Clinical Brief

Congenital Malformations of the Lung

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ABSTRACT

Congenital malformations of the lung are rare and vary widely in their presentation and severity. The most common manifestation of the congenital cystic disease of the lung at newborn and early infancy is respiratory distress. Later on in life, cysts usually lose this compressive character and may remain asymptomatic until infection occurs, while producing cough, dyspnea, and thoracic pain. The purpose of this study is to review authors institutional experience of congenital cystic lung disease, with specific reference to diagnosis, treatment, as well as outcome, furthermore, to present some cases with unusual clinical manifestations. **[Indian J Pediatr 2007; 74 (2) : 192-194]** *E-mail: jafariroohi@yahoo.com*

Key words : *Lobar emphysema; Pulmonary sequestration; Bronchogenic cyst; CCAM*

Congenital malformations of the lung and mediastinum vary considerably in their embryology, presentation, management, and out come.[1] They consist of pulmonary sequestration (PS), cystic adenomatoid malformation (CCAM), congenital lobar emphysema (CLE), and bronchogenic cyst (BC). These four anomalies present a different clinical picture, are often difficult to diagnose, and require surgical management.[2]

In two referral centers of Tabriz University of Medical Sciences, The authors evaluated the medical records of surgery realm patients across the March 1996 and March 2005 records with the discharge diagnosis of lung congenital malformation was documented with exclusive attention upon the manifestation which related to the age, method of diagnosis and outcome.

CASE REPORTS

Congenital lobar emphysema (n=7)

Through these patients, 4 were referred by main feature of respiratory distress from birth. Three remainder patients were misdiagnosed, 2 of them were refered as a refractory pneumonia and the other one misdiagnosed as a pneumothorax at a peripheral hospital and an intercostals drain was inserted increasing the respiratory distress, requiring an emergency lobectomy. There was patent ductus arteriosus in 1 case of CLE. All the patients underwent surgery procedures.

About the location of the lesion; 2 in right upper lobe, 1 in right lower lobe and in 4 subjects the lesion was located in left upper lobe. The recovery was uneventful in all seven patients.

Bronchogenic cyst (n=6)

The most common presentation comprised cough, suppurative bronchitis and hemoptysis with one case of massive hemoptysis. In all cases, the delay between appearance of symptoms, discovery of the cyst, and operation varied from a few hours to 6 years (mean,2 years and 2 months). Plain X-ray along side chest CT scans were chosen as diagnostic modalities for all of patients, which revealed involvement of right upper lobe in two cases, right middle lobe in 1 case, right lower lobe in 1 and left lower lobe in 2 cases. Partial absence of pericardium was seen in one of our cases in this group as a very unusual associated anomalies. All the subjects were asymptomatic and long-term follow-up of pulmonary function is good despite minor radiological anomalies such as compensatory lobar hyperinflation.

Congenital cystic adenomatoid malformation (n=3)

The presenting symptom was respiratory difficulty or distress. Resembling the prior groups, chest radiography and computerized tomography was the choice of evaluation. In two patients left - upper lobe were affected and one subject had bilobar involvement of upper and middle lobes of left side. Patients had no complications.

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Pulmonary Sequestration (n=2)

Patients were presented with chronic cough and hemoptysis. In chest radiography of first patient consolidation of left - lower lobe was detected, likewise a similar consolidation was found in the right lower lobe of second case. Further evaluation was performed by computerized tomography scan. In the first case calcified hydatid cyst of left - lower lobe was erroneously reported (Fig. 1). In 31 for former and 23 months for later, follow-up period none of them revealed any sign or symptom of physical or respiratory limitations. The distribution of the lesion is shown in Table 1.

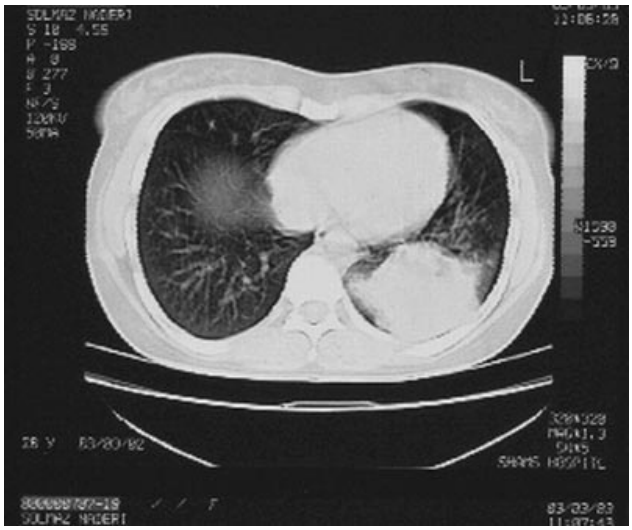

Fig 1. Intralobar pulmonary sequestration. CT scan showing calcified mass in left lower lobe misdiagnosed as a hydatid cyst.

DISCUSSION

CLE was the most common lesion in authors series. It is a marked pulmonary hyperinflation state that resembles all of the clinical features of obstructive emphysema. Respiratory symptoms appear as the lobe progressively over inflates; half of patients present within the first 2 days of life.[3]

This is a condition manifested during the first few months of life as progressively hyper expansion of one or more lobes of the lung. CLE usually occurs in the upper lobes of the lung (left greater than right), followed next in frequency by the right middle lobe, but it also can occur in the lower lobe.[4] In the present survey, upper lobes were afflicted in 6 patients, only one of authors cases suffering from lower lobe involvement. Males were affected twice as often as females, as previously reported.[5]

It is very important not to confuse CLE with a pneumothorax, as happened in one of their cases, an intercostals tube further increase the patient's respiratory distress, as reported earlier by Man *et al.*[5]

Surgical management consisted of lobectomy in 6 cases and segmentectomy in one. There was no morbidity or mortality in this group.

BS, the second common lesion in the present study, was diagnosed in 6 of the 16 patients. The clinical presentation of lung bronchogenic cysts (BS) is variable, such as respiratory distress at birth to late appearance of multifarious symptoms. Later on in life, cysts usually lose this compressive character and may remain asymptomatic until infection occurs; producing cough, dyspena, and thoracic pain.Lobectomy is the standard procedure, as the cyst is often surrounded by areas of atelectasis and pneumonia. Anatomic segmentectomy is reasonable for a small cyst.[6]

TABLE 1. Clinical Profiles of the Patients

Case	Age/sex	Disease	Symptom (I MS D H)	Treatment
1	13 days / M	CLE	+ (- + +-)	RUL
2	2 months / F	CLE	+ (- + +-)	emergent RUL
3	15 days / M	CLE	+ (- - + -)	Segmentectomy
4	43 days / M	CLE	+ (- + + -)	LUL
5	2 months / M	CLE	+ (- + + -)	LUL
6	30 days / F	CLE	+ (- + + -)	RLL
7	4 months / M	CLE	+ (- + + -)	LUL
8	12 years / M	BC	+ (+ - - +)	Cystectomy
9	14 years / M	BC	+ (+ - - -)	LLL
10	27years / F	BC	+ (+ - - +)	Segmentectomy
11	34 years / M	BC	+ (+ - - +)massive	emergent RML
12	13 years / M	BC	+ (+ - - -)	RLL
13	34 years / M	BC	+ (+ - - -)	LLL
14	24 days / M	CCAM	+ (- + + -)	LUL
15	22 days / M	CCAM	+ (- + + -)	LUL
16	9 days / M	CCAM	+ (- + + -)	Bilobectomy LU+M
17	26 years / F	ILS	+ (+ - - +)	RLL systemic blood supply
18	25 years / F	ILS	+ (+ - - -)	LLL

I, infection; MS, mediastinal shift; D, dyspnea. H, hemoptysis; RUL, right upper lobectomy; RML, right middle lobectomy; LUL, left upper lobectomy; LLL, left lower lobectomy; BC, bronchogenic cyst; CCAM, congenital adenomatoid malformation; CLE, congenital lobar emphysema; EPS, extralobar pulmonary sequestration.

Only two cases were treated with cystectomy and segmentectomy, and the other cases required lobectomy. One patient at the age of 34 years required emergency lobectomy because of massive hemoptysis due to rupture of cyst, but authors did have not any complications such as tension pneumothorax or empyema due to this event.

Most CCAM occurs in the left lower lobe.[4] However, occasionally CCAM may involve more than one lobe , as occurred in one patient in our series who had concordance involvement of both the left middle and upper lobe. The authors have three case of CCAM diagnosed in new born period and dyspnea was the most common presenting symptom. CT scan confirms the diagnosis, reveals associated anomalies, and better depict the anatomic extent of malformations.[7] In cases with bilobar or bilateral lesions prognosis is known to be poor because of pulmonary hypoplasia of the residual lung.[8]

Pulmonary sequestration is uncommon and consists of a mass of lung tissue, usually in the left lower chest. Intralobar sequestration more commonly occurs within the parenchyma of the lower lobe, but can occur on the right.[4] In authors patients which one of them had lesion on right lower lobe and the other one had it in left side. In first case who was suffering from left lower intralobar lesion which was erroneously reported hydatid cyst in chest CTscaning, furthermore was substantiated by serological studied. This patient sustained surgery by prediagnosis of hydatid cyst. At the time of operation the authors found a non-functioning lung tissue mass separated from the normal bronchopulmonary tree and vascularised by an aberrant systemic artery. Lobectomy was performed successfuly. In their experience, this pathology reaches clinical significance at the time of infection of the sequestered tissue.

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