Congenital Lobar Emphysema in An Adult Patient: Case Report

Erişkin Bir Olguda Konjenital Lober Amfizem

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ABSTRACT Congenital lobar emphysema (CLE) is a rare bronchopulmonary malformation which may cause severe respiratory distress with symptoms such as cough, dyspnea, tachypnea, retraction of the chest, wheezing and cyanosis in the neonatal period. Several mechanisms of CLE have been proposed, including dysplastic or absent bronchial cartilage, endobronchial obstruction, extensive mucosal proliferation and infolding, extrinsic compression of the bronchi by the aberrant cardiopulmonary vasculature, and diffuse bronchial abnormalities. However, approximately half of these are due to unknown causes. Typical findings include overinflation of a pulmonary lobe with mediastinal shift to the contralateral side and compression in the adjacent parenchyma. Most children with CLE present symptoms before 6 months of age. Diagnosis of CLE in adulthood is unusual, and patients often present with mild symptoms. We discussed a case of CLE in an adult who diagnosed intraoperatively and treated with lobectomy.

Key Words: Congenital, emphyema, adult, surgery

ÖZET Konjenital lober amfizem (KLE) yenidoğan döneminde siyanoz, "wheezing", göğüsün çökmesi, dispne ve takipne gibi ciddi solunum sıkıntısı belirtileri gösteren nadir bir bronkopulmoner malformasyondur. KLE etiyolojisinde; bronşiyal kıkırdağın agenezi veya displazisi, endobronşiyal obstrüksiyon, yoğun mukozal proliferasyon veya içe katlanması, aberan kardiyopulmoner vasküler yapılar ile bronşun dıştan basıya uğraması ve diffüz bronş anormallikleri gibi birçok nedenler ileri sürülmüştür. Ancak yarıya yakınının nedeni henüz açıklanabilmiş değildir. Tipik bulguları, karşı tarafa mediastinal şift ile birlikte pulmoner lobun aşırı şişmesi ve komşu parankimin kompresyonudur. KLE genellikle çocuklarda ilk 6 ay öncesi görülmektedir. Bundan dolayı erişkinlerde KLE semptomları genellikle hafif derecededir. Bu çalışmada, intraoperatif tanısı konulan ve lobektomi uygulanan KLE'li erişkin bir olgu tartışılmıştır.

Anahtar Kelimeler: Konjenital, amfizem, erişkin, cerrahi

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Congenital lobar emphysema (CLE) is a rare bronchopulmonary malformation that presents before 6 months of age. CLE is characterized by overinflation of a pulmonary lobe with mediastinal shift to the contralateral side and compression in the adjacent parenchyma. CLE usually presents just after the birth with symptoms like wheezing, cyanosis, retraction of the chest, tachycardia, dyspnea and tacypnea. Any infant with CLE and moderate to severe respiratory symptoms should be treated with lobectomy.[1,2] In literature, there are only a few cases that were diagnosed in the adulthood.[3,4]

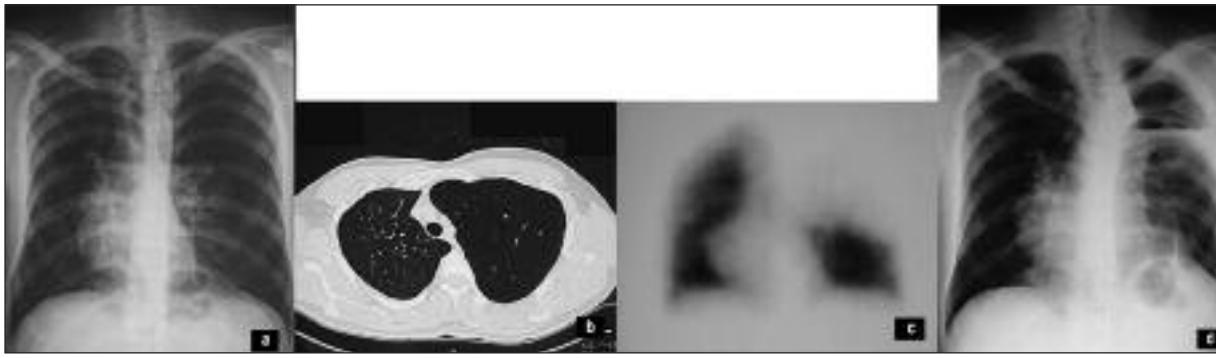

FIGURE 1: Hyperlucency on the upper zone of the left hemithorax and mediastinal shift to the right side in preoperative chest x-ray **(1a)**; hyperinflation of the left upper lobe, shift of the mediastinal structures to the right side and diminished anterior posterior diameter of the chest on CT **(1b)**; decreased perfusion in the involved lobe **(1c)**; the left lower lobe filled only 2/3 of the hemithorax in the postoperative third month chest x-ray **(1d)**.

CASE REPORT

A 22 year-old man was referred to our outpatient clinic with several years history of dyspnea. He used to smoke 1.5 boxes of cigarette/year. In his physical examination; he had pectus excavatum, and kyphoscoliotic chest deformity. Thoracic vibration and breath sounds were decreased in the upper zone of the left hemithorax and there was hypersonority with percussion. The laboratory parameters were in normal limits. Preoperative spirometry results were as follows: FVC: 5.28 L (104.5%), FEV_1: 3.44 L (79.9%), FEV_1/FVC: 78.3%. Preoperative arterial blood gases measurements were in normal limits. On chest X-ray there was hyperlucency in the upper zone of the left hemithorax and mediastinal shift to the right side (Figure 1a). Hyperinflation of the left upper lobe shifted the mediastinal structures to the right side. The anterior-posterior diameter of the chest was diminished in computerized tomography (CT) (Figure 1b). We did not observe any pathologic conditions on fiberoptic bronchoscopy (FOB). A lung perfusion scan was performed and a decreased perfusion of the involved lobe was demonstrated (Figure 1c). We determined that the lower lobe filled only 2/3 of the hemithorax in the postoperative third month chest x-ray (Figure 1d).

Our differential diagnosis included bullous lung disease, Swyer James McLeod syndrome, and lobar emphysema. We performed left sided thoracotomy under endotracheal double lumen tube anesthesia. The emphysematous upper lobe was still over inflated after single lung ventilation (Figure 2a). The lobe felt like sponge rubber, did not deflate, and bounced back into shape after it was

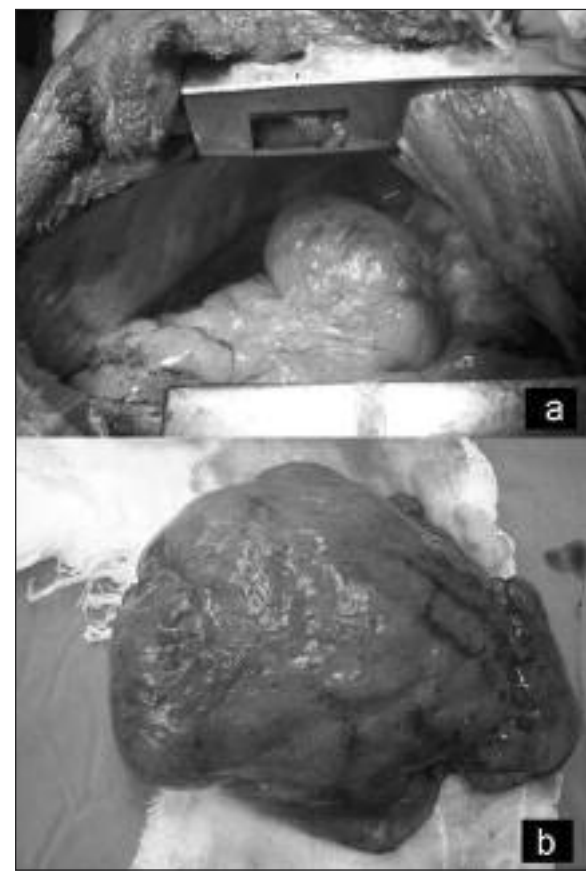

FIGURE 2: (a,b): The emphysematous upper lobe was still overinflated after single lung ventilation (2a), the lobe felt like sponge rubber, did not deflate, and bounced back into shape after it was compressed. After lobectomy, the emphysematous upper lobe did not deflate (2b).

compressed. After lobectomy, the emphysematous upper lobe did not deflate (Figure 2b). With these findings, we concluded that the patient had CLE. Histopathologic examination revealed multiple bullous ectasia near the pleura. There were fibrous thickening, hyalinization, focal lymphocyte accumulation, and calcification and ossification areas on the wall of bulla. The alveoli were lined by thin, depressed pneumocyt cells and this was consistent with widespread emphysema. The peripheral bronchus was normal, but no cartilage was seen in the distal bronchus. There were local fibrosis and lymphocyte infiltration in the wall of bronchus. The postoperative period was uneventful. A spirometry test and arterial blood gas measurements were performed in the 3rd month of the follow up. Spirometry results were as follows: FVC: 3.36 L (66.5%), FEV_1: 2.79 L (64.6%), FEV_1/FVC: 99.6%. These measurements were as follows in 12th month postoperatively: FVC: 4.86 L (96.5%), FEV_1: 4.19 L (86.2%), FEV_1/FVC: 101.6%. The patient is still under follow up.

DISCUSSION

Congenital lobar emphysema (CLE) is one of the unusual childhood respiratory tract disorders that is characterized by overinflation of a pulmonary lobe, air trapping in the affected lobe, compression of the adjacent parenchyma, mediastinal shift to the contralateral side and herniation of emphysematous lobe across the opposite side of the chest. Several mechanisms identified in the etiology of CLE are; dysplastic bronchial cartilage, endobronchial obstruction, hypoplastic or absent bronchial cartilage and extrinsic bronchial obstruction due to an aberrant cardiopulmonary vasculature.[1,5,6]

However, it is difficult to determine the exact cause of CLE, and no apparent cause is identified in approximately half of the cases. It is seen in boys more than the girls. The most frequently affected lobe is the left upper lobe which is followed by the right middle lobe and the right upper lobe.[1,2,4,5] Our case was a young man and had CLE in his left upper lobe as most frequently reported in the literature.

Nearly half of the infants with CLE develop symptoms in the first few days of the life and 95% of the children present before 6 months of age. Some patients are misdiagnosed as pneumothorax or airway foreign body. The most common symptoms are tachypnea, dyspnea, cyanosis, retraction of the chest, wheezing and coughing. The breath sounds are decreased at the affected lobe.[1,7] As in our case, only a few cases have been reported who were diagnosed in adulthood.[3]

CLE in adulthood is usually asymptomatic or presents with very mild symptoms including coughing, dyspnea and respiratory infections. In some patients CLE is associated with other congenital or acquired diseases such as pectus excavatum, right-sided aortic arch, congenital heart defects, bilateral diaphragmatic eventration and mediastinal bronchogenic cyst.[1,5,6] Our case had slowly increasing respiratory distress. On physical examination, he had pectus excavatum and kyphoscoliosis. The breath sounds were decreased at the affected lobe on auscultation.

CLE is still confused with other emphysematous diseases and congenital cystic adenomatoid malformation (CCAM). CLE is diagnosed with clinical, radiological and histological findings. A plain chest X-ray is the initial step of radiological evaluation for the diagnosis of CLE. Typical findings seen on the chest X-ray are; hyperlucency in the involved lobe, mediastinal shift to the contalateral side and flattening of the ipsilateral diaphragm. The radiologic findings of CCAM are the presence of a solid mass covered with multiple cystic areas, and the mediastinal shift.[1,6,8] In our case, hyperlucency in the left upper lobe and mediastinal shift to the right side were seen on chest X-ray. The left lower lobe was compressed because of the overinflation of the left upper lobe and there was mediastinal shift to the right side on CT. No bronchial obstruction or mediastinal masses were seen.

Some authors recommend a radionuclide ventilation-perfusion scan to confirm that the involved lobe is not functioning. Characteristic findings on a radionuclide ventilation-perfusion scan is the decreased perfusion (secondary to the compression

of surrounding blood vessels) and the reduced ventilation of the involved lobe.[1,5,6] In our case, a perfusion scan was performed and decreased perfusion was seen in the left upper lobe, however the perfusion was normal in the left lower lobe and in the right lung.

It is difficult to diagnose CLE preoperatively in an adult. Intraoperative findings may be helpful for the diagnosis.[1,5,6] The intraoperative findings in our case were the emphysematous upper lobe which was still overinflated after single lung ventilation, sponge-like appearance of the lobe and bouncing back into shape after it was compressed, and after lobectomy, the failure of the emphysematous upper lobe to deflate. We concluded that the patient had CLE with these findings.

Several mechanisms of CLE have been proposed, including dysplastic or absent bronchial cartilage, endobronchial obstruction, extensive mucosal proliferation and infolding, extrinsic compression of the bronchi due to an aberrant cardiopulmonary vasculature, and diffuse bronchial abnormalities. However, approximately half of the cases are due to unknown causes.[1,5,8] In our case, we couldn't find any definitive cause.

The universally accepted treatment of CLE with moderate to severe respiratory symptoms is lobectomy. Infants who are asymptomatic or those with milder symptoms may be carefully observed although this remains controversial. The treatment of CLE in adulthood is surgical resection in order to avoid the compression of the other lobe and mediastinal shift.[1,2,4,5] We performed lobectomy in our case because of increasing respiratory distress. After left upper lobectomy, the remaining lower lobe had only filled the half of the left hemithorax on the postoperative 17th day. The left lower lobe filled only the 2/3 of the hemithorax during at the postoperative 3rd month follow up. Our patient is under follow up for 12 months and the space in left hemithorax regresses gradually.

The findings suggest that the prognosis in patients with CLE diagnosed in early infancy is good after lobectomy, and an adaptive mechanism occurs in a period from a few months to a few years. There is a permanent loss of lung volume and function after lobectomy in adults.[1,2,4,5] In our case, spirometry values decreased in postoperative 3rd month when compared to the preoperative values. The spirometry values returned to normal after the 12th month.

CLE is usually diagnosed in the childhood. If it is seen in adulthood, it can be misdiagnosed. We should keep in mind CLE in adult patients with dyspnea, mediastinal shift and air trapping in the chest X-ray.

REFERENCES

1. Reynolds M. Congenital lesions of the lung. In: Shields TW, Locicero J, Ponn RB, Rusch VW, eds. General Thoracic Surgery. 6th ed. Philadelphia: Lippincort Williams and Wilkins; 2005. p.1104-5.
2. McDonald CF, Pierce RJ, Barter CE, Chou ST, Daniel FJ. Congenital lobar emphysema requiring surgery in adult life. Aust N Z J Med 1986;16(4):501-5.
3. Canneto B, Carretta A, Vagani A, Puglisi A, Pansera M, Zannini P. [Congenital lobar emphysema in adults]. Minerva Chir 2000;55(5): 353-6.
4. Critchley PS, Forrester-Wood CP, Ridley PD. Adult congenital lobar emphysema in pregnancy. Thorax 1995;50(8):909-10.
5. Shanmugam G. Adult congenital lung disease. Eur J Cardiothorac Surg 2005;28(3): 483-9.
6. Stanton M, Davenport M. Management of congenital lung lesions. Early Hum Dev 2006; 82(5):289-95.
7. Toksoy N, Yalın T, Bernay F, Arıtürk E, Gürses N. [Congenital lobar emphysema]. Turkiye Klinikleri J Pediatr 1999;8(4):226-8.
8. Zylak CJ, Eyler WR, Spizarny DL, Stone CH. Developmental lung anomalies in the adult: radiologic-pathologic correlation. Radiographics 2002;22:S25-43.