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Congenital Lobar Emphysema

CASE REPORT

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ABSTRACT

Congenital lobar emphysema is a rare developmental abnormality characterized by overinflation of one or more of the pulmonary lobes. This disease presents as respiratory distress because of the ventilation perfusion mismatch as a result of the hyperinflated lung causing compression atelectasis on the ipsilateral or contralateral side with mediastinal shift. Surgical resection of the affected lobes is indicated in both, unless symptoms resolve with conservative therapy. Here, we present a 40-day-old female child requiring right middle lobectomy who presented with respiratory distress caused by congenital lobar emphysema.

Keywords: Congenital lobar emphysema, respiratory distress, lobectomy

INTRODUCTION

Congenital lobar emphysema (CLE) is a congenital disease characterized by overinflation of one or more pulmonary lobes. Its prevalence is between 1/20,000 and 1/30,000, and its incidence varies from 1/70,000 to 1/90,000. While respiratory distress deteriorates rapidly in some infants, it has an insidious onset and slower course in others (1-3). Some patients, however, can remain asymptomatic for years (4, 5).

CLE is most frequently seen in the left upper lobe of the lung (43%–50%), followed by the right middle lobe (32%–35%), right upper lobe (20%), and right lower lobe (less than 1%) (2, 3, 6). In this study, we discuss the case of a patient, in accordance with literature, who presented with the complaints of cough, cyanosis, and discomfort and was diagnosed with CLE and whose condition improved after right middle lobectomy.

CASE REPORT

A 40-day-old girl presented with the complaints of discomfort, cyanosis after cough, and difficulty in breathing lasting for 3 days. From her medical history, it was learned that she had been born at term (birth weight, 4 kg) as a second child from the second pregnancy of a 30-year-old healthy woman through ceasarean section and that she had not had any problem after the delivery. Her parents were not consanguineous, and she had a 3-year-old healthy brother. On physical examination, it was detected that her body temperature was 36.6°C, respiratory rate was 60/min, oxygen saturation was 90%, pulse was 144/min, body weight was 4900 g, and height was 55 cm. On auscultation, lung sounds were decreased in the right side and there were bilateral crepitant rales and intercostal retractions. In laboratory analyses, the level of hemoglobin was 11.6 g/dL, white blood cell count was 11,600/mm³, platelet count was 462,000/mm³, CRP value was 3.45 mg/L, and liver and kidney tests were normal. Posteroanterior chest radiography revealed a remarkable overinflation in the right side and right paracardiac reticular infiltration (Figure 1). The patient was given oxygen with mask and appropriate fluids, and antibiotic therapy was initiated for possible infection. Due to accompanying intermittent expiration length and rhonchi, salbutamol therapy was tried, but improvement was not observed. The results of echocardiography were normal. In the follow-up examinations, intermittent increase and decrease, the absence of fever, normal acute phase reactants, and herniation of the right lung toward the left side in the control chest radiography indicated the coexistence of congenital malformation rather than infection (Figure 2). In contrast-enhanced thoracic computed tomography (CT), there was a significant emphysematous overinflation in the middle lobe of the right lung and accordingly, mediastinal shift to the left side. Moreover, atelectatic changes were observed in the upper lobe posterior segment and lower lobe superior segment of the right lung and in the lower lobe posterobasal segment of the left lung (Figure 3). Histopathological examination of the patient having undergone the right middle lobectomy was consistent with emphysema (Figure 4). After her health status got better, she was discharged from the hospital.

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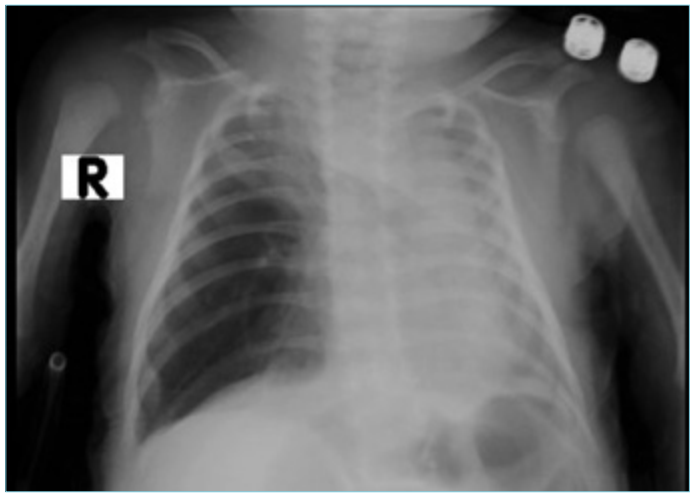

Figure 1. In the first posteroanterior chest radiography of the patient, apparent overinflation is observed in the right side

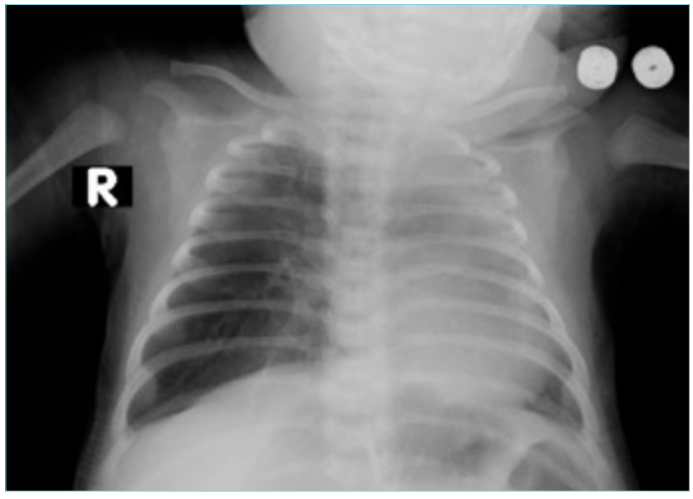

Figure 2. In the posteroanterior chest radiography performed due to the lack of improvement in patient's health status, overinflation in the right side, the shift of the heart and mediastinum toward the left side, and the herniation of the right lung toward the left side in the posterior mediastinum are observed

DISCUSSION

Congenital lobar emphysema is a rarely seen developmental anomaly of the lungs. Its causes cannot be found in 50% of cases. Among the definable causes, the most common one is congenital cartilage defect (25%), whereas the other 25% comprise of mucosal thickening, mucus plugs, abnormal cardiopulmonary structure, and occasionally bronchial obstruction associated with intrathoracic mass (7).

The onset time and severity of clinical findings vary depending on the size of the affected lobe, compression on the surrounding lung tissue, and the weight of mediastinal shift. Symptoms generally appear within the first 6 months (1, 5, 8). In the study of Thakral et al. (2), it was observed that the symptoms began within the first 6 weeks in 17 of 21 patients with the diagnosis of CLE (81%), between 6 weeks and 6 months in 2 patients (9.5%), and after 6 months in 2 patients (9.5%) (2). In our patient, the signs of respiratory stress began within 6 weeks and chest radiography and thoracic CT revealed an apparent overinflation and mediastinal shift in the right side.

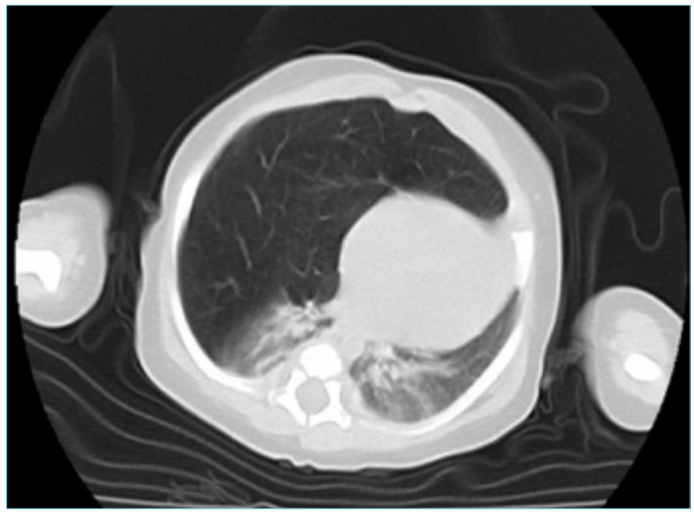

Figure 3. In the thoracic computed tomography, apparent emphysematous overinflation in the middle lobe of the right lung and, accordingly, mediastinal shift toward the left side are observed

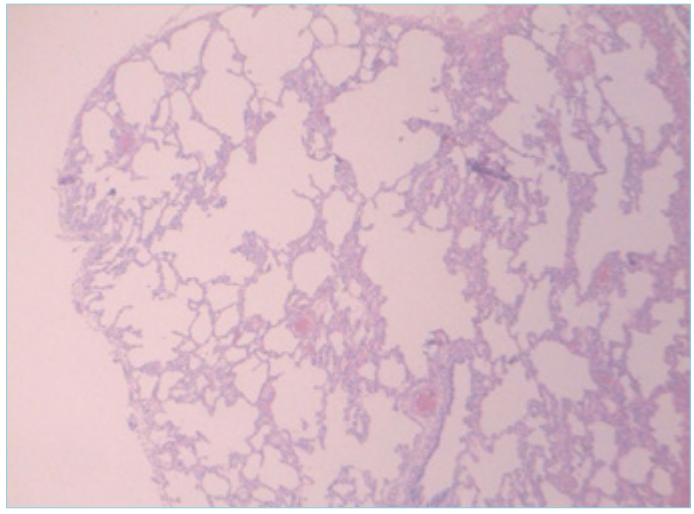

Figure 4. It is observed that the alveolar walls are ruptured and the alveoli are expanded (Hematoxylin eosin ×40)

The most common symptoms include tachypnea, respiratory stress, cough, wheezing, cyanosis, recurrent lung infection, and growth retardation. Similar findings can also be observed in bronchopneumonia, cyanotic congenital heart diseases, other congenital anomalies of the lungs, cystic fibrosis, and aspiration of foreign bodies. Therefore, these disorders should be ruled out (2, 3, 5, 8). Moreover, cardiovascular anomalies coexist with CLE in 14% cases (3). In our patient, the initial diagnosis was bronchopneumonia primarily due to the symptoms and findings and treatment was initiated considering it. However, because of the lack of clinically notable improvement, overinflation of the right lung in the chest radiography, absence of fever, and normal acute phase reactants, infection was ruled out and congenital anomalies were considered.

The result of echocardiography performed for possible cardiac anomaly was evaluated to be normal.

In the diagnosis of congenital lobar emphysema, chest radiography is helpful as well as clinical findings. Overinflation in the affected lobe, mediastinal shift to the contralateral side, and flattening in the ipsilateral diaphragm can be observed in the chest radiography. The findings of the radiography can sometimes be confused with pneumothorax. CT helps to confirm the diagnosis in atypical cases, to eliminate extrinsic mass that can lead to overinflation in the lungs, and to demonstrate the bronchial anatomy before surgery. Furthermore, it is useful for ruling out vascular anomalies and differentiating pneumothorax, pneumatocele, diaphragmatic hernias, or cystic adenomatoid malformation (2, 5, 9, 10). In our patient, CLE was suspected due to the findings of clinical monitoring and radiography, but the final diagnosis was established with CT.

Conservative treatment is recommended for asymptomatic patients or for patients with mild symptoms. In patients having severe respiratory distress, surgical resection of the affected lobe is the treatment approach that is mostly accepted (1, 2, 7, 9). Because our patient had an apparent respiratory distress and she did not benefit from conservative treatment, surgical treatment was performed. Then, her respiratory distress improved; she is still being followed up without any problem.

CONCLUSION

The diagnosis of congenital lobar emphysema cannot be easily made because it imitates many diseases that are frequently seen in children. In such cases, lack of adequate response to the treatment given, reviewing the findings of radiography again, and, above all, suspicion of CLE will help clinicians considerably.

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Authors' Contributions: Conceived and designed the experiments or case: EAA. Performed the experiments or case: EAA, ÜMŞ, ÖM, AK, MK, YAT. Analyzed the data: EAA. Wrote the paper: EAA. All authors read and approved the final manuscript.

Conflict of Interest: No conflict of interest was declared by the authors.

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