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Konjenital Lobar Amfizem

CONGENITAL LOBAR EMPHYSEMA

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Özet

1993-2001 yılları arasında cerrahi olarak tedavi edilen 4 konjenital lobar amfizemli olgu gözden geçirildi. Olguların 3'ü erkek, 1'i kızdı ve yaşları 28 gün, 2, 3 ve 10 aydı. Tekrarlayan akciğer enfeksiyonları, dispne, siyanoz ve sıkıntılı solunum ana bulgulardı. Olguların 3'ünde sol üst lob tutulmuştu. Göğüs x-ray ve bilgisayarlı tomografi tutulmuş lobların hiperaerasyonunu gösteriyordu. Bütün olgularda lobektomi yapıldı ve postoperatif dönem sorunsuz geçti. Solunum sıkıntısı ile gelen her infantta konjenital lobar amfizemden şüphelenilmelidir. Başarılı medikal tedaviler bildirilmesine rağmen, tutulan lobun cerrahi rezeksiyonu kabul edilen tedavi yöntemidir.

Anahtar kelimeler: Konjenital lobar amfizem, lobektomi

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Summary

We reviewed four patients with congenital lobar emphysema who had been treated surgically between 1993-2001. Three were boys and one was girl aged 28 days, 2, 3 and 10 months. Recurrent attacs of chest infections, dyspnea, cyanosis and respiratory distress, were the main presentation. The affected side was the left upper lobe in three patients. Chest x-ray and computerized tomography scans showed hyperaeration of the affected lobes. Lobectomy was performed in all cases and postoperative course was uneventful. Congenital lobar emphysema should be suspected in any infant presenting with breathlessness. Despite reports of succesful conservative management, the accepted treatment is surgical lobectomy of the affected lobe.

Keywords: Congenital lobar emphysema, lobectomy

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Introduction

Congenital lobar emphysema (CLE) is a rare condition of respiratory distress during infancy, and it is characterized by hyperinflation of a lobe in the absence of extrinsic bronchial obstruction [1]. The left upper lobe is involved most often, followed in incidence by involvement of the right middle lobe [1]. Gross and Lewis, published the first successful surgical treatment of CLE. Clinically the condition may present varying severity. In approximately one-third of cases, severe respiratory distress occurs soon after birth, and rapid deterioration may follow. The remaining two-third of patients have a slower onset, usually at one or two months of age, often coinciding with respiratory infection [2]. Chest radiograph is sufficient to establish the diagnosis in the vast majority of patients. The treatment of choice is surgical removal of the affected lob in patients who have persistent or progressive, severe respiratory distress that those not respond to medical treatment.

Cases

We reviewed four patients with CLE who had been treated surgically between 1993-2001. Four children consisting of 3 boys and 1 girl aged 28 days, 2, 3 and 10 months. Recurrent attacks of chest infection, dispnea, ciyanosis and respiratory distress, which started during the early neonatal period, were the main presentation. On admission three babies had tachypnea, retraction of the chest wall and cyanosis, and another had episode of cyanosis after birth. Each received several forms of medical treatment without benefit. One patient associated with imperforate anus and others had no any congenital anomalies. The patient who had imperforate anus had been treated with a right transverse loop colostomy two days after birth at another institute, but CLE not recognized at the same time. In all cases, chest x-ray and computed tomography (CT) was diagnostic of CLE. Chest x-rays showed hyperaeration of the affected lobes in all cases (Figure 1). Mediastinal shift and lobar or segmental atelectasis with herniation of the emphysematous lobe were seen in three cases. Computed tomography of the chest was performed in three patients and showed hyperaeration of the affected lobe, mediastinal shift and atelectasis in all patients, and consolidation in 2 patients (Figure 2). Pulmonary perfusion scan with technetium 99m macroaggregates of human serum albumin (99mTc MAA) was used in 2 patients and showed loss of perfusion in the affected lobes (Figure 3). The affected side was the left upper lobe in three patients, and right upper lobe in one patient. Tube thoracostomy was performed in one patient who had misdiagnosed initially as pneuomothorax. A chest drain was inserted with no further air drainage no clinical

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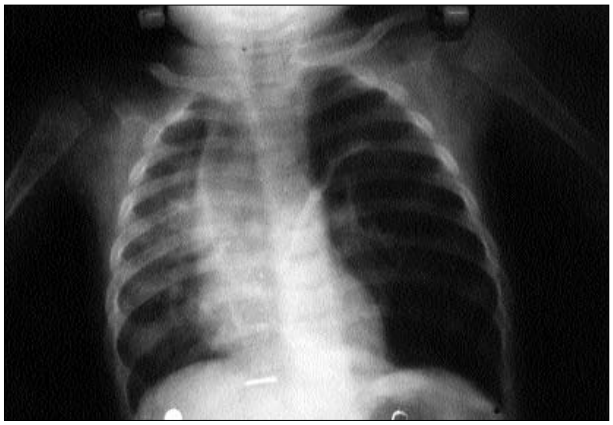

Figure 1. Chest x-ray showing left upper lobe emphysema with mediastinal shift in preoperative period.

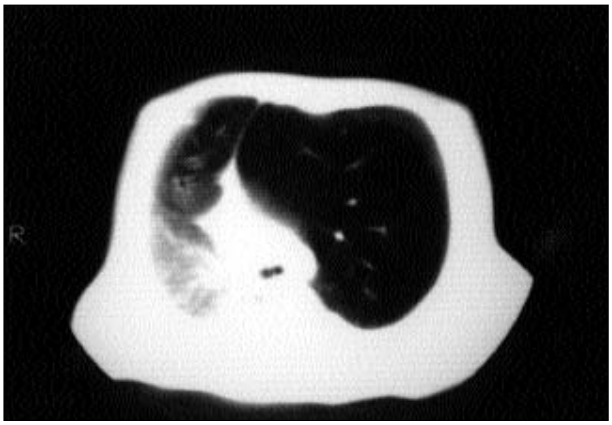

Figure 2. Chest CT showing emphysematous left upper lobe, mediastinal shift and pneumonic consolidation in the right lung.

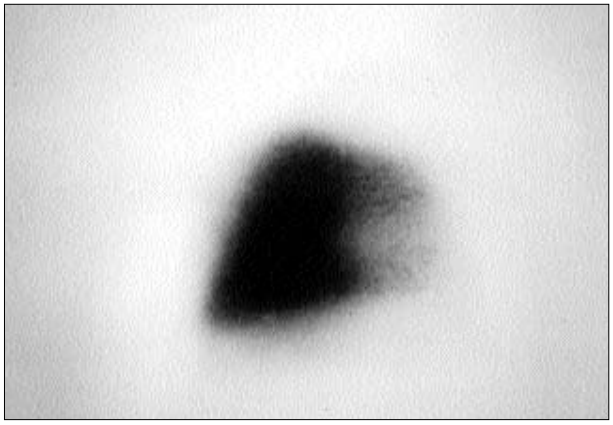

Figure 3. Pulmonary perfusion scan [^{99m}Tc MAA] showing loss of perfusion in the left upper lobe.

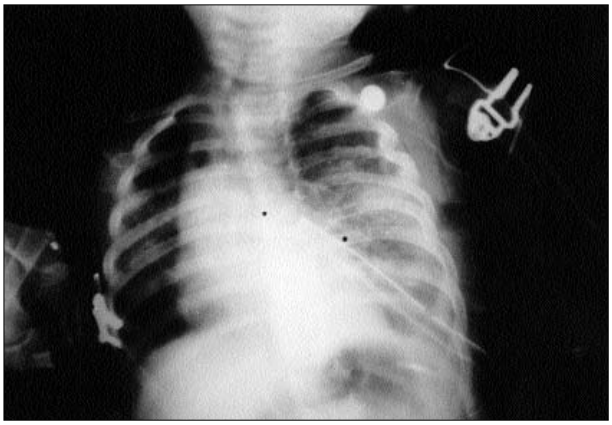

Figure 4. Chest x-ray showing loss of emphysematous lobe on postoperative third day.

improvement. Bronchoscopy was performed in two patients in order to determine the presence or absence of abnormalities of the bronchial tree mucous plaque or a foreign body. No foreign body aspiration or mucous plaque was found. To exclude any associated cardiac defect or vascular abnormalities that could accompany CLE, echocardiography was performed in all patients and no additional cardiac anomaly was found. Lobectomy was performed in all patients (Figure 4). Postoperative course was uneventful for all patient. One patient was lost to follow-up after the first postoperative check-up. The remaining two patients were found to be well at regular follow visits. The data about clinical features, diagnostic methods, and treatment are summarized in (Table 1).

Discussion

Congenital lobar emphysema is a rare cause of respiratory distress during infancy which is cured by surgery. Normally male babies are affected more often than females. Congenital lobar emphysema should be suspected in any infant presenting with breathlesness. It may present immadiately at birth, in the neonatal period or in infancy [2]. It is usally unilateral, affecting the left upper lobe, followed by the right middle lobe and right upper lobe, althoug bilateral involvement has been reported [3]. However, in our series the left upper lobe was most common affected (3/4). The exact cause of CLE is to determine, and no apparent cause is found in 50% of cases [4]. The current theory suggests inadequate cartilagenous support of the bronchus, has been by. It has been found an increase in the number of alveoli in the affected lobe.

Congenital lobar emphysema produces symptoms in infancy. Often, a history of tachypnea, retraction of the chest wall, and wheezing since birth exists [1,2]. In our cases, the onset of the symptoms was typically in early infancy. Physical examination reveals a shift of the trachea and mediastinum to the contrlateral hemithorax. Breath sounds are decreased on the affected side, with associated hyperresonance. The diagnosis is usually confirmed on a plain chest radiograph, often with mediastinal displacement and herniation of the affected lobe may compress the contrlateral lung [2], and in these cases lung isotope scanning is indicated to differentiate CLE from hypolasia of the contrlateral lung [5]. On a plain chest x-ray, careful inspection of vascular markings reduces the risk of

Table 1. Clinical Features, diagnosis, management and results of patients with CLE.

Case no.	1	2	3	4
Sex	male	male	female	male
Age	28 days	2 months	3 months	10 months
Age of onset of symptoms	2 days	2 days	1 week	2 months
Presenting symptoms	Tachypne, cyanosis	Tachypnea, dispnea, cyanosis	Tachypnea, cyanosis	Cough, respiratory infection, dyspnea
Additional anomalies	-	Imperforated anüs	-	-
Affected lobe	LUL	RUL	LUL	LUL
Diagnostic methods	Chest x-ray, CT, PS	Chest x-ray, CT, PS	Chest x-ray	Chest x-ray, CT
Treatment	Lobectomy	Lobectomy	Lobectomy	Lobectomy
Results	Well at 1 year	Well at 3 years	Lost to follow-up after first check-up	Well at 5.5 years

CT = computed tomography; LUL = left upper lobe; RUL = right upper lobe; PS = perfusion scan

misdiagnosis of this lesion as a tension pneumothorax. One of the our patients initially interpreted as pneumothorax, and diagnostic aspiration was performed at another institution. A chest drain was inserted with no further air drainage and no clinical improvement. Attempts at needle aspiration must be avoided, for this almost results in a tension pneumothorax that can be fatal. The radiological differential diagnosis includes pneumothorax, pneumatocele, atelectasis, or hypoplasia of the lung with hyperinflation of the contrlateral lung, diaphragmatic hernia, and congenital cystic adenomatoid malformation [8]. In an infant with mild to moderate respiratory distress, CT can establish the diagnosis of CLE by showing the hyperlucent expanded lobe and stretched, attenuated vessels and CT can also exclude extrinsic causes of lobar emphysema, such as vascular anomalies or mediastinal mass. Chest CT was performed in three patients and confirmed the radiographic finding. Bronchoscopy can worsen patients condition, but may be useful in excluding non-opaque foreign as the cause of the hyperinflation in older children [5]. Bronchoscopy was performed in one patient aged ten months, who presented with acute respiratory symptoms suggestive of foreign body inhalation. The ventilation / perfusion radionucleide scan is useful in evaluating the function of the affected and compressed lobes. However, it is difficult to perform in a small infant.

Despite reports of successful conservative management [6] the accepted treatment is surgical lobectomy of the affected lobe. Operative mortality rate is 3% to 7% whereas with conservative therapies it is 50 to 75% [7]. The conservative treatment may be considered in selected infants and older children with mild symptoms. In neonates and infants with severe respiratory distress, emergency thoracotomy may be life saving. In the present study emergency lobectomy was performed in one patient. At operation, the chest is opened as soon as possible after induction of anesthesia. Positive-pressure ventilation causes further overinflation of the involved lobe and increases the risk of mediastinal shift and cardiac arrest. Before resection, the mediastinum must be carefully examined for lesions that could have obstructed the bronchus.

We feel that surgical excision of the affected lobe could be managed as soon as possible in all infants younger than 2 months, and in infants older than 2 months who present with severe respiratory symptoms. Conservative management can be performed in older children who present with mild symptoms with a close follow-up of the patient.

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