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Surgical treatment of congenital lobar emphysema: A report of nine patients

Konjenital lober amfizemin cerrahi tedavisi: Dokuz olgu sunumu

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Background: In this study, we aimed to present the outcomes of surgical treatment performed on nine pediatric patients with congenital lobar emphysema (CLE).

Methods: Between March 1996 and February 2011, nine children (5 boys, 4 girls; mean age 19.9 months; range 40 days-6 years) diagnosed with CLE who underwent pulmonary resection in our clinic were retrospectively analyzed. The symptoms, localization of the emphysematous lobe, diagnostic work-up, treatment modality, morbidity and mortality rates, and length of hospital stay were reviewed.

Results: Dyspnea was present in all of the patients, but it was severe in four patients. Emphysema was detected in the right middle lobe in three patients, the left upper lobe in three patients, the right upper lobe in one patient, and the left lower lobe in one patient. In the ninth and final patient, both the upper and middle right lobes were diseased. For all of the patients, the affected lobes with congenital emphysema were resected. None had postoperative morbidity or mortality, and the mean postoperative length of hospital stay was 5.8 days (range, 3-7 days).

Conclusion: Pulmonary resection in children is a treatment modality for CLE with low morbidity and mortality rates that eliminates the disease rapidly, safely and completely.

Key words: Child; congenital lobar emphysema; pulmonary resection.

Amaç: Bu çalışmada konjenital lober amfizem (KLA)'li dokuz çocuğa uygulanan cerrahi tedavinin sonuçları sunuldu.

Çalışma planı: Mart 1996 - Şubat 2011 tarihleri arasında kliniğimizde KLA tanısı konulan ve pulmoner rezeksiyon uygulanan dokuz çocuk hasta (5 erkek, 4 kız; ort. yaş 19.9 ay; dağılım 40 gün-6 yıl) geriye dönük olarak incelendi. Hastaların semptomları, amfizemli lobun yerleşim yeri, tanısı, tedavi şekli, morbidite ve mortalite oranları ve hastane yatış süreleri gözden geçirildi.

Bulgular: Dispne hastaların tümünde vardı, fakat hastalık dördünde ciddi idi. Amfizem üç hastada sağ orta lobda, üç hastada sol üst lobda, bir hastada sağ üst lobda ve bir hastada sol alt lobda tespit edildi. Dokuzuncu ve son hastada ise sağ üst ve orta lob birlikte tutulmuş idi. Konjenital amfizemden etkilenen loblar tüm hastalarda rezeke edildi. Ameliyat sonrası morbidite ve mortalite hiç kimsede gözlenmedi ve ameliyat sonrası ortalama hastane yatış süresi 5.8 (dağılım 3-7 gün) gün idi.

Sonuç: Konjenital lober amfizemli çocuklarda pulmoner rezeksiyon düşük morbidite ve mortalite oranları ile hızlı, güvenli ve hastalığı tamamen ortadan kaldıran bir tedavi yöntemidir.

Anahtar sözcükler: Çocuk; konjenital lober amfizem; pulmoner rezeksiyon.

Congenital lobar emphysema (CLE), a rare lesion that forms during the embryological development phase of the lung, is an uncommon clinical entity that generally presents as acute respiratory distress. The incidence of CLE is stated to be between 1/70.000 and 1/90.000 live births.[1] Congenital lobar emphysema is typified by hyperinflation of one or more lobes of the lung, which leads to compression atelectasis on the ipsilateral or

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contralateral side of the lung followed by a mediastinal shift.

Diagnosis of CLE in most cases occurs within six months of birth. Respiratory distress is the most common symptom that manifests during the perinatal period, and recurrent infections (together with respiratory distress) are frequently seen later in life.[2] In this study, we present the results of surgical treatment in our CLE patients over a 15-year period for the purpose of adding our knowledge of these cases to the published material available in the literature.

PATIENTS AND METHODS

Nine children (5 boys, 4 girls; mean age 19.9 months; range 40 days-6 years) were diagnosed with CLE in our clinic between March 1995 and February 2011. Diagnostic methods included posteroanterior (PA) chest radiography and computer tomography (CT). All of the patients underwent surgical treatment according to the localization of the lesion. The records of the patients were evaluated by the clinical findings, lesion location, surgical procedure, postoperative hospitalization time, and surgical outcome.

RESULTS

Dyspnea was present in all patients, and it was severe in four of the nine patients. A cough was present in eight cases, fever in seven, tachypnea in five, and wheezing in three; one patient had cyanosis and oxygen dependence. Emphysema was located in the right middle lobe in three cases, the left upper lobe in three cases, the right upper lobe in one case, the left lower lobe in one case, and in both the upper right and middle right lobes in one case (Figures 1-3). In all patients, the affected lobes were resected through a standard posterolateral thoracotomy (Table 1). Pathological examinations confirmed the presence of CLE in all cases.

All of the patients' symptoms improved after surgery. No postoperative morbidity or mortality occurred, and the average postoperative hospital stay was 5.8 days (range; 3-7 days).

DISCUSSION

Congenital lobar emphysema was first reported in 1954, and it is usually characterized by hyperinflammation in one pulmonary lobe.[3] The left upper lobe (43%) and the right middle lobe (32%) are most often involved, followed by the right upper lobe (20%) and bilateral involvement (20%).[4-6] Although more than one lobe may be affected, involvement of the lower lobes is rare. The incidence in male children is three times higher than that in female children.[2] In our study, all of the lobes, except for the right lower lobe, were involved at least once, and the right middle lobe and the left upper lobe were involved with the same frequency. One case presented in which the right middle lobe and the right upper lobe were both involved. Congenital lobar emphysema settled in the left lower lobe in one case, and we could not find any other instances of this described in the literature. No cases involving bilateral involvement were observed.

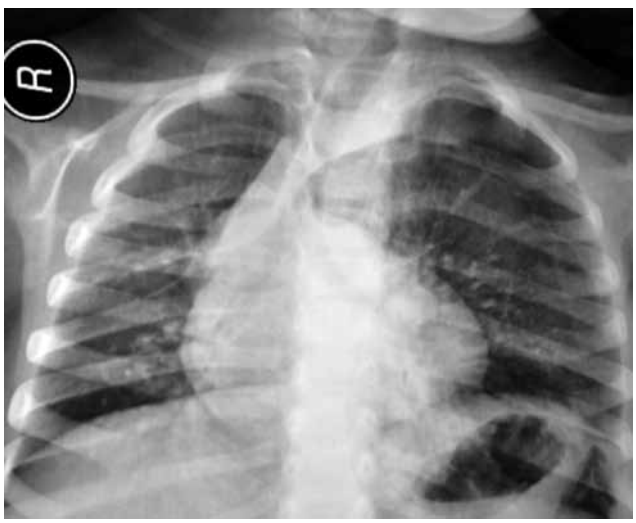

Figure 1. A patient with a hyperlucent lower lobe of the left lung that herniates to the right, with the displacement of the mediastinal structures also to the right; the upper lobe of the left lung and the right lung were compressed.

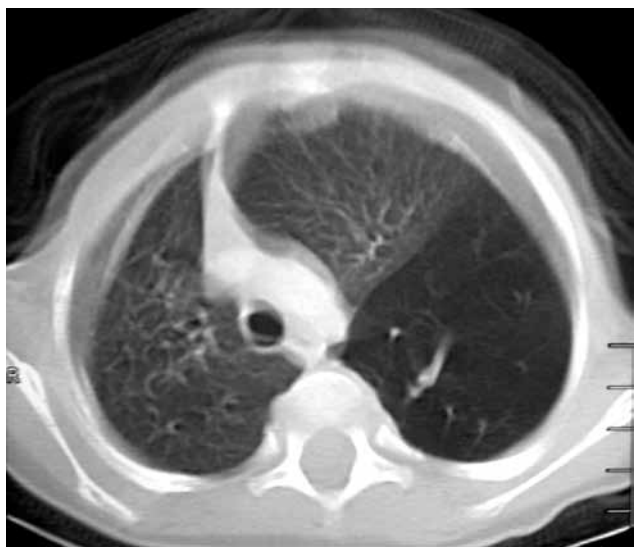

Figure 2. Computed tomography shows the hyperinflation of the entire lower lobe of the left lung, with patent vascular structures and a slight mediastinal shift to the right.

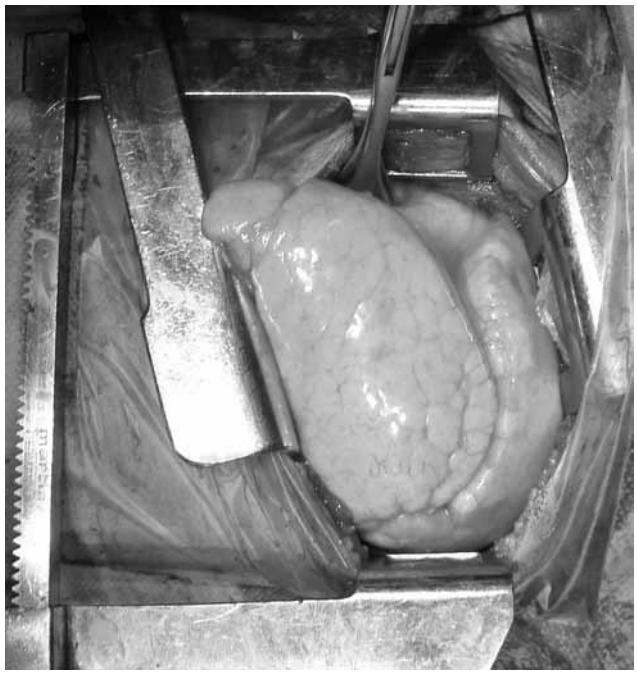

Figure 3. The upper and lower lobe of the left lung as viewed during surgery.

The etiology for CLE is unknown in almost half of patients. Entrapment of air due to the valve effect of the dysplastic bronchial cartilage, mucous plaques in the bronchus, aberrant veins compressing the bronchus, and bronchial disorders caused by infections are some of the known etiologies. In addition, one etiology of CLE involves polyalveolar lobe formation by numerous, normal-sized alveoli, but no destruction occurs at the alveolar wall. However, the alveolar count is three-five times higher than that for normal parenchyma.[2,7-9] The incidence rate of coexisting CLE and cardiovascular anomalies is 14%.[10] Anomalies such as renal agenesis, renal cyst, pectus excavatum, and diaphragmatic hernia as well as gastrointestinal and extremity anomalies may also coexist.[11,12] However, these congenital anomalies did not appear in our nine cases.

Myers[13] has classified CLE into three clinical types: CLE type 1 is symptomatic in infants, CLE type 2 in adolescents, and CLE type 3 signifies that the patient is asymptomatic (incidental diagnosis). Most patients are symptomatic in the neonatal period, and types 2 and 3 are rare. Symptom onset usually occurs in the first week in half of the patients and within the first six months of life in the remaining patients while the number of reported cases with CLE detected in adulthood is very limited. Congential lobar emphysema is the most common etiology for neonatal respiratory distress syndrome, which is usually seen in the first six months of life.[2] In our study, all patients had respiratory distress to varying degrees. The other most commonly observed symptoms were cough and fever. The mean age of symptom onset was 19.9 months, which is compatible with type 1 CLE. No cases of adolescent, asymptomatic, or incidental diagnosis were uncovered.

On physical examination, hypersonority, reduced respiratory sounds, and deviation of the trachea to the opposite side have been detected in the involved side of the lungs, with the compression of healthy pulmonary tissue resulting in dyspnea, cyanosis, reduced venous circulation, hypertension, and eventually cardiac arrest. The diaphragm may shift downwards bilaterally. Direct thoracic radiography is usually sufficient to establish a diagnosis of CLE, but in suspected cases, CT also aids in the diagnosis.[14] On thoracic radiographs and CT scans, hyperlucency, a collapsed adjacent lobe, and a mediastinal shift as well as a hyperinflated lobe with herniation to the other side have been observed. Tense and thinned veins have also been observed in the emphysematous lobe on CT.

The diagnosis was obtained for all of our cases via PA chest X-ray images and thoracic CT scans. The differential diagnosis of CLE usually includes pneumothorax, pulmonary hypoplasia, pneumatocele

Table 1. Characteristics of cases

Age	Gender	Localization	Treatment
40 days	Female	Right middle lobe	Middle lobectomy
45 days	Male	Right middle lobe	Middle lobectomy
2 months	Male	Left upper lobe	Upper lobectomy
6 months	Female	Left upper lobe	Upper lobectomy
18 moths	Male	Right middle lobe	Middle lobectomy
18 moths	Female	Left upper lobe	Upper lobectomy
2 years	Female	Lef lower lobe	Lower lobectomy
3 years	Male	Right upper lobe	Upper lobectomy
6 years	Male	Right upper and middle lobe	Bilobectomy superior

and endobronchial mass, and congenital cystic adenomatoid malformation. A congenital diaphragmatic hernia and foreign body aspiration should also be kept in mind. In many cases, tension of the lobe (excessive air) is mistaken for pneumothorax, and a thoracostomy is performed. This does not lead to clinical relief but causes the clinical picture to deteriorate.[2]

In infants with severe respiratory symptoms, pulmonary resection is needed to avoid morbidity and mortality. The recommended treatment is a lobectomy. However, some authors have reported that conservative treatment can be applied in asymptomatic patients or in patients with minimal symptoms.[12,15] Infants with severe respiratory distress may need an urgent thoracotomy and a lobectomy. During surgery, excessive expansion of the lungs should be avoided until the thorax is opened to prevent an increase in the mediastinal shift and compressive shock. After the thorax is opened, the emphysematous lobe should be removed from the chest, and the lobectomy should commence. Our cases were all treated surgically. A lobectomy was applied in eight of the cases while one patient underwent a bilobectomy. No postoperative morbidity and mortality were observed, and all patients were completely cured.

To conclude, CLE is a rare, but potentially life-threatening abnormality which affects infants. Neonatal and childhood cases with progressive respiratory distress must be considered as possible CLE, and the diagnosis should be confirmed by radiological evaluation. Pulmonary resection has excellent results in most of the symptomatic cases.

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