Original Article

Access this article online
Website: www.afrjpaedsurg.org
DOI: 10.4103/0189-6725.150987
Quick Response Code:

Congenital cystic lesions of lung in the paediatric population: A 5-year single institutional study with review of literature

Shibsankar Barman, Kartik Chandra Mandal, Rajarshi Kumar, Sumitra Kumar Biswas, Madhumita Mukhopadhyay[1], Biswanath Mukhopadhyay

ABSTRACT

Background: The aim was to evaluate the clinical presentation, investigation modalities, operative management, pathology, outcome (morbidity and mortality) and short term follow-up of congenital cystic lesions of the lung. **Materials and Methods:** This is a retrospective study. Study period was 5 years (December 2008-November 2013) in the Department of paediatric surgery, Nil Ratan Sircar Medical College. Study population: Total number = 10 patients. Age range: 2 days-7 years. (Neonate-4). Male and female ratio = 1:1. **Result:** Among 10 cases of lung cyst four having congenital lobar emphysema, four having congenital pulmonary airway malformation, one sequestration and one teratoma. All patients have undergone surgical excision in terms of lobectomy or excision of the lesion. Post-operative histopathology confirmed the diagnosis. Recovery was uneventful. **Conclusion:** Although our experience is limited, operative management of lung cysts seems to be safe with rewarding results. However we are yet to encounter many of the other varieties of the cysts found in the lung, which may be associated with other congenital anomalies and have an impact on prognosis.

Key words: Congenital lung cyst, congenital lobar emphysema, congenital pulmonary airway malformation, lobectomy

INTRODUCTION

Congenital cystic lesions of lung present an interesting spectrum of pathology in the paediatric population. Congenital lobar emphysema (CLE), congenital pulmonary airway malformation (CPAM), Sequestration, intrapulmonary cystic teratomas are rare conditions found in children. In 1954, Gross and Lewis, published the first case report of CLE.[1] CLE and CPAM underwent lobectomy, intralober cystic teratoma and extralobar sequestration undergone excision of the lesion. The aetiology of CLE was unknown, in over half of the reported cases. Intrinsic obstruction may result from cartilaginous deficiency, bronchial stenosis or redundant bronchial mucosa. Abnormal vessels or lymph nodes are usually responsible for extrinsic compression.[1]

MATERIALS AND METHODS

This was a five years (December 2008-November 2013) retrospective study in the department of Paediatric Surgery, N R S Medical College.

Patients with progressive respiratory distress and repeated respiratory tract infection confirmed both clinically and radiologically were included in our study.

RESULTS

There were patients, with age range of 2 days to 7 years. The Male and female ratio = 1:1. Among the 10 cases of lung cyst four cases are CLE and two cases of CLE involving left upper lobe and two cases of CLE involving left lower lobe [Table 1]. Left hemithorax was bulged; breathe sound diminished, hyper resonance, mediastinal shifting to right in all cases. Diagnostic difficulty was pneumothorax, congenital diaphragmatic hernia. Diagnosis was confirmed by chest-X-ray PA and lateral view, contrast enhanced computed tomography (CT) scan thorax done in two cases. Blood gas analysis was not done in any case. Lobectomy was performed in all cases of CLE [Figure 1]. Chest X-ray was done on the third post of day to confirmed lung expansion and chest tube was removed on the third post of the day. Patients were discharged on fifth or sixth post-operative

Department of Paediatric Surgery, Nil Ratan Sircar Medical College and Hospital, [1]Department of Pathology, Institute of Post Graduate Medical Education and Research, Kolkata, West Bengal, India

Address for correspondence:
Dr. Kartik Chandra Mandal,
Department of Paediatric Surgery, Nil Ratan Sircar Medical College and Hospital, 138 AJC Bose Road, Kolkata - 700 014, West Bengal, India.
E-mail: kartikchandramandal@yahoo.in

Table 1: Detail parameters of our series (n = 10)

Age at presentation	Sex	Clinical features	Investigations	Diagnosis	Management and recovery
1 year	Female	Repeated attacks of respiratory infection and breathlessness	Chest X-ray: Large cystic lesions in right lower lobe, mediastinal shift CT scan: Few air containing cysts noted in the right lung, mediastinal shift to left with atelectasis of right middle lobe. Upper part of left lower lobe shows patchy consolidation	Right CPAM	Operation: Right thoracotomy+right lower lobe lobectomy Recovery: Uneventful
4 days	Female	Breathlessness and diminished air entry left hemi thorax	Chest X-ray: Large cystic lesions in left lower lobe CT scan: Not done	Left CLE	Operation: Left thoracotomy+left lower lobe lobectomy Recovery: Uneventful
2 months	Male	Respiratory distress, repeated RTI and diminished air entry left hemi thorax	Chest X-ray: Large cystic lesions in left upper lobe CT scan: Not done	Left CLE	Operation: Left thoracotomy+left upper lobe lobectomy Recovery: Uneventful
3 days	Male	Dyspnoea since birth and poor feeding diminished breath sound in right lower and mid zone. No peristaltic sound in chest. Abdomen scaphoid	Chest X-ray: Air filled spaces looking like bowel loops in right hemi thorax, visible bowel loops in abdomen CT scan: Not done	Right CPAM	Operation: Right lower lobectomy Recovery: Initially a diagnosis of CDH was made and abdomen opened up to find no pathology, incision extended as right thoracotomy
7 years	Male	Repeated attacks of pulmonary infection and occasional attacks of respiratory distress. Dull on percussion on left side of the chest and diminished air entry on the left side	Chest X-ray: Soft tissue shadow left lung CT scan: Heterogeneous left lung lesion	Immature teratoma of left lung	Operation: Left thoracotomy+excision of the lesion Recovery: Uneventful
5 years	Male	Recurrent episode of RTI since birth and diminished air entry on the left side	Chest X-ray: Ovoid opacity in left lower lung CT scan: Bronchgenic cyst left lower lobe	Pulmonary sequestrati-on of left lung	Operation: Left thoracotomy and lobectomy Recovery: Uneventful
2 years	Male	Repeated attacks of pulmonary infection and occasional attacks of respiratory distress, diminished air entry on the left side	Chest X-ray: Radiolucency in upper left lung with mediastinal shifting CT scan: Confirm the diagnosis	Left CLE	Operation: Left thoracotomy+left upper lobectomy Recovery: Uneventful
2 days	Female	Respiratory distress since birth and diminished air entry on the left side	Chest X-ray: Inconclusive CT scan: Not done	Left CPAM	Operation: Left thoracotomy+left lower lobectomy Recovery: Uneventful
11 days	Female	Respiratory distress since birth, difficulty in feeding and diminished air entry and resonant percussion on the left side	Chest X-ray: Soft tissue shadow left lung with mediastinal shifting CT scan: Collapsed and consolidation of right lung and left lower lobe, mediastinal shift to right, small pneumothorax	Left CLE	Operation: Left thoracotomy+left lower lobectomy Recovery: Uneventful
2 years	Female	Repeated episodes of RTI, occasional respiratory distress since birth and diminished air entry on the right side	Chest X-ray: Radiolucent right hemithorax CT scan: Huge multiloculated cystic areas replacing almost whole right lung	Right CPAM	Operation: Right thoracotomy+right lower lobectomy Recovery: Uneventful

CT: Computed tomography; CPAM: Congenital pulmonary airway malformation; CDH: Congenital diaphragmatic hernia; CLE: Congenital lobar emphysema; RTI: Respiratory tract infection

day. Pre-, per- and post-operative clinical examinations and pulse oximetry monitoring was done in every case.

One patient with lung cyst involving left upper lobe, presented with repeated chest infection, occasional respiratory distress without mediastinal shift. Breath sounds were normal. Patient was treated by physician initially. Chest X-ray and CT scan thorax shows left upper lobe heterogeneous lesion. On exploration through left posterior thoracotomy in fourth intercostals

space, the cyst was in left upper lobe with well-defined margin, encapsulated and cyst was removed without capsular breach. Histopathological report showed immature teratoma of left upper lobe.

Another patient presented with repeated attack of the respiratory tract infection. Breath sound was diminished, dull on percussion on left lower chest without mediastinal shift. Diagnostic confusion was with left sided congenital diaphragmatic hernia. CT scan thorax confirmed as a case of lung cyst of the left lower lobe. Left thoracotomy was done through sixth intercostals space, left antero-lateral. Extralobar mass with solid and cystic component with separate vascular supply and compressed left lower lobe was found. Histopathological report came as extralobar pulmonary sequestration.

Four patients with CPAM were operated, of which three cases involving right lower lobe and one case involving left lower lobe. Patient in neonatal age group presented with respiratory distress, in post neonatal age presented with repeated respiratory tract infection. Chest X-ray done in all cases, CT scan done in two post neonatal patient showing multiloculated cystic space occupying lesion compressing surrounding lung tissue. Right lower lobectomy done in three, left lower lobectomy in one case [Figure 2]. Histological report confirmed as CPAM in all cases [Figure 3]. Among the four cases of CPAM, we found three Type I and one Type II. Pre-operative bronchoscopy was done in one case (2 years female patient with right CPAM) to exclude intraluminal bronchial obstruction. Operative procedures were uneventful in all the cases and they were doing well in the post-operative period.

We have encountered air leak during left lower lobectomy (2 days female patient with left CPAM) from left lower lobe bronchus. It was detected and repaired with interrupted prolene stitches. Post-operative period was uneventful.

Follow-up period varied from 6 months to 1 year. In 1 year follow-up only one patient developed respiratory tract infection, which was managed conservatively.

DISCUSSION

Congenital cystic lesions of lung in children are a spectrum of anomaly. Early diagnosis, investigations, and management of these potentially life-threatening anomalies rewarded with good result. These include CLE, CPAM, pulmonary sequestration, intrapulmonary cyst etc.

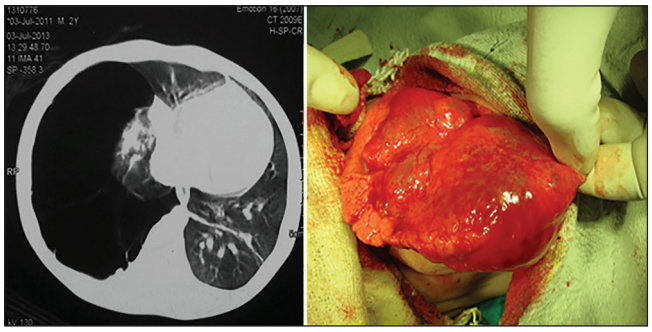

Figure 1: Coronal section of computed tomography showing right sided congenital lobar emphysema with intra-operative excision

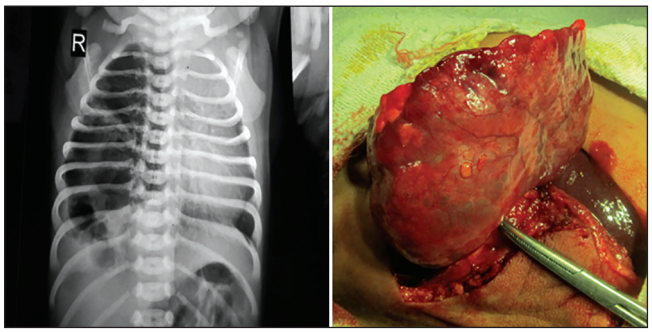

Figure 2: X-ray chest PA view showing right congenital cystadenomatoid malformation with intra-operative excision

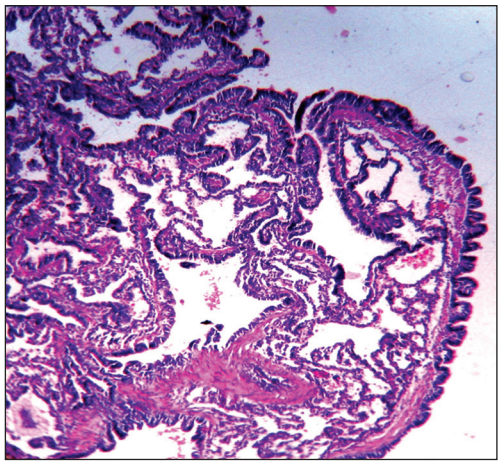

Figure 3: Histopathological section showing congenital cystadenomatoid malformation

Hence, there is a high possibility that the diagnosis of congenital lung malformations would be missed, unless specifically kept in mind and sought for by advanced imaging studies as pneumonia is the most common cause of respiratory morbidity in children, especially in developing countries.[2]

Congenital lobar emphysema was first reported in 1954 by Gross and Lewis.[1] It usually involves left upper, right middle and right upper lobe. Lower lobe involvement is rare. In our series, we found all patients having

involvement of the lower lobe. Regarding aetiology there are number of theories, but most accepted and recent theory was obstruction of bronchioles either extrinsic or intrinsic causing air trapping. Usually, they are presented with respiratory distress, but may be asymptomatic. Confirmation of diagnosis was done by contrast enhanced CT thorax. Neonates with respiratory distress diagnosed with X-ray chest. Symptomatic group are treated with lobectomy and asymptomatic patients can be managed conservatively. Operative mortality rate is 3-7%, whereas with conservative therapies it is 50-75%. Hence, conservative management should be reserved only for patients with milder symptoms or no distress at all.[3]

Congenital pulmonary airway malformation accounts for 25% cases of lung cyst. Usually lower lobe involvement is common. Congenital cystadenomatoid malformation (CCAM) was first acknowledged in the medical literature by Ch'in and Tang in 1949.[4] CCAM is recently renamed as CPAM.[5] A classification system was proposed by Stocker *et al*. who classified into three types based on histopathology.[6] CPAM classification scheme (clinico-pathological) has been revised in 2002 by Stocker and categorized them as - Type 0: Trachea-bronchial, Type 1: Bronchial-broncheolar, Type 2: Broncheolar, Type 3: Broncheolar-alveolar duct, Type 4: Distal acinar.[7] CPAM is caused by an embryonic insult which leads to abnormal development of terminal bronchioles. Usually, an entire lobe of the lung is replaced by a nonworking cystic piece of abnormal lung tissue in most cases. In our series, we found two patients having involvement of right lower lobe, one having right middle and one left lower lobe diagnosed at neonatal and post neonatal age. Outcome of the foetus with CPAM is good, in rare cases cystic mass compress around normal lung and as well as heart and can be life threatening for foetus. Antenatal diagnosis has done by prenatal ultrasonography (USG) revealed echogenic mass, displacement of heart, pushed diaphragm, absence of visible lung tissue. Antenatally diagnosed CPAM should be monitored closely and successful excision after delivery. Large macrocystic lesion requires in-utero Harrison thoraco-amniotic shunt and very large mass with respiratory compression may require EXIT procedure. Extreme cases with compression of heart require foetal intervention. Non-immune hydrops fetalis survival may increase with prenatal steroid.[8,9] All patients are managed with lobectomy in our series with good post-operative outcomes.

Extralobar sequestration accounts 25% cases of bronchopulmonary sequestration. Males are more affected than female. Left lower lobe involvement was more than right lower lobe. Diagnosis should be done by CT scan with three-dimensional reconstruction. Most of the cases CT scan with clinical and chest X-ray correlation confirm the diagnosis.[10,11]

Pulmonary teratoma is a rare germ cell tumour. First case of intrapulmonary teratoma reported by Morh in 1839.[12] Patient usually presented with repeated chest infection, chest pain, cough, and haemoptysis.

Pulmonary teratoma usually occurs in left upper lobe with mature cystic teratoma being more common. Immature teratoma is a rare germ cell tumour particularly in children. In our series we have reported a case of intrapulmonary immature cystic teratoma of left upper lobe. Surgical resection is the treatment of choice and complete removal leads to a long recurrence free survival.[13]

On an average 15-20% cases of CPAM are associated with other anomalies commonly cardiac and renal.[14] Type 2 CPAM (Bronchiolar) commonly associated with other anomalies (cardiac, renal, intestinal atresia, skeletal anomalies etc.) approximately in 60% cases. Truncus arteriosus and tetralogy of Fallot are most common cardiac anomalies associated with CPAM.[15] Extra lobar sequestration is more commonly (50%) associated with CPAM and other congenital anomalies.

In our study, we have performed echocardiography and USG of the abdomen in all patients. We have not found any associated anomalies.

Surgical resection is the treatment of choice in cystic lung lesions in the paediatric population. In cases of cystic pulmonary lesions with uncertain radiological findings, surgery is indicated in order to perform histological examination of the lesion and to prevent infection and the potential risk for neoplastic transformation.

REFERENCES

1. Irving IM. Malformations and acquired lesions of lungs, pleura and mediastinum. In: Lister J, Irving IM, editors. Neonatal Surgery. London: Butterworth & Co. Ltd.; 1990. p. 265-71.
2. Reddaiah VP, Kapoor SK. Epidemiology of pneumonia in rural underfives. Indian J Pediatr 1990;57:701-4.
3. Holliday MA, Segar WE. The maintenance need for water in parenteral fluid therapy. Pediatrics 1957;19:823-32.
4. Ch'in KY, Tang MY. Congenital adenomatoid malformation of one lobe of a lung with general anasarca. Arch Pathol (Chic) 1949;48:221-9.
5. Stocker JT, Mani H, Husain AN. The respiratory tract. In: Stocker JT, Dehner LP, editors. Pediatric Pathology. 3rd ed. Philadelphia: Lippincott/Williams & Wilkins; 2011. p. 441-515.

6. Stocker JT, Madewell JE, Drake RM. Congenital cystic adenomatoid malformation of the lung. Classification and morphologic spectrum. Hum Pathol 1977;8:155-71.
7. Stocker JT, Husain AN. Cystic lesions of the lung in children: Classification and controversies. In: Timens W, Popper H, editors. Pathology of the Lung. Vol. 39. Medknow: European Respiratory Monograph; 2007. p. 1-20.
8. Curran PF, Jelin EB, Rand L, Hirose S, Feldstein VA, Goldstein RB, *et al*. Prenatal steroids for microcystic congenital cystic adenomatoid malformations. J Pediatr Surg 2010;45:145-50.
9. Peranteau WH, Wilson RD, Liechty KW, Johnson MP, Bebbington MW, Hedrick HL, *et al*. Effect of maternal betamethasone administration on prenatal congenital cystic adenomatoid malformation growth and fetal survival. Fetal Diagn Ther 2007;22:365-71.
10. Wu N, Sun Y, Zheng QF, Lü C, Yan S, Zhang LJ, *et al*. Diagnosis and treatment of intralobar pulmonary sequestration. Zhonghua Yi Xue Za Zhi 2007;87:2627-31.
11. Ooi GC, Cheung CW, Lam WK, Tsang KW. Pulmonary sequestration: Diagnosis by magnetic resonance angiography and computed tomography. Chin Med J (Engl) 1999;112:668-70.
12. Collier FC, Dowling EA, Plott D, Schneider H. Teratoma of the lung. AMA Arch Pathol 1959;68:138-42.
13. Takeda S, Miyoshi S, Ohta M, Minami M, Masaoka A, Matsuda H. Primary germ cell tumors in the mediastinum: A 50-year experience at a single Japanese institution. Cancer 2003;97:367-76.
14. Laje P, Liechty KW. Postnatal management and outcome of prenatally diagnosed lung lesions. Prenat Diagn 2008;28:612-8.
15. Priest JR, Williams GM, Hill DA, Dehner LP, Jaffé A. Pulmonary cysts in early childhood and the risk of malignancy. Pediatr Pulmonol 2009;44:14-30.

Cite this article as: Barman S, Mandal KC, Kumar R, Biswas SK, Mukhopadhyay M, Mukhopadhyay B. Congenital cystic lesions of lung in the paediatric population: A 5-year single institutional study with review of literature. Afr J Paediatr Surg 2015;12:66-70.

Source of Support: Nil. **Conflict of Interest:** None declared.