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Congenital emphysema as a rare etiology of scoliosis

Skolyozun nadir bir nedeni olarak doğumsal amfizem

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ABSTRACT

Congenital emphysema is characterized by the hyperinflation of related lung field, compression of emphysematous lung space to the adjacent normal lung tissue, and mediastinal shift in neonatals. Scoliosis is a condition in which vertebrae are curved laterally. Although there are many factors causing scoliosis, to our knowledge, scoliosis developing secondary to emphysema was not reported in the literature. In this article, we present a scoliosis case which we believe developed due to congenital pulmonary emphysema.

Keywords: Congenital emphysema; pneumonectomy; scoliosis.

ÖZ

Doğumsal amfizem, yenidoğanlarda ilgili akciğer alanının hiperinflasyonu, amfizematöz akciğer alanının çevredeki normal akciğer dokusuna basısı ve mediastinal kayma ile karakterizedir. Skolyoz ise vertebraların yana doğru eğrildiği bir durumdur. Skolyoza neden olan birçok etken olmasına rağmen, bildiğimiz kadarıyla literatürde amfizeme sekonder gelişen skolyoz bildirilmemiştir. Bu yazıda, doğumsal pulmoner amfizeme bağlı geliştiğini düşündüğümüz bir skolyoz olgusu sunuldu.

Anahtar sözcükler: Doğumsal amfizem; pnömonektomi; skolyoz.

Congenital lobar emphysema is characterized by progressive air trapping usually in one lobe, hyperaeration, compression of the affected lobe on the adjacent lung tissue, and herniation of the affected lobe on the opposing lung. The left upper, right upper, and middle lobes are primarily affected. In the neonatal period, patients usually present with severe and progressive respiratory distress while recurrent lower respiratory infections, wheezing, a chronic cough, and symptoms related to the signs of compression are seen in older children.

CASE REPORT

An 18-year-old female patient came to an orthopedic outpatient clinic with a complaint of back pain that had lasted for two years. She was referred to our department because of the presence of scoliosis and an emphysematous change in the left lung. A physical exam revealed prominent scoliosis and less participation of the left lung in breathing. In addition, chest radiography detected broad hyperaeration along with cystic changes, a deviation of the mediastinum to the right side, and prominent scoliosis in the left hemithorax (Figure 1), and thoracic computed tomography (CT) found diffuse emphysematous lesions, which were similar to air cysts, and variable wall thickness in the left lung. Furthermore, we noted that the mediastinum deviated to the opposite side because the cystic structures were filled with air in the left lung and that scoliosis deformation to the other

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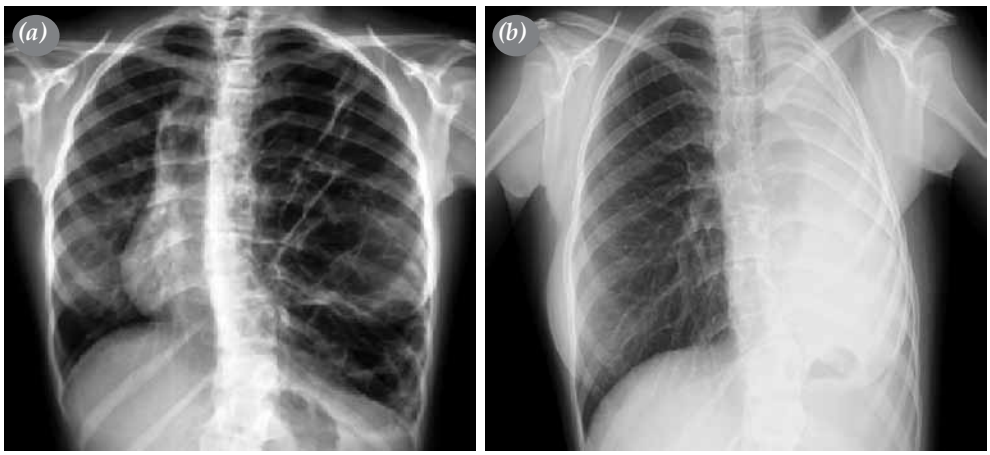

Figure 1. (a) Cystic changes in the left lung and scoliosis were shown in the preoperative posteroanterior chest X-ray **(b)** Postoperative posteroanterior first-year control chest X-ray revealed that mediastinum returned to its normal position.

side in the vertebra was also present (Figure 2). The fiberoptic bronchoscopic examination was normal, and pulmonary ventilation/perfusion scintigraphy detected that ventilation and perfusion were distinctively diminished in the left lung (Figure 3). However, no cardiac or vascular abnormalities were detected on echocardiography, and no positive extra signs, in particular those related to Marfan syndrome, were found on the patient's opthalmology and rheumatology examinations.

A left thoracotomy was then performed, and emphysematous left lung was observed. In fact, the lung had still not deflated even after it was removed via a left pneumonectomy (Figure 4). The pathological analysis revealed congenital emphysema, and the patient was discharged on the seventh postoperative day. On control chest radiography performed at the first postoperative year follow-up, the expansion capacity of the right lung had advanced, the mediastinum returned to its normal position, and the vertebral scoliosis was recovered slightly (Figure 1).

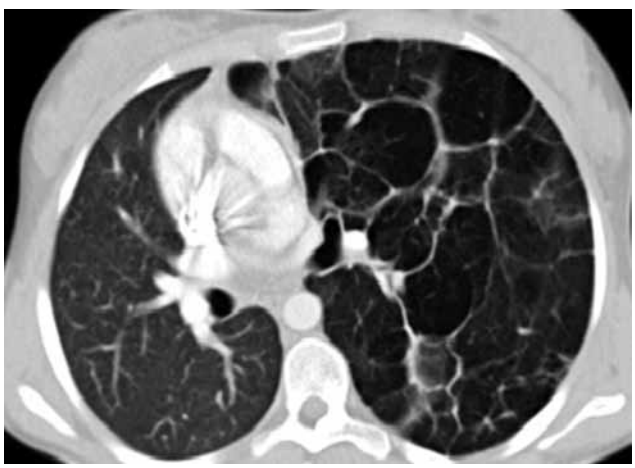

Figure 2. Thoracic computed tomography image revealed cystic changes in the left lung.

DISCUSSION

Agenesis or dysplasia of bronchial cartilage, endobronchial obstruction, intense mucosal proliferation, external compression of the bronchi by aberrant cardiopulmonary vascular structures, and diffuse bronchi abnormalities are among the possible etiological factors associated with congenital lobar emphysema, but nearly half of the causes have not yet been discovered. The patients usually have a large barrel chest due to the internal compression of the lung, and it is possible for the other lung to be compressed in some cases because of the mediastinal shift that formed due to the intensity of the emphysema. Furthermore, Aydin et al.[1] reported that cardiac abnormalities, renal agenesis, renal cysts, pectus excavatum, and diaphragm abnormalities may accompany congenital lobar emphysema. In addition, adolescent scoliosis was present along with the congenital lobar emphysema in our case.

The most appropriate treatment for this type of emphysema is surgical resection of the affected lung parenchyma. In previous studies in which volume reduction surgery was performed, there was less chest wall compliance and flattening of the diaphragmatic dome, improved expiratory airflow, and increased inspiratory capacity in patients with emphysema and bullous lung disease.[2,3] In our case, the inflation of the right lung decreased, the mediastinum returned to

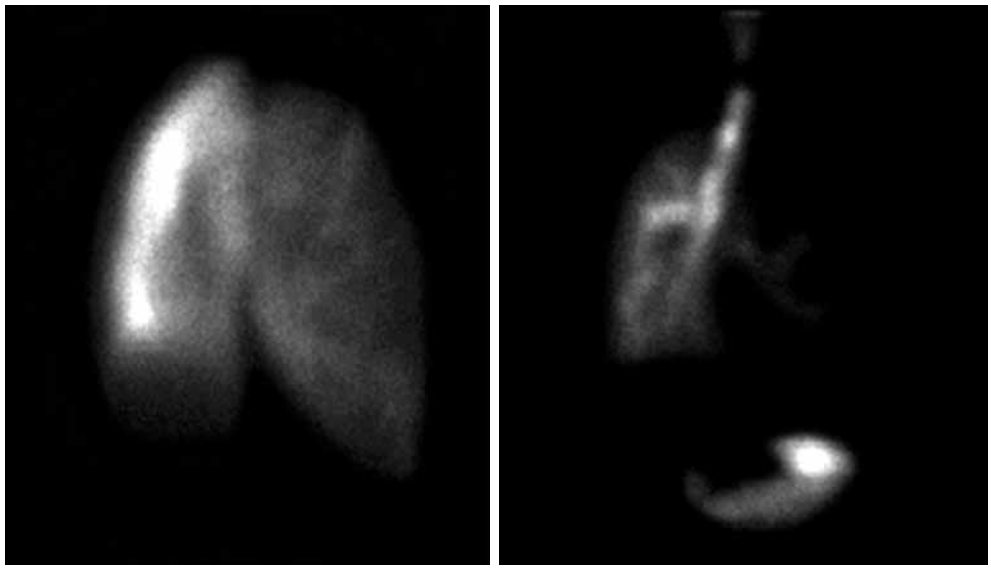

Figure 3. Pulmonary ventilation-perfusion sintigraphy showed ventilation-perfusion mismatch in the left lung.

its normal position, and there was some improvement in the scoliosis when we compared the pre- and postoperative first-year chest radiographies.

Scoliosis is a medical condition in which a person's spine is curved from side to side. It can be classified as either congenital, idiopathic, traumatic, or neuromuscular or be related to mesenchymal disease (Marfan syndrome and Ehler-Danlos syndrome), rheumatological disease, extraspinal contracture (post-empyema, burns, or bone infection), metabolic disease (rickets or osteogenesis imperfecta), the lumbosacral joint (spondilolysis/spondilolysthesis), or a tumor.

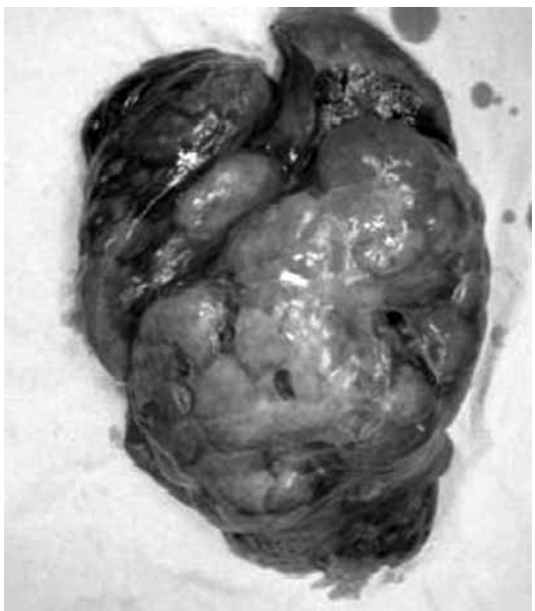

Figure 4. The left lung was still not deflated even after removed via left pneumonectomy.

Idiopathic scoliosis is the most common form, and while its etiology is still unclear, many explanatory theories exist such as muscle anomalies, connective tissue defects, and trauma.[4-6] Idiopathic scoliosis can be divided into three subgroups based on the age when the condition began: infantile (0-3 years old), juvenile (3-10 years old), or adolescent (10 years old-maturity). The restrictive type of pulmonary diseases are observed in the infantile idiopathic scoliosis since the curve occurs earlier, and the impact to the development of the pulmonary parenchyma is greater as well.[7] On the other hand, Koumbourlis[8] asserted that the lung is not affected to a higher degree in adolescent scoliosis since lung maturation has mostly reached its peak by then.

In our case, there were no significant symptoms or scoliosis viewed on sight during the infantile and juvenile periods. However, thoracic scoliosis, the total emphysematous state of the left lung, and shifting of the mediastinal organs to the opposite side were observed on the chest radiography that was performed when the patient consulted with an orthopedist for here back pain during adolescence.

Since compressions and contractures in the body take place, especially in adolescent idiopathic scoliosis,[4-6] we believe that the adolescent scoliosis

in our patient formed because of compression caused by her congenital emphysema. Hence, our findings indicate that emphysema should be included as a part of the broad etiological classification of idiopathic scoliosis.

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