial Atresia	1	3.3
AVM	0	0.0
Pulmonary Sequestration	1	3.3
Pleural Effusion	8	26.7
Pneumothorax	4	13.3
Empyema	0	0.0
Cavitating Lesion	4	13.3
Mediastinal Shift	1	3.3
Atelectasis	3	10.0
Consolidation	10	33.3
Cystic Lesion With?	3	10.0
Pleuropulmonary Blastoma	1	3.3

As per table 4 The most common finding on CT chest was cystic lesions present in 73.3% of the patients, followed by consolidation in 33.3% patients; pleural effusion in 26.7% patients; pneumothorax in 13.3% patients; bronchiectasis, atelectasis each in 10% of the patients. Pleuropulmonary Blastoma was present in 3.3% patients and mediastinal shift in 3.3% patients.

Table 5: Distribution of study patients as per surgical procedure

Surgical procedure	Frequency	Percentage
RPLT with upper lobectomy	5	16.7
RPLT with middle lobectomy	4	13.3
RPLT with lower lobectomy	5	16.7
LPLT with upper lobectomy	3	10.0
LPLT with middle lobectomy	1	3.3
LPLT with lower lobectomy	7	23.3
RPLT with upper and middle lobectomy	1	3.3
RPLT with excision of cyst	2	6.7
LPLT with excision of cyst	1	3.3
ICTD placement	1	3.3
Total	30	100
RPLT= Right posterolateral thoracotomy; LPLT= Left posterolateral thoracotomy; ICTD = Intercostal chest tube drainage		

As per table 5 most common procedure performed was left lower lobectomy in 23.3% of the patients followed by right upper lobectomy in 16.7% patients; right lower lobectomy in 16.7% patients; right middle lobectomy in 13.3% patients; left upper lobectomy in 10% patients; excision of cyst was performed in 10% patients (combined left and right) and only ICTD placement was performed in 1 patient.

Table: 6 Congenital lesions, number, sex distribution, age of patients at surgery,presentation, site of lesion, surgery, and diagnostic investigation (n = 30)

Pathology	No. ofcases (%)	Age at Surgery	Sex M:F	Presenting Symptoms	Site of lesion (No. of patients)	Surgery	Diagnostic Test
CPAM	18 (60%)	2 months	8:10	Respiratory	RUL(5)	Lobectomy of affected lobe(s)	USG (30%)
		to 4 years		distress, respiratory	RML(3)		
		(mean age : 12.4months)		tract infection	RLL(5) LUL(1)		CT (70)
					LML(1)		
					LLL(1)		
BC	3 (10%)	1 month 4 months	0:3	Recurrent chest infections	RUL;2 LUL;1	Cyst Excision	?USG (33.3) CT (100)
		9 months					
CLE	3 (10%)	2 months 3 months 2 months	1:2	Respiratory distress, chest infection	LUL;2 LLL:1	Lobectomy of affected lobe(s)	CT (100)
PS	2 (6.7%)	2 years 2 years	1:1	Fever, chest infection	LLL;2	Lobectomy of affected lobe(s)	CT (100)
BA	2 (6.7%)	14 months 2 years	1:1	Fever, Respiratory tract infection	RML;1 LLL;1	Lobectomy of affected lobe(s)	CT (100)
PPB	1 (3%)	18 months	0:1	Respiratory distress, fever on and off	LLL;1	Lobectomy of affected lobe (s)	CT (100)
1 more patient suspected of PPB also died during preoperative optimization.							

As per table 6 CPAM was the most common CLM found in 60% of the patients. Age distribution was 2 months to 4 years with a mean of 12.4 months. Male: Female ratio was 4:5.

Most common symptom was respiratory distress/respiratory tract infection. Thoracotomy of the affected side was performed in all patients. Lobectomy of the affected lobe(s) was performed in all patients. Most common site of lesion was RUL and RLL. Bronchogenic cyst was found in 10% of the patients. Age distribution was 1 month to 9 months with a mean of 4.6 months. All patients of Bronchogenic cyst were females. Most common symptom was recurrent chest infections. Thoracotomy of the affected side was performed in all patients. Pulmonary Sequestration was found in 2 (6.7%) of the patients. Both patients were of same age 2y (24 months).

Male: Female ratio was 1:1. Fever with or without chest infection was the most common symptom. Thoracotomy of the affected side was performed in both the patients. Lobectomy of the affected lobe was performed in both patients. Lesion was found in LLL in both the cases.

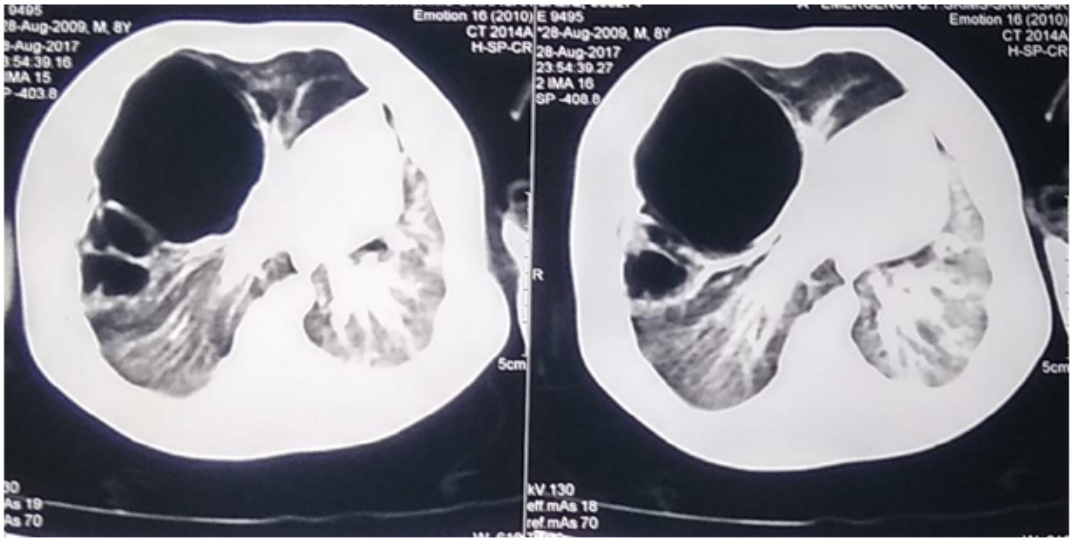

Figure 1: CT chest showing large cyst in right upper lobe CPAM

Discussion

In our study there were a total of 30 cases; 18 cases (60%) were Congenital cystic adenomatoid malformation (CPAM), 3 cases (10%) were Congenital Lobar Emphysema (CLE), 3 cases (10%) were Bronchogenic Cyst (BC), 2 (6.7%) cases each of Pulmonary Sequestration (PS) and Bronchial Atresia (BA), 1 (3.3%) histologically proven case of Pleuropulmonary Blastoma (PPB) type II and 1 suspected case of pleuropulmonary Blastoma.

These findings are corresponds to findings Basant kumar's [9] report who reported 56% of cases as CPAM; 20% as CLE; 12% as PS; and 12% as BC. Our findings are supported by Jean Marie [10]. These findings are further supported by MacSweeney [11] who reported 56% of cases as CPAM; and 18% of cases as Pulmonary Sequestration.

In our study the pre-operative diagnosis of congenital lung malformations on chest x- ray showed cystic lucencies in all patients of CPAM; in 66.6% of CLE patients; all patients of BC; 50% of PS patients; 50% of BA patients. The pre-operative diagnosis on chest X-ray showed mediastinal shift to opposite side in 38.8% of CPAM patients; in 66.6% of CLE patients; and in 33.3% of BC patients. The pre-operative diagnosis on chest X-ray showed pneumothorax in 22.2% of CPAM patients. The pre-operative diagnosis on chest X-ray showed Atelectasis in 22.2% of CPAM patients; 33.3% of CLE patients; 66.6% of BC patients; 50% bronchial atresia patients. These findings are confirmed by Jean Marie et al [12] and Donald H Hulinde [13].

The results were consistent with the study conducted by Woo Sun Kim [14] in which 90% of the patients had cystic lesions. The results were also supported by the study conducted by Junpei Ikezoe [15] who conducted a review of CT scans of 23 patients of bronchopulmonary sequestration and found that only 4 patients were diagnosed on the basis of CT findings combined with appropriate history. The results were also supported by the study conducted by Lena N. Naffaa [16] who studied imaging findings in 4 patients of Pleuropulmonary Blastoma and found that all patients had findings suggestive of Pleuropulmonary Blastoma. In our study we found that 22 (73.3%) of the patients were diagnosed preoperatively, whereas in 8 (26.7%) of the patients, the diagnosis was inadequate or doubtful.

While the decision making for children with symptomatic CLM's is reasonably straight forward and that is resection of the abnormality, there is on-going debate regarding the need for and the timing of surgery in children with asymptomatic lesions. For those who support surgical intervention, the safety of pulmonary resection in infants and children is no longer a source of debate as the outcomes are generally good. Complication rates after surgery range between 6% and 9% and are mostly related to prolonged air leak. [17] Mortality is a very rare occurrence in experienced hands and is thus not a valid outcome measure.

In our study, the most common procedure performed for CLM's was Lobectomy which is approved by studies of Rothenberg [18] and Cano I [19] who have found in their studies that complete resection of congenital lung lesion is achieved by lobectomy and remains the treatment of choice in many centers. Lobectomy for CLM's is also favoured by studies of Muller20 and Khosa21 who after their respective studies have recommended formal lobectomy over segmental resection. Traditionally, lobectomy occurred as an open surgical procedure through posterolateral thoracotomy.

Conclusion

CPAM is the most common CLM in paediatric age group, in the Kashmir valley, with a female preponderance. Recurrent respiratory tract infections with or without distress is the most common mode of presentation and any patient with these symptoms should be evaluated by USG chest in periphery or CT if available to rule out CLM's. Antenatal USG capabilities are paramount to the early diagnosis of CLM's and should be enhanced in the peripheries. Surgery is the treatment of choice in all CLM's and should be performed as soon as possible after preoperative optimization. Complete gross removal of disease is necessary to prevent/decrease the chances of malignancy/recurrence and should be always kept in mind.

Pathologist's reports should describe the lesion, including the type, as described in widely used classifications as some lesions require more aggressive follow up. Use of thoracoscopic approaches for removal of CLM's having proved to be beneficial and should be started. Patients with CLM's who have been operated are an important group for education, and should be made to understand the need for regular follow up, so that recurrence/transformation to malignancy can be detected and treated in time.

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