Original Article

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Surgery Section

Thoracic Mass Lesions in Children and their Management: A Prospective Interventional Study

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ABSTRACT

Introduction: Thoracic mass lesions in children are heterogeneous group of disorders, out of which developmental malformations such as Congenital Pulmonary Airway Malformation (CPAM), bronchogenic cysts and Congenital Lobar Emphysema (CLE) are common pathology. Primary pulmonary malignancies like carcinoid tumour and pulmonary blastoma may rarely be encountered.

Aim: To analyse the clinical pattern, diagnosis and management of children with thoracic mass lesions.

Materials and Methods: This prospective interventional study was conducted from June 2018 to May 2021, on 19 patients diagnosed and operated for thoracic mass lesions. It was carried out in the Department of Paediatric Surgery, Sardar Vallabhbhai Patel Post Graduate (SVPPG) Institute of Paediatrics, Srirama Chandra Bhanja (SCB) Medical College, Cuttack, Odisha, India. The final diagnosis was only based on histopathological study. The parameters assessed were demographic features, operative findings and pathological diagnosis. Statistical analysis was done using Microsoft Excel Q1 Macros software. Data was presented as mean, median with Confidence Interval (CI) and percentages (%).

Results: A total number of 19 cases were operated, out of which 12 were boys and seven were girls. The median age at presentation was three month (range: five days to five years). Right side of thorax was more commonly affected in 14 cases and left side in five cases. CLE was the most common pathology (eight cases) followed by CPAM (six cases). Bronchogenic cysts and lymphangioma were detected in two cases each. Pulmonary blastoma was the only malignant lesion found. There was no mortality in the present study.

Conclusion: The patients with thoracic mass lesions usually present before the age of six months and CLE was detected as the most common finding. A conclusive diagnosis can be obtained only from histopathology.

Keywords: Congenital lobar emphysema, Congenital pulmonary airway malformation, Pulmonary blastoma

INTRODUCTION

Thoracic mass lesions in children are heterogeneous group of disorders, which may be solid or cystic. The incidence is reported as 30-42 cases per 100,000 individuals and they constitute 5-18% of congenital malformations [1]. Paediatric lung masses are ten times more likely to be benign developmental lesions than a neoplasm. Among the primary lung neoplasms, malignancies outnumber benign tumours with a ratio of 3:1 [2]. The common childhood thoracic mass lesions include CPAM, bronchogenic cyst, pulmonary sequestration and CLE [2].

In spite of the rarity, CPAM is described as the most common pulmonary malformation [3-5]. Although, it was reported since 1949 by Ch' in and Tang, the exact aetiology of this malformation is still unknown [4,6]. It is described as proliferation of terminal respiratory bronchioles resulting in formation of cystic and non cystic pulmonary mass [5-7]. CLE was first reported by Nelson in 1932 and is described as hyperinflation of one or more lobes of lung causing compression of adjacent lobe and mediastinal shift [8-11]. The aetiology is unknown in 50% cases. Bronchial cartilage dysplasia, extrinsic bronchial compression by vessels or cysts and endobronchial obstruction are important causes in rest of the cases [5,7,9]. Bronchogenic cysts are also rare anomalies with incidence of 1 in 42,000 to 68,000 individuals [12].

The rarity of these malformations and lack of adequate exposure pose specific problems in the management in newborns and infants. Studies on thoracic masses during childhood are also limited. Thus, the present study was carried out with an objective to analyse the clinical pattern, diagnosis and management of children with thoracic mass lesions, operated in a tertiary care hospital, Cuttack, India.

MATERIALS AND METHODS

This prospective interventional study was conducted in the Department of Paediatric Surgery, SVPPG Institute of Paediatrics, SCB Medical College, Cuttack, Odisha, India, from June 2018 to May 2021. Institutional Ethics Committee approval was obtained (IEC/36/2020/128) and study was carried out accordingly. Patients admitted with thoracic mass lesions were taken as study population.

Inclusion criteria: Patients below the age of 14 years, who were operated for thoracic mass lesions and a confirmatory diagnosis was made by histopathological study were included in the study.

Exclusion criteria: Children in whom a provisional diagnosis was made based on chest radiography but, surgery was not performed due to refusal by parents and patients who left against medical advice before completion of treatment were excluded from the study.

Study Procedure

All participants complying to the above criteria within the three year period were included (n=19) by purposive sampling. The diagnosis was based on clinical and radiological features. It was confirmed by intraoperative findings and further histopathological study. The parameters compared were demographic profile, operative findings and histological features in different types of thoracic mass lesions.

Routine blood investigations and X-ray chest were done. Computed Tomography (CT) scan of chest was done for better delineation of the mass. A provisional diagnosis was made and surgery was planned under general anaesthesia. Thoracotomy was performed on the affected side of chest. Intraoperative findings were noted in detail and excised specimens were sent for histopathological study to the Department of Pathology. Intensive Care Unit (ICU) and ventilatory support was provided according to need. Postoperative

radiography of chest was done to find out lung expansion. After recovery, children were discharged and advised to attend Outpatient Department (OPD) after two weeks. Subsequent follow-up was made every month for three months, followed by every six months thereafter. A detailed clinical examination and routine haematological investigations were done during each visit. Chest radiography was performed at third month follow-up to check lung expansion. The total follow-up duration ranged from six months to three years.

STATISTICAL ANALYSIS

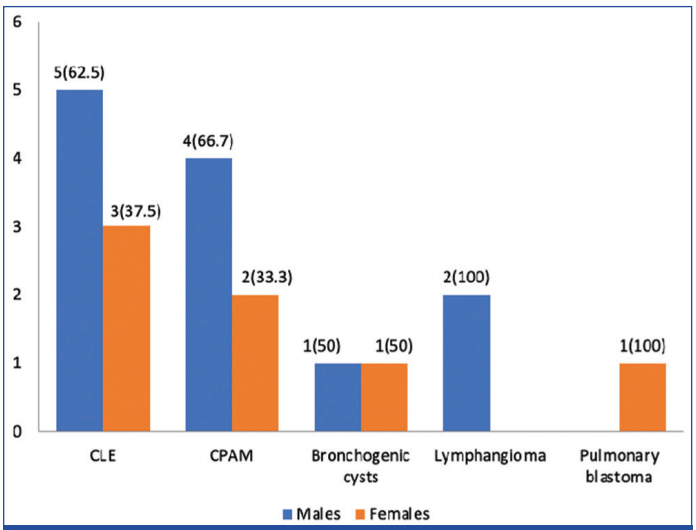
Statistical analysis was done using Microsoft Excel Q1 Macros software. Data was presented as mean, median with CI and percentages (%).

RESULTS

A total number of 19 thoracic mass lesion cases were operated during the three years study period [Table/Fig-1]. The median age at presentation was three months and ranged from five days to five years. Newborns accounted for 8 (42.10%) cases and 15 (78.94%) patients were below the age of six month. The median weight was 3.8 kg (range: 1.6-13 kg). Right side of thorax was involved in 14 cases and left side in five cases. There were 12 males and seven females; M:F=1.7:1 [Table/Fig-2]. CLE was the most common in pathology (eight cases) followed by CPAM (six cases). Bronchogenic cysts and lymphangioma were detected in two cases each. Pulmonary blastoma was the only malignant lesion found in this study.

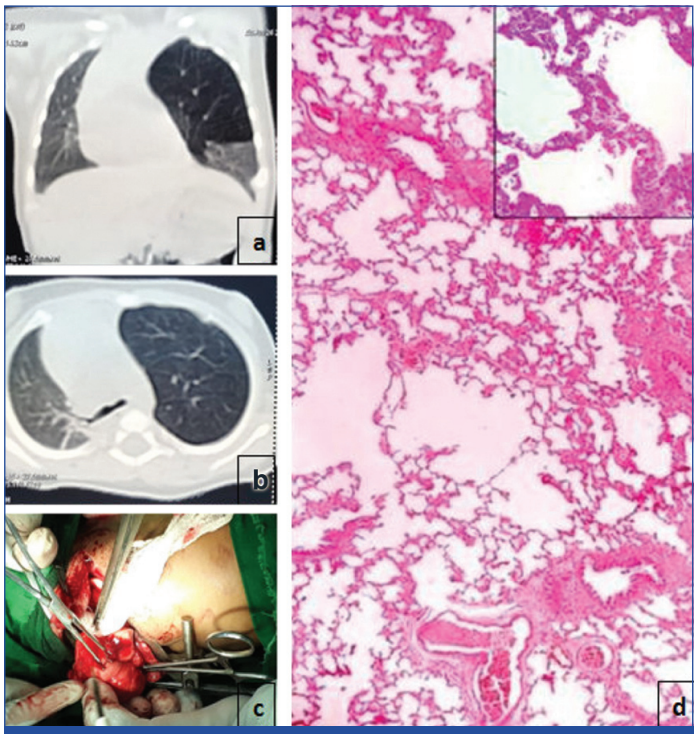
Thoracic mass lesion	No. of cases (%)	Median age at presentation	Median weight (kg)	Site (Right side/ Left side)
CLE	8 (42.1)	2 months	3.15	RUL-4, RML-2, LUL-2
CPAM	6 (31.6)	3 months	4.15	RML-2, RLL-2, LLL-2
Bronchogenic cyst	2 (10.5)	2.5 years	8.4	RML-1, RLL-1
Lymphangioma	2 (10.5)	3.5 years	12.7	R-1, L-1
Pulmonary blastoma	1 (5.3)	5 days	2.5	RLL
Total	19	3 months	3.8	R-14, L-5

[Table/Fig-1]: Thoracic mass lesions operated during the study period.
CLE: Congenital lobar emphysema; CPAM: Congenital pulmonary airway malformation bronchogenic cysts; R: Right; L: Left; RUL: Right upper lobe; RML: Right middle lobe; LUL: Left upper lobe; RLL: Right lower lobe; LLL: Left lower lobe



[Table/Fig-2]: Gender distribution {n(%)} in the study group.
CLE: Congenital lobar emphysema; CPAM: Congenital pulmonary airway malformation bronchogenic cysts

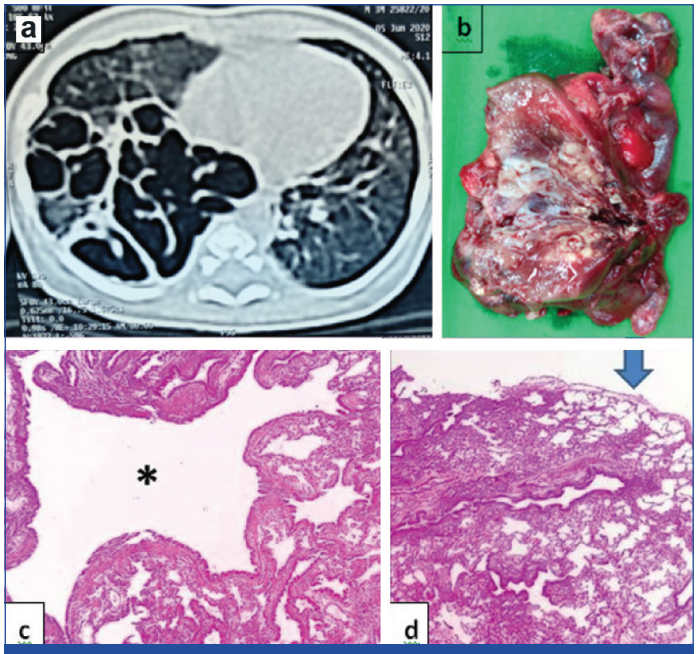
Congenital lobar emphysema: The mean age at presentation of CLE patients was two months (range: one month to four months, CI: 1.26-2.97). Male infants were more commonly affected than females (M:F=1.67:1). The mean weight was 3.29 kg (range: 1.6-4.5 kg, CI: 2.67-3.88). All the patients presented with respiratory distress. X-ray chest followed by CT scan was done in all cases [Table/Fig-3]. Intraoperatively, Right Upper Lobe (RUL) was found to be most commonly affected (four cases) followed by Right Middle Lobe (RML) and Left Upper Lobe (LUL); two cases each. Thoracotomy and resection of the involved lobe was performed. All the patients required postoperative ventilatory support. Complications included pneumonia in two cases, which required change of antibiotics and wound infection in one case, which was settled with daily dressing. However, they recovered and X-ray chest showed compensatory lung expansion. The histopathology study of resected lobes revealed distension of alveolar spaces. The infants were discharged with advice to attend OPD for follow-up after two weeks. All the patients were clinically stable at first follow-up. At three and six month follow-up, they were asymptomatic, gaining weight and chest radiography revealed normal lung growth.



[Table/Fig-3]: A three-month-old female infant presented with respiratory distress: a) CT scan coronal view showing hyperinflation of Left Upper Lobe (LUL) and compression of lower lobe; b) Axial CT window revealed crossing of overinflated lobe to opposite side and mediastinal shift; c) Intraoperative picture of emphysematous Left Upper Lobe (LUL) through thoracotomy incision; d) Photomicrograph shows distended alveolar sacs without destruction of septa (H&E, 10x). Inset: Lining of dilated sac by flattened simple squamous cells. Consistent with congenital lobar emphysema (H&E, 40x).

Congenital pulmonary airway malformation: The age at presentation in CPAM patients varied from seven days to six months, with a median of three months (CI: 1.52-4.79). The mean weight at presentation was 3.83 kg (range: 2.5-4.7 kg, CI: 3.12-4.53). Male babies were twice more commonly affected than females. Cough along with fever was the presenting feature in four cases and respiratory distress was the mode of presentation in two neonates. Right side of lung was affected in four cases (RML: two cases, Right Lower Lobe (RLL): two cases) and Left lower lobe in rest two cases. Thoracotomy of the affected side and lobectomy of the involved lobe was performed in all cases [Table/Fig-4]. Postoperatively, they were kept in ICU with ventilatory support. Complications included pneumonia in one case and wound infection in another case. The first case required change of antibiotic and second one required daily dressing. All the patients were discharged and advised to attend OPD with histopathology report. The histology showed cystic structures of variable size. The linings were composed of pseudostratified columnar epithelium

with interspersed mucus cells. During first follow-up at two weeks of discharge, the infants were found to be asymptomatic with normal chest findings. At subsequent three and six follow-ups, they were found to be clinically stable and chest X-ray showed compensatory pulmonary growth.

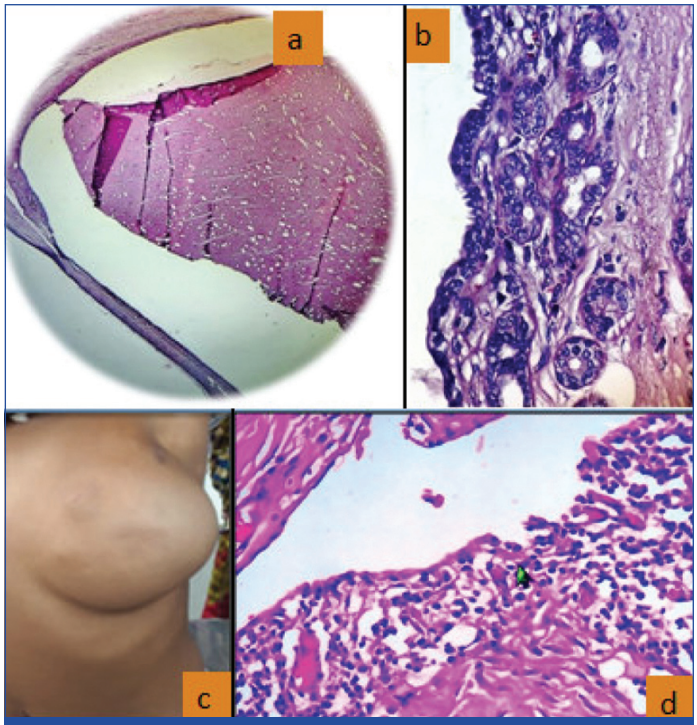


[Table/Fig-4]: A three-month-old male infant presented with cough and fever: a) CT scan chest showing multiple cysts in Right Lower Lobe (RLL); b) Thoracotomy and excision of Right Lower Lobe (RLL) done; c) Photomicrograph shows variably sized cystic structures in lung parenchyma. One dilated cyst lined by pseudostratified columnar epithelium (*); d) Interspersed mucus lining cells (arrow). Suggestive of congenital pulmonary airway malformation (H&E, 40x).

Bronchogenic cyst: Out of two cases of bronchogenic cyst, one patient was a 12-day-old newborn female and other was a five-year-old male child. The newborn presented with respiratory distress in second week of life and the child presented with fever along with cough. RLL and RML were the sites of involvement in these cases respectively. Right side thoracotomy and excision of the cyst was done in both the cases. The newborn required postoperative ventilatory support and the child maintained good saturation with oxygen inhalation. Histopathology revealed bronchogenic cyst in these cases [Table/Fig-5a,b]. The patients recovered well and discharged.

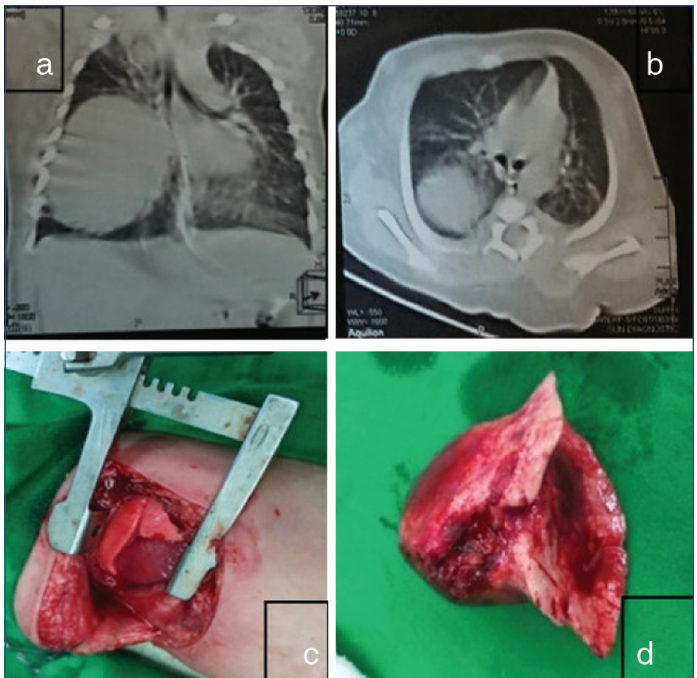
Lymphangioma: Both the patients of lymphangioma were males, with a mean age of 3.5 years and mean weight of 12.7 kg. Right side of chest was affected in one case and left side in another case [Table/Fig-5c,d]. Thoracotomy and resection of lymphangioma was done in both the cases. None of these patients required postoperative ventilatory support. Postoperative complications included prolonged lymphatic fluid leak from chest tube in one case and wound infection in another case. Histopathological examination showed flattened benign endothelial lining cells over fibrotic scanty stroma infiltrated with aggregates of lymphocytes. The cavity of the cystic structures contained floating lymphocytes in proteinaceous fluid like materials revealing diagnosis of lymphangioma. They were discharged and during follow-up both the patients were clinically stable. There was no recurrence on radiological examination.

Pulmonary blastoma: A five-day-old female newborn presented with respiratory distress and X-ray chest plain showed homogenous opacity in right lower zone. Ultrasonography suggested hypoechoic lesion without vascularity. This prompted us to do CT scan, which detected a well marginated soft tissue mass of size 4.5×5 cm in right lower lung [Table/Fig-6]. During thoracotomy, the tumour was found to involve entire lower lobe and it was completely resected. Histopathology and subsequent Immunohistochemistry (IHC) revealed

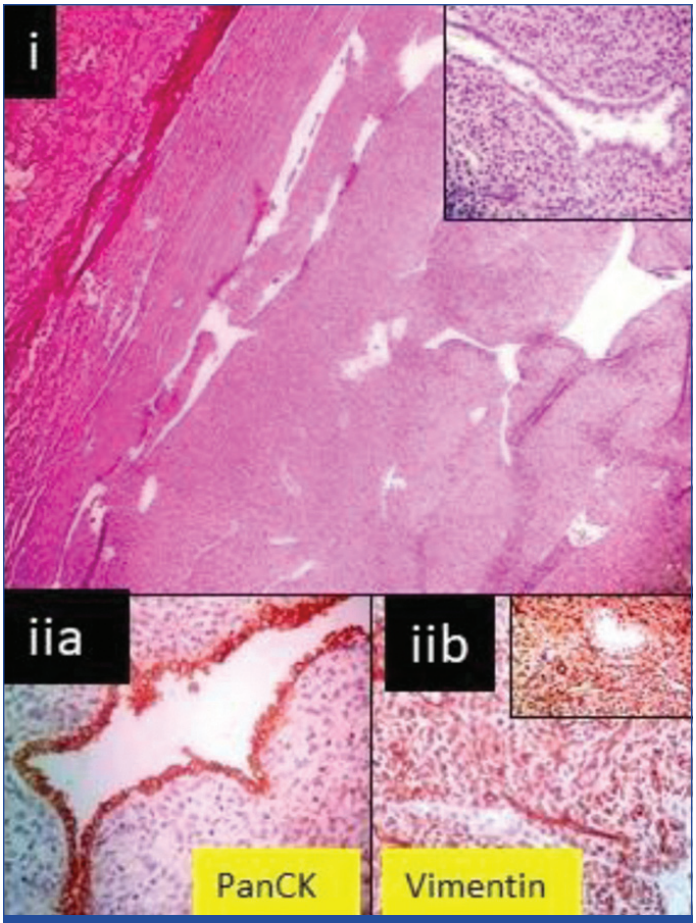


[Table/Fig-5]: a) Unilocular cyst containing viscous fluid (H&E, X40); b) The cyst wall recapitulates the bronchial lining with variable amounts of seromucinous glands consistent with diagnosis of bronchogenic cyst; c) Photograph of a four-year-old child with lymphangioma; d) Large irregular vascular spaces lined by flattened bland epithelial cells and the collagenous stroma contains lymphocytes (black arrow) suggestive of lymphangioma (H&E, 400X).

biphasic pulmonary blastoma, a rare pathology [Table/Fig-7]. Biphasic appearance of the tumour was based on detection of epithelial and primitive stromal components. Epithelial components showed branching glandular tubules resembling fetal lung. Stromal components seemed primitive blastematous cells. IHC revealed strongly positive PanCK on glandular cells and stromal cells were vimentin positive. Neuroendocrine differentiation was not noticed in the case by negative chromogranin and synaptophysin markers. During first follow-up at two weeks of discharge, the neonate was totally asymptomatic. On subsequent follow-up, she was clinically stable, gaining weight and chest radiography showed normal lung growth.



[Table/Fig-6]: A five-day-old female neonate presented with respiratory distress (a&b) CT scan chest showing a well marginated soft tissue mass in Right Lower Lobe (RLL) region; c) Right side thoracotomy-intraoperative photograph; d) Excised pulmonary tumour.



[Table/Fig-7]: Histopathology of the excised pulmonary tumour (i) Well circumscribed, unencapsulated tumour mass lying at the interface of normal lung tissue (H&E, 40X); (Inset) branching glandular structure lined by malignant cells with sub- and supranuclear glycogen vacuoles (H&E, 400X); (ii a) Pancytokeratin positive in glandular cells; (ii b) Vimentin positive in stromal cells and negative in glandular lining cells (Inset), consistent with diagnosis of pulmonary blastoma.

DISCUSSION

Although, patients below the age of 14 years were included, the maximum age observed in this study was five years. CPAM is reported as most common pulmonary malformation in most of the literatures [3-5]. However, in the present study it was the second common finding following CLE. Similar to the existing literature it was found that, lung masses in children are mostly benign developmental lesions than a neoplasm, as 94.7% cases were benign developmental malformations [2]. Pulmonary blastoma was the only malignancy observed in the study. Males outnumbered females, which may be due to the fact that, male infants were more often presented to us. Overall, right side of thorax was predominantly affected, a finding which was not highlighted previously.

Congenital lobar emphysema is a rare anomaly with reported incidence of 1 in 20,000-30,000 births [13]. Although, a male preponderance reported in some studies, there is no clear consensus regarding gender preference [8,11,14]. A male preponderance was marked in the present study. The mean age at presentation was two months in this study. But a higher mean age was reported in other studies [Table/Fig-8] [8,11,14]. Most of the newborns and infants present with respiratory distress [5,8,11]. Similar observation was also made in the present study. It has been frequently reported that, LUL is most commonly affected followed by RML and RUL [5,7,11]. However, a different scenario was observed in this study. RUL was most frequently affected in the study (50% cases). A bilobar involvement was reported by Nazem M and Hosseinpour in 17% of their cases, but only unilobar involvement was seen in this study [11]. The complications like pneumonia and wound infection were similar to other studies [11,14]. Surgical excision is the treatment of choice and postoperative mortality is reported in 3-7% cases [13]. A higher mortality rate (13.3%) was observed in series by Nazem M and Hosseinpour [11]. However, there was no mortality among CLE patients in the present study. This may be a reflection of gradual understanding about this disease, refinement in surgical practice and ventilatory support.

The commonly quoted incidence of 1 in 11,000-35,000 live births about CPAM seems to be underestimated [15]. Due to widespread and advancement of antenatal screening, the disease is increasingly recognised. A recent population based prospective study detected an incidence of 1 in 7,200 live births [6]. So, the incidence is in rising trend. Pneumonia is the usual presenting feature but, neonates may present with respiratory distress [3,5]. This was well experienced in the present study, as both of our neonates presented with respiratory distress. The disease may not be noticed till childhood, when they present with recurrent chest infection [6]. A male preponderance was detected in some studies, but the gender predominance is not established [7,15]. Males were twice more commonly affected in this study.

It has been reported that, CPAM affects both sides of lung equally [7]. But, Muller CO et al., reported more frequent involvement of left side [3]. However, the authors found a right sided preponderance (2:1). The predominant involvement of lower lobes in CPAM is well observed in this study, as upper lobes were not involved in any case. Similarly, the unilobar nature of this disorder as marked in other series is also found here, as any bilobar or multilobar involvement was not encountered [7]. Multilobar involvement is reported in 1-2% cases of CPAM [5]. Surgical excision is the treatment of choice for symptomatic patients. It is also widely advocated for asymptomatic patients because of risk of infection, pneumothorax and malignancy [3,5]. If not operated, most of the asymptomatic children will also become symptomatic (80%) [3]. A clear association between CPAM and malignant tumours (8.6% cases) have been reported in a review of paediatric pulmonary neoplasms [16]. Lobectomy should be preferred over parenchyma saving resections because of difficulty in accurate delineation of margins of lesion within a lobe by preoperative CT scan, adequate compensatory lung growth in infants and complications such as air leak or bleeding, that are

Author's name and year	Place of study	Study period	Number of subjects	Age of children considered	Parameters compared	Conclusion
Nazem M and Hosseinpour [11]	Iran	1996-2008 (12 years)	30	3.5 m to 9.5 m	a. Mean age at presentation-6.4 m b. Most common lobe involved-LUL (50%) c. Mortality-13.5%	In contrast to other studies, the present study revealed: a. an early age at presentation b. RUL was most common involved lobe c. gradual improvement in detection, surgical management and prognosis of these infants
Cataneo DC et al., [8]	Brazil	1979-2009 (30 years)	20	9 d to 4 yr	a. Mean age-6.9 m b. Most common lobe involved- LUL (35%), c. Mortality-nil	
Abdel-Bary M et al., [14]	Egypt	2015-2019 (5 years)	31	2 m to 6 m	a. Mean age at presentation-111 d b. Most common lobe involved-LUL (62%) c. Mortality-nil	
Present study	India	2018-2021 (3 years)	8	1 m to 4 m	a. Mean age at presentation-2 m b. Most common lobe involved- RUL(50%) c. Mortality-nil	

[Table/Fig-8]: Comparison of findings in Congenital Lobar Emphysema (CLE) patients with similar studies [8,11,14].
m: Months; d: Days; yr: Year; LUL: Left upper lobe; RUL: Right upper lobe

associated with segmentectomy [3,5]. Lobectomy was performed in all cases in this study.

Bronchogenic cysts are developmental malformation of foregut or tracheobronchial tree due to abnormal budding [12,17]. Accordingly, the location of cysts is quite variable. It is mentioned that two thirds are within lung parenchyma and rest in the mediastinum [7]. Lung parenchyma was the site of lesion in both of cases in the present study. Bronchogenic cysts may be asymptomatic and detected incidentally on chest radiography, performed for other purpose. Symptomatic patients usually present with cough, fever and dyspnoea [17]. Similarly, both the patients in this study presented with features of pneumonia. Although, it is mentioned that, cysts have no specific side predilection, both the cases were found on right side of chest [7]. Similar to other reported studies bronchogenic cysts were located in lower lobes in this study, as upper lobes were not affected [18,19]. However, Sarper A et al., reported upper lobe involvement in four of their cases, out of six cases in their study [17]. Patent connection to tracheobronchial tree is unusual and when present leads to superadded infection [7,17]. None of the present cases had communication to airways and complete resection was performed. Bronchogenic cyst should be resected due to risk of infection, rupture, haemorrhage and malignancy [7,12]. The lifetime risk of malignant transformation is reported as 0.7% [20].

Vascular malformations are rarely found in mediastinum, but when encountered, they are usually lymphatic malformations [21]. Lymphangiomas constitute 5-6% of thoracic masses [22]. They were detected in 10.5% of cases in the present study. Although, it is reported that most of the cases present before two years of age, in this study patients presented at three and four years age [22,23]. Lymphangiomas confined solely to mediastinum are very rare (1% cases) and vast majority are extensions of cervical lesions [22-24]. One of the patients had associated cervical extension and another was having axillary extension. Mediastinal lymphangiomas need surgical removal, but total excision is difficult in many cases and may not be possible in some children [21-23]. Incomplete excision patients should be treated with adjuvant sclerotherapy [21,24]. Although, postoperative recurrence is reported as a major complication, authors did not encountered any recurrence [24].

Among paediatric pulmonary masses primary tumours, metastatic tumours and non neoplastic lesions occur with a ratio of 1:5:60 [2,25]. So, primary pulmonary malignancies are rare in children but when encountered, carcinoid tumours and Pleuropulmonary Blastoma (PPB) are the common pathologies [2,26]. PPB occurs almost exclusively during childhood and are monophasic tumours, containing only mesenchymal malignant components [2,27]. The newborn with pulmonary malignancy detected in the present study was found to be having biphasic histology, containing mesenchymal as well as epithelial components. She was diagnosed as Classical Biphasic Pulmonary Blastoma (CBPB), which is different from PPB. The diagnosis was based on microscopic findings and IHC on the resected specimen. Pulmonary blastoma is distinguished from another more common paediatric tumour of lung, PPB, which shows mature and immature cartilaginous areas. The glandular structures are occasionally detected with ciliated columnar epithelium overlying condensed small round cells [28,29]. CBPB is usually seen among adult smokers with peak incidence at fourth decade of life [27,30]. Finding such an aggressive, adult onset malignancy in a five-day-old newborn is extremely rare and a remarkable feature in the present study.

Limitation(s)

The major limitation of this study is limited number of patients, because of rarity of these malformations and impact of COVID-19 crisis. Again, this is a unicentric study having inherent problems. A multicentric study will certainly provide further insight and new array of management approach.

CONCLUSION(S)

Thoracic mass lesions in children are mostly developmental malformations and usually present before the age of six month. A high index of suspicion is required for understanding these rare disorders, especially in infants presenting with progressive respiratory distress. In contrast to existing literature, where CPAM is described as most common pathology, CLE was the most common finding in the present study. Right side of thorax was more commonly affected than left side. Primary pulmonary malignancies such as pulmonary blastoma may be encountered rarely, even during neonatal period. Early identification and complete surgical resection are mainstays of treatment. Although, radiological studies suggest the lesion, histopathology is the gold standard for conclusive diagnosis.

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