Congenital Lobar Emphysema That Seen In Two Siblings
İki Kardeşte Gözlenen Konjenital Lober Amfizem

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Abstract

Congenital lobar emphysema (CLE) is usually characterized by hyper-inflammation in one lobe. The left upper lobe (50%) and the right middle lobe (30%) are most often involved, followed by the right upper lobe. Family history and CLE relation did not mentioned in literature. In this article we present two siblings with CLE; one of them was 3 years old boy with right upper lobe CLE patient and the other one is, his 40 day old sister with right middle lobe CLE.

Keywords: Congenital lobar emphysema, sibling

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Introduction

CLE is rare lesion that forms during the embryological development phase of the lung. It is usually characterized by hyper-inflammation in one lobe. Majority of patients take diagnose of CLE within 6 month of birth. Respiratory distress is the common presenting symptom at perinatal period. Recurrent infections together with respiratory distress are common presenting symptom in later periods of life ¹.

Congenital heart failure, renal agenesis and pectus excavatum are common reported congenital pathologies that associated with CLE ². Together with one article that presents twin brothers with CLE, family history and CLE relation did not mentioned in literature. In this article we present two siblings with CLE; one of them was 3 years old boy with right upper lobe CLE patient and the other one is, his 40 day old sister with right middle lobe CLE.

Cases

Case 1. A 3 year-old male child who had suffered from cough, fever and dyspnea was admitted to our clinic. The symptoms of the patient were started 6 months before. Symptoms had a gradual onset and had shown progression with time. Examination of respiratory system revealed less movement of right upper chest, trachea was shifted to the left, vocal resonance was decreased in right side and on auscultation diminished breath sound...
was found on right side. Thorax computed tomography (CT) showed prominent hyperaeration and stretched attenuated vessels in the upper lobe of the right lung. Middle and lower lobes of the right lung and the left lung were compressed. There was to left shift in the mediastinum and herniation of the hyperinflated lobe. Right thoracotomy was performed to patient. At middle and lower lobes of the right lung there was atelectasis. The patient underwent a right upper lobectomy. His respiratory functions were not anomaly in the period postoperative. The patient discharged home on postoperative fifth day.

Case 2. Five year after the first case the second child of the same parents was evaluated. A 40 days old female child who was tachypnea, cough, wheezing, cyanosis, serious dyspnea and dependent on oxygen was admitted in our hospital. Thorax computed tomography showed prominent hyperaeration and stretched attenuated vessels in the middle lob of the right lung. Upper and lower lobes of the right lung and the left lung were compressed. There was to left shift in the mediastinum and herniation of the hyperinflated lobe (Fig 1). Right thoracotomy was performed to patient (Fig 2). At gross examination, the bronchi were extremely thin and poorly developed. At upper and lower lobes of the right lung there was atelectasis The patient underwent a right middle lobectomy. The patient discharged home on postoperative third day.

There is no relationship between parents and socioeconomic level of family was moderate.

**Discussion**

Congenital lobar emphysema was first reported in 1932 by Nelson. Vogt-Moykopf reported the rate as 3.5% in their 117 congenital lung anomalies series. It is usually characterized by hyper-inflammation in one lobe. The left upper lobe (50%) and the right middle lobe (30%) are most often involved, followed by the right upper lobe. One of two patients of our study was male and his emphysematous lesion located in right upper lobe. Lesion of the female patient was located in the right middle lobe.

The etiology for CLE is not known for 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 infections causing bronchial disorders are some of the known etiologies. In addition, one etiology of CLE involves polyalveolar lobe formation by numerous alveoli with normal size. Anomalies such as cardiac anomalies,
renal agenesis, renal cyst, pectus excavatum, diaphragmatic hernia, and extremity anomalies may also coexist. However in our cases cardiac and other anomalies were absent. Thompson et al. first reported CLE in twins in 2000. Our two sibling patients are the first reported sibling CLE cases who are not twin.

The onset of the symptoms is usually in the first week in half of the patients and within 6 months in the other half. In our study one of the patients is 3 years old and the other one is 40 day old. Type of CLE is type I in both patients. Symptoms of older sibling were started after two years old and get progressively worse thereafter. On the other hand symptoms of younger sibling were started within a week after birth and the patient was dependent to oxygen supply.

Direct thoracic radiography is usually sufficient in establishing a diagnosis of CLE. In suspected cases, CT aids in diagnosis. On thoracic radiograph and CT, hyperlucency, collapsed adjacent lobe, and mediastinal shift as well as hernia of the hyperinflamed lobe to the other side are observed. On CT, tense and thinned veins are also observed in the emphysematous lobe.

In infants with severe respiratory symptoms, pulmonary resection is needed to avoid morbidity and mortality. The recommended treatment is lobectomy. However, some authors have reported that, in asymptomatic patients or in patients with minimal symptoms, conservative treatment can be applied. In our study, emphysematous lobes of the patients were resected and any postoperative complication did not observed during the follow up.

In conclusion, CLE is seen very rarely. It may lead to life-threatening respiratory distress in infants. The presented cases were the first in literature as sibling CLE cases that are not twins. Familial predisposition may be taken into account for CLE. Surgical treatment is required to eliminate symptoms, prevent complications, and establish a histopathological diagnosis.

References