CASE SERIES

Variability of presentation and surgical approach in Congenital Cystic Lesions of Lung: A retrospective study in children presenting in Mayo Hospital

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Abstract

The objective was to study the clinical presentation and surgical outcome in children with congenital cystic lesions of the lung. The medical records of 11 patients operated in the department of paediatric surgery, from January 2014 to December 2017, were evaluated retrospectively. Median age was 18 months (1-108). Respiratory distress was seen in 5 (45.4%) patients, recurrent chest infections in 4 (36.4%) patients and only 2 (18.2%) presented after birth. One patient of congenital lobar emphysema was misdiagnosed as pneumothorax and four patients of recurrent chest infection had been misdiagnosed as pulmonary tuberculosis. All patients underwent lateral thoracotomy. There was no mortality, median length of hospital stay was 4 days (4-5) and only one patient needed postoperative ventilation. On follow up, 10 (90.9%) patients had attained normal level of physical activities. We conclude that increasing awareness of these lesions can prevent misdiagnosis and unnecessary tube thoracostomy and anti tubercular therapy in children.

Keywords: Congenital cystic lesions of lung, Congenital cystic adenomatoid malformation, Congenital lobar emphysema, Bronchopulmonary sequestration, Bronchogenic cyst.

Introduction

Congenital cystic lesions of lung occur due to defective bronchoalveolar development. These are also known as bronchopulmonary foregut malformations, comprising congenital cystic adenomatoid malformation (CCAM), congenital pulmonary airway malformation (CPAM), congenital lobar emphysema (CLE), bronchopulmonary sequestrations (BPS) and bronchogenic cysts. The incidence of congenital cystic lesions of lung is in the range of 1 per 8,300 to 35,000 live births.[1,2]

Clinical presentations are variable in respect of severity and time of presentation. It may present antenatally as polyhydramnios or hydrops foetalis and foetal death. Postnatally, it may remain asymptomatic or may present with respiratory distress or recurrent chest infections.[3,4] Very rarely, it may undergo malignant transformation and cause pleuropulmonary blastoma.[3-5]

Treatment modalities for cystic lesions of lung include expectant treatment for asymptomatic patients and operative treatment for symptomatic patients carried out either by thoracoscopy or thoracotomy or embolisation of the feeding artery of the BPS.[1,6]

These lesions are often confused as pulmonary tuberculosis in children presenting with recurrent chest infections and pneumothorax in neonates presenting with respiratory distress.[7,8] In order to avoid misdiagnosis, all children with recurrent chest infections or incidentally found cysts in chest X ray (CXR) should undergo computed tomography (CT Scan) of chest.[4] It is needless to emphasise that by sharing our experience with regards to a timely diagnosis and early intervention, we not only can update and enhance the existing knowledge, but also can spread awareness amongst physicians and paediatricians.

Case Series

After obtaining ethical approval and informed consent from parents, medical records of patients operated for congenital cystic lesions of lung in the department of paediatric surgery, Mayo Hospital, Lahore, were reviewed from January 2014 to December 2017. Data included information about antenatal record, age (in months) and weight (in kilograms) of the patient at time of presentation, gender of the patient, findings of CXR and CT Scan chest for lesion site, mediastinal shift, collapse of healthy lung tissue and/or herniation of affected area, aberrant vessel if any, and clinical presentation.

History of respiratory distress was taken as dyspnoea, wheezing and cyanosis. Recurrent chest infection was labeled as fever, cough, tachypnoea with antibiotics usage twice or more in one year. Haemoptysis or blood

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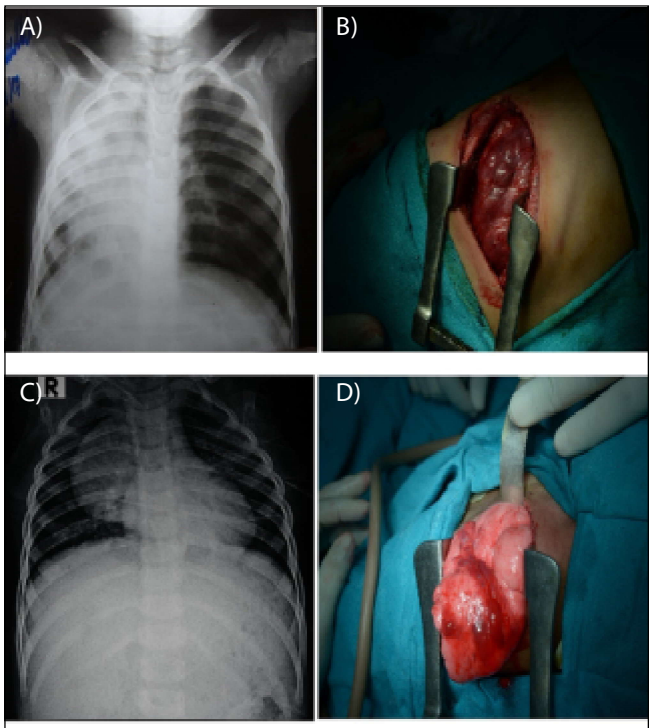

Figure: Selected images of some patients: A) CXR showing right upper lobe collapse with mediastinal shift and left lung hyperinflation. B) Thoracotomy of same patient showing right upper lobe CPAM. C) CXR showing right paratracheal cyst. D) Thoracotomy of CLE patient showing popping out of emphysematous lung.

in sputum and considered massive if it exceeded 8 ml/day. Findings of bronchoscopy were recorded if done. Operation was muscle sparing lateral thoracotomy on the affected side. Procedures for example cyst excision, segmentectomy, lobectomy and pneumonectomy were carried out and findings were recorded. Post-operative data included post-operative complications such as wound infection, respiratory tract infection, need of mechanical ventilation and tube thoracostomy duration. Length of hospital stay and histopathology of resected specimen was also recorded. Patients' parents were called in for follow up and after informed consent, level of physical activities according to the age, whether normal or restricted was also recorded.

Data was entered in SPSS version 23. Median and inter quartile range (IQR) and standard error of mean (SEM) of categorical variables - age, weight, length of hospital stay, chest tube and mechanical ventilation duration were calculated. Frequency and percentage of descriptive variables were also calculated.

In last 3 years, a total 11 patients were operated for cystic lung lesions. Only 2 (18.2%) were diagnosed antenatally. Seven (63.6%) were male and four (36.4%) were female. Median age was 18 months with IQR 1.0-108.0 months and SEM 17.91 (minimum age 8 days; maximum age 12 years). Median weight was 7 kg with IQR 3.5-25.0 kg and SEM 3.45. Regarding clinical presentation, 5 (45.4%) patients presented with respiratory distress while 4 (36.4%) presented with recurrent chest infection and 2 (18.2%) presented after birth. The 4 patients labeled as having recurrent chest infection, had been given anti tubercular therapy (ATT). Patient with BPS had haemoptysis and a bronchoscopy was done only on this patient, which revealed no abnormal findings. CXR and CT Scan chest revealed herniation of affected lobe, with mediastinal shift in 2 patients of CLE on the left side and right para tracheal cyst in case of bronchogenic cyst (Figure 1-C). Lesion site of CCAM was left upper lobe in 2 patients, left lower lobe in 2 patients, right upper lobe in 1 patient, right middle and lower lobes in 1 patient and right lower lobe in 1 patient. CXR of one patient with CCAM showed collapse of right upper lobe of lung with mediastinal shift and left lung hyperinflation (Figure 1-A). Patient of BPS had a multicystic mass in left lower chest with systemic supply by an aberrant vessel originating from coeliac trunk. Seven patients (63.6%) were diagnosed as having CCAM, 2 (18.2%) had CLE, 1 (9.1%) had bronchogenic cyst and 1 (9.1%) had BPS. Demographic data according to different types is given in Table 1.

All patients underwent lateral muscle sparing thoracotomy. Seven (63.6%) patients had left thoracotomy, with 3 (27.3%) patients undergoing left upper lobectomy, 3 (27.3%) left lower lobectomy and 1 (9.1%) having segmentectomy. Four (36.4%) patients

Table-1: Demographic data of patients with congenital cystic lung lesions including pre-operative status.

	CCAM	CLE	BPS	Bronchogenic Cyst
Sex				
Male	5	1	0	1
Female	2	1	1	0
Age (months)	(1.25 - 120)*	0.25, 2	108	18
Weight (kg)	(3.75 - 29)*	3.4	25	8
Antenatal diagnosis	Yes (1) No (6)	Yes (1) No (1)	No	No
Pulmonary Tuberculosis Misdiagnosis	Yes (3) No (4)	No	Yes	No
Mechanical Ventilation	No	Yes (1)	No	No

*Data presented as inter quartile range.
CCAM: Congenital Cystic Adenomateoid Malformation
CLE: Congenital Lobar Emphysema, BPS: Bronchopulmonary Sequestration

Table-2: Congenital Cystic Lung Lesions Clinical presentation, Differential Diagnosis and treatment.

	Clinical presentation	Differential Diagnosis	Treatment
CCAM	Limited to 1 lobe (n=6), limited to 2 lobes (n=1). Antenatal diagnosis (n=1), All symptomatic. Symptoms of recurrent chest infection (n=3), respiratory distress (n=3)	Tuberculosis, Abscess Bronchiectasis	Surgical resection (n=7, left upper lobectomy (n=2) Left lower lobectomy (n=2) Right upper lobectomy (n=1) Right middle and lower lobectomy (n=1) Right lower lobectomy (n=1)
CLE	Limited to 1 lobe (n=2), Antenatal diagnosis (n=1). All symptomatic. Respiratory distress (n=1)	Pneumothorax	Surgical resection (n=2). Left upper lobectomy (n=10). Left lower lobectomy (n=1)
BPS	Presented with recurrent chest infection (n=1)	Abscess, Pneumonia Tuberculosis	Surgical resection (n=1). Left Lower Secgmentectomy (n=1)
Bronchogenic Cyst	Presented with right paratracheal cyst (n=1). Respiratory distress (n=1)	Infected cyst	Surgical excision of cyst (=1)

had right thoracotomy, with 1 (9.1%) undergoing cystectomy, 1 (9.1%) right lower lobectomy, 1 (9.1%) right upper lobectomy and 1 (9.1%) right middle and lower lobectomy. Per operatively, multicystic lesions were noted in the affected lung lobe (Figure 1-B) or the affected lung popped out of thoracotomy incision in case of CLE (Figure 1-D). The details of surgical approach, differential diagnosis and presentation according to the different types of cases is summarised in Table 2.

Postoperatively, only 1 (9.1%) patient needed mechanical ventilation and he was successfully weaned off within 24 hours. Median chest tube drainage was 2 days with IQR 2-3 days and SEM 0.3. Median length of hospital stay was 4 days with IQR 4-5 days and SEM 0.42. Histopathology report showed cysts lined by columnar epithelium in cases of CCAM, alveolar distention in CLE cases and in case of bronchogenic cyst, lining of respiratory epithelium. Almost all patients (90.9%) had attained normal level of physical activities according to their age, except 1 (9.1%) undergoing two lobectomies

Discussion

In developed countries, almost all cases of congenital cystic lesions of lung are diagnosed antenatally, but in our set up due to lack of proper antenatal checkups, only 2 cases were detected before birth. These findings are similar to studies done by Karunasumetta et al and Raman et al.[7,9]

In our series, 63.6% were male. Minimum age was 8 days and maximum age was 12 years. Other studies have also shown male preponderance and variable age at the time of diagnosis.[1,3,9]

Respiratory distress was noted in 45.4% of the cases. One case of CLE was misdiagnosed as pneumothorax and tube thoracostomy was done. Not recognising CLE and misinterpreting it as pnemothorax on CXR, leads to unnecessary tube thoracostomies in these patients and in Prabhu et al's study, around 30% of the patients underwent intercostal drainage due to misdiagnosis.[8] Misdiagnosis can be avoided by proper interpretation of CXR.

In our series, next common presentation was recurrent chest infections in 36.4% of the patients. All were misdiagnosed as having pulmonary tuberculosis and were given ATT. After failure of improvement in symptoms, patients were investigated and appropriate referrals were made. A CT Scan of the chest helps in differentiating these lesions from tuberculosis and other acquired lung anomalies as these lesions are commonly discrete with multiple air filled cysts in case of CPAM; has anomalous blood supply in case of BPS and in the case of bronchogenic cyst, the cyst has higher than water attenuation.[4] These findings are similar to the study conducted by Raman et al, emphasising the need to evaluate children with recurrent chest infections for congenital cystic lesions of lung, even in tuberculosis prevalent areas.[7]

Asymptomatic patients are managed conservatively, but in our series all patients were symptomatic and underwent thoracotomy with no mortality. All attained normal level of activities according to their age, except one child who had undergone two lobectomies. Our findings are consistent with other studies showing that lung tissue regenerates in children.[1,9] Rothenberg et al recommended segmentectomy when bilateral or extensive disease was noted in order to preserve lung functions, or when the disease was limited to an anatomic segment.[10] We did segmentectomy in only one case. In rest of the cases it involved lobe or more than one anatomic segment. In our series, the most common lesion was CPAM, involving all

lung lobes. The second most common lesion was CLE and it was on the left side. Literature review also suggests the same.[1,2]

Although thoracoscopy causes lesser pain and entails shorter hospital stay, yet thoracotomy and thoracoscopy are both equally feasible surgeries for such lesions. In our series, median hospital stay was 4 days, which is same after thoracoscopy.[6,10]

The wide range of ages, and the few number of cases in our series, made it difficult to generalise the results.

Conclusion

Focusing on antenatal diagnosis and increasing awareness of congenital cystic lesions of lung, can prevent misdiagnosis and unnecessary tube thoracostomy and ATT in children.

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