Case Report

Congenital lobar emphysema

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ABSTRACT

Congenital lobar emphysema is a very rare congenital cystic malformation of the lung that can cause acute respiratory distress in early life. This paper reviews 6 cases of congenital lobar emphysema seen over a period of 10 years. The medical records of children with the diagnosis of congenital lobar emphysema were retrospectively reviewed for age at diagnosis, sex, presenting symptoms, investigations, treatment and outcome. There were 4 males and 2 females, and all of them presented before 6 months of age. Three presented with recurrent chest infection, while the other 3 had acute respiratory distress soon after birth. In all, the diagnosis was confirmed by chest x-ray, and the left upper lobe was affected in all of them. Although congenital lobar emphysema is rare, clinical awareness of this condition is important for early diagnosis and effective surgical treatment.

Keywords: Congenital lobar emphysema, congenital lung cysts, acute respiratory distress.

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Congenital cystic diseases of the lung comprise a group of closely related, rare, but potentially life-threatening anomalies. These include congenital cystic adenomatoid malformation, pulmonary sequestration, bronchogenic cyst and congenital lobar emphysema (CLE). Congenital lobar emphysema is characterized by massive distension of the affected lobe of the lung, usually the left upper lobe or the right middle lobe.[1] Expansion of the affected lobe as a result of air trapping and compression of the surrounding lung as well as the contralateral lung may cause life-threatening respiratory distress, which if not recognized and treated, may be fatal. We draw attention to this condition by reporting our experience with 6 cases, discussing their clinical features, diagnosis, and management.

Case Report. Over a period of 10 years from 1990 to 2000, 12 children with congenital cystic diseases of the lung were treated at Qatif Central Hospital, Qatif, Kingdom of Saudi Arabia (KSA). Six of them had congenital lobar emphysema. The medical records of these patients were retrospectively reviewed for age at presentation, sex, presenting symptoms, investigations, treatment and outcome. The operative details and histopathology were obtained from the operative notes and histopathology report. Their clinical and demographic features are shown in **Table 1**. There were 4 males and 2 females. Their ages at the time of surgery ranged from 7 days to 8 months. Three patients (number 2, 4 and 6 in **Table 1**) presented immediately after birth with acute respiratory distress, and whereas patient number 2 was referred for surgery soon, patient 4 on the other hand stayed in the hospital for 1½ months and was discharged home. This patient had 2 subsequent attacks of acute respiratory distress before he was referred for surgery. Patient number 6 presented immediately after birth with acute respiratory distress, but his condition improved subsequently and he was discharged home after one week. He was referred for surgery at the age of 4 months when he sustained an attack of acute respiratory distress. Patient number 3 presented at the age of one month with chest

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Table 1 - Clinical data for patients with congenital lobar emphysema.

N	Age	Sex	Birth Weight (kg)	Presentation	Site	Treatment
1	2 1/2 months	F	3.65	Recurrent attacks of chest infection from one month of age	LUL	Left upper lobectomy
2	7 days	M	4	Acute respiratory distress soon after birth	LUL	Left upper lobectomy
3	8 months	M	3.5	Recurrent attacks of chest infection from one month of age	LUL	Left upper lobectomy
4	4 months	F	3	Acute respiratory distress soon after birth	LUL	Left upper lobectomy
5	1 month	M	3.2	Recurrent attacks of chest infection	LUL	Left upper lobectomy
6	4 month	M	4.68	Acute respiratory distress soon after birth	LUL	Left upper lobectomy
N - number; F - female; M - male; LUL - left upper lobe						

infection and was diagnosed to have CLE, but the parents refused surgery. This patient suffered recurrent attacks of chest infection and finally had surgery at the age of 8 months. Patients number one and 3 were cousins. Patients number 5 had 2 attacks of pneumonia while patients number one had 3 attacks of pneumonia prior to surgery. In all 6 patients the left upper lobe was affected. The diagnosis in all of them was made by chest radiograph (**Figure 1**) which showed distension and dilatation of the left upper lobe as well as shift of the mediastinum and herniation of the emphysematous lobe to the right side. Five of our patients had computerized tomography (CT) scan of the chest which showed hyperinflation of the left upper lobe of the lung with right mediastinal shift and herniation of the emphysematous lobe to the right side behind the sternum pushing the trachea to the right. None of our patients had associated anomalies. All our patients underwent left thoracotomy and left upper lobectomy. Histological examination showed large markedly over distended alveolar spaces without tissue destruction consistent with CLE. Patient number one had, in addition, interstitial alveolar lymphocytic infiltrate, alveolar edema, congestion, focal increase in interstitial fibrous tissue and a striking number of desquamative mononuclear cells with prominent cytomegalovirus inclusions. Post-operatively all the patients did well except patient number one who developed a broncho-pleural fistula. This was treated conservatively but did not close and required operative closure. In spite of this the air leak persisted postoperatively for about 10 days before it finally stopped spontaneously. On follow-up ranging from 8 months to 9 years all patients are asymptomatic and well.

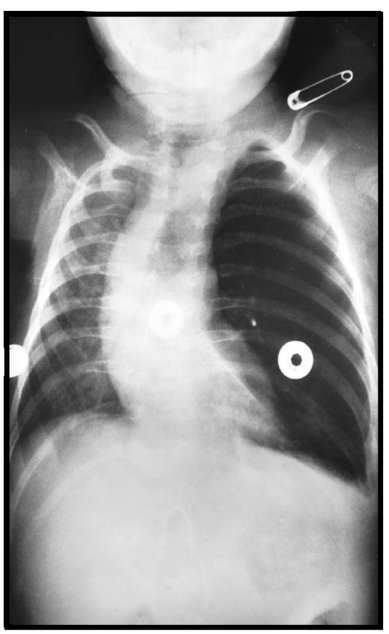

Figure 1 - Chest radiograph of patient number 3 showing massive distension of the left upper lobe. Note also the mediastinal shift and herniation of the distended lobe to the opposite side.

Discussion. Congenital lobar emphysema is a comparatively rare condition. Nuchtern and Harberg during a 10-year-period performed 1051 thoracic surgical operations, 22 (2.1%) of them were for congenital lung cysts, and of these only 6 (0.6%) were for CLE.[1] Wesley et al over a 10-year-period reviewed 22 patients with congenital lung cysts, 3 of them were for CLE.[2] In a collective series of 57 cases of congenital lung cysts reported from Riyadh, KSA, there were 37 cases of CLE.[3] Over a 10-year-period, we treated 12 children with congenital cystic diseases of the lung, 6 of them had CLE.

The exact etiology of CLE is not known. Several factors have been proposed, but congenital deficient cartilaginous support of the involved bronchus is present in 50% to 60% of these cases.[4,5] Hislop and Reid described the findings of a polyalveolar lobe in

some of these patients where the alveolar size may be normal, but the alveolar number is increased 3 to 5 folds.[6] Other rare identifiable causes include extraluminal obstruction and compression of the affected bronchus by abnormal blood vessel or congenital lung cyst, congenital bronchial stenosis and redundant bronchial mucosal flaps.[4,7-9] All of these factors lead to a ball-valve effect that permits inflation of the affected lobe during periods of negative intrathoracic pressure, but collapses and obstructs the affected bronchus with expiration. Ultimately this leads to air trapping and over expansion of the affected lobe of the lung. One of our patients was found on pathological examination to have an associated cytomegalovirus interstitial pneumonia. The significance of this from an etiological point of view is not exactly known. Interestingly, of the 6 patients treated, only this patient developed persistent post operative air leak.

Congenital lobar emphysema typically affects young infants less than 6 months of age and it is more common in males than females.[4] All our patients presented before 6 months of age and there were 4 males and 2 females. Although the majority of patients with CLE present early with acute neonatal respiratory distress, a small group of them may be asymptomatic or present at a later age with recurrent chest infection. In more than half the patients the left upper lobe is affected, followed by the right middle lobe. Although none of our patients had associated congenital malformations, CLE is frequently associated with congenital heart disease, which should be looked for during the evaluation of these patients.[10] We found chest radiograph to be diagnostic of CLE, but sometimes if the affected lobe is grossly distended it is not uncommon for this to be confused with tension pneumothorax. This is specially so in an infant with acute respiratory distress, but they can be differentiated by close and careful inspection of the chest radiograph by the presence of lung markings in CLE. Although this was not helpful in our patients, CT scan of the chest may be valuable in locating extrinsic etiological factors such as bronchogenic cyst, which must be considered whenever CLE is diagnosed.[11-13] The early age onset of CLE precludes the possibility of aspiration of a foreign body as an etiological factor. In addition, aspiration usually affects the lower lobe. When bronchoscopy is considered necessary, it should be performed in the operating room. The possibility of air trapping and progressive respiratory distress during the procedure must always be kept in mind and preparations should be made for emergency thoracotomy if necessary. Surgical treatment of CLE is total lobectomy which must be performed early to overcome the potentially life threatening acute respiratory distress and reduce the possibility of subsequent infection. Al-Bassam et al managed 5 children out of 37 with CLE conservatively and all did well with no surgical intervention.[3] Such a policy of conservative management can be adopted for patients who are stable, asymptomatic and can be followed-up closely. The importance of careful anesthetic induction cannot be over emphasized as these patients may not tolerate positive pressure ventilation which can lead to air trapping with rapid and massive enlargement of the affected lobe, mediastinal shift and cardiac arrest.[14] With improved safety of pediatric anesthesia, we found surgical resection to be both safe and effective in the management of CLE. Total lobectomy is tolerated well in infants and children as growth and expansion of the remaining lung tissue is known to occur in children up to the age of 5 years with subsequent total lung volume and function ultimately returning to normal.[15,16]

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