CASE REPORT OLGU SUNUMU

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Mixed Type Congenital Pulmonary Airway Malformation: A Rare Case Report

Mix Tip Konjenital Pulmoner Havayolu Malformasyonu: Nadir Görülen Bir Olgu Sunumu

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ABSTRACT Congenital pulmonary airway malformations are rare developmental lung airway malformations that lead to cystic and/or adenomatous pulmonary areas. First described by Stocker in 1897, congenital cystic adenomatoid malformation was renewed in 2001 and named as congenital pulmonary airway malformation. It is divided into 5 types according to its clinical and pathological features. The incidence of this disease, which is rarely seen in the neonatal period, is 1/25,000-1/35,000 pregnancy. Definitive diagnosis is made after pathological examination. We aimed to present a case of mixed type congenital pulmonary airway malformation in which we performed left pneumonectomy at the age of 12.

Keywords: Congenital pulmonary airway malformation; congenital anomaly; lung

ÖZET Konjenital pulmoner havayolu malformasyonu, kistik ve/veya adenomatöz pulmoner alanlara yol açan nadir gelişimsel akciğer hava yolu malformasyonlarıdır. İlk olarak Stocker tarafından 1897 yılında tanımlanan konjenital kistik adenomatoid malformasyon, 2001 yılında yenilenerek, konjenital pulmoner havayolu malformasyonu olarak adlandırılmaktadır. Klinik ve patolojik özelliklerine göre 5 tipe ayrılmıştır. Yenidoğan döneminde oldukça az görülen bu hastalığın görülme sıklığı 1/25.000-1/35.000 gebeliktir. Kesin tanı patolojik inceleme sonrası konur. On iki yaşında sol pnömonektomi yaptığımız mix tip konjenital pulmoner havayolu malformasyonu olgusunu sunmayı amaçladık.

Anahtar Kelimeler: Konjenital pulmoner havayolu malformasyonu; konjenital anomali; akciğer

Congenital cystic adenomatoid malformation, first described by Stocker in 1897, was renewed in 2001 and named as congenital pulmonary airway malformation (CPAM). It is a congenital hamartomatous disease of the lung and is divided into 5 types according to its clinical and pathological features.[1,2] Of these subtypes, only type 3 is adenomatoid. The incidence of this disease, which is seen very rarely in the neonatal period, is 1/25,000-1/35,000 pregnancies.[1,2] Definitive diagnosis is made after pathological examination. We aimed to present a case of mixed type CPAM, which was rarely seen in the literature, in which we had previously had two wedge resections and left pneumonectomy at the age of 12 and whose definitive diagnosis was made by histopathological examination.

CASE REPORT

Our case was being followed up by the pediatric clinic at an external center since birth due to frequent lower respiratory tract infections and severe cough.

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The patient was referred to our clinic after a posterior-anterior chest radiograph (PACR) was taken when his respiratory distress increased when he was 4 years old. In his physical examination, there were decreased breath sounds in the left hemithorax. The patient also had an epilepsy diagnosis. The patient's laboratory tests were hypoxic. Since the PACR taken in our clinic showed severely increased aeration on the left side and the mediastinum shifted to the right, a thoracic computed tomography (CT) was requested. In the thoracic CT, the heart and mediastinum were deviated to the right, a giant bullae with 14x10 cm axial dimension containing septations in the left lung upper lobe, and emphysematous changes and bullae in other parenchymal areas were observed (Figure 1). The left lung lower lobe appeared completely collapsed and air bronchograms were observed inside. There were areas of collapse and consolidation in the paramediastinal areas with wide spread air bronchograms in the right lung. The findings suggested congenital lobar emphysema. Since the patient's definitive diagnosis was not clear and the patient's symptoms were severe, lobectomy was not performed. The case was approached symptomatically and instead of lobectomy, thoracotomy and wedge resection were performed on the bullous areas compressing the normal lung parenchyma. The patient, who achieved bilateral lung expansion in postoperative PACRs, was excised on the 26th day. His pathology was evaluated as "compatible with bullae formation". When the patient was 7 years old, the event developed in a similar manner. When the patient came for the 2nd time, pneumonectomy was required, but since the decision to perform pneumonectomy would be a very big decision for a child patient, a symptomatic approach was again taken and the child was waited to grow up. This situation was explained to the patient's family and the family's approval was obtained. The same procedures were repeated surgically and his pathology was reported as "alveolar destruction areas are observed, these findings do not support congenital lobar emphysema". When the patient was 12 years old and the symptoms continued, a thoracic CT (Figure 2) showed a bullae formation covering a large part of the upper lobe of the left lung, measuring approximately 132 mm in diameter at its largest dimension. There were also large bullae further down. Atelectasis changes were observed in the lung tissue adjacent to the bullae. Increased nodular density was observed in the lower lobe of the left lung. The patient underwent left pneumonectomy after family consent was obtained (Figure 3). The pathology of the pneumonectomy material was reported as "CPAM supported. Although the exact type determination was not given, it was observed to be widespread CPAM type 2, and compatible with CPAM type 1 in focal areas" (Figure 4, Figure 5). The thoracic drain was terminated on the 1st postoperative day. The patient, whose postoperative clinical, laboratory and physical examination findings showed significant im-

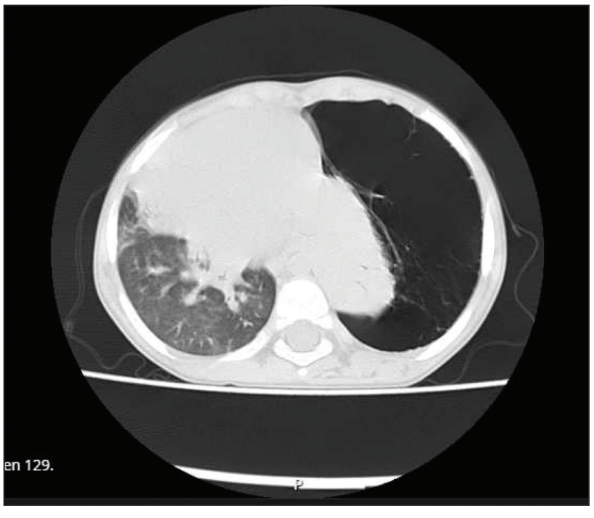

FIGURE 1: Section of the first thoracic tomography scan taken when the case was referred to our clinic.

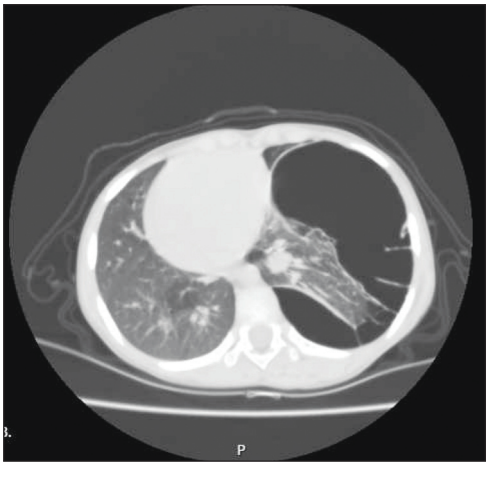

FIGURE 2: Section of the control thorax tomography taken before pneumonectomy.

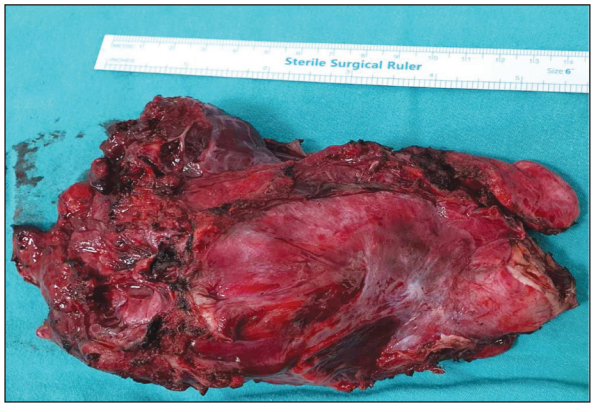

FIGURE 3: Macroscopic view of the left lung pneumonectomy material.

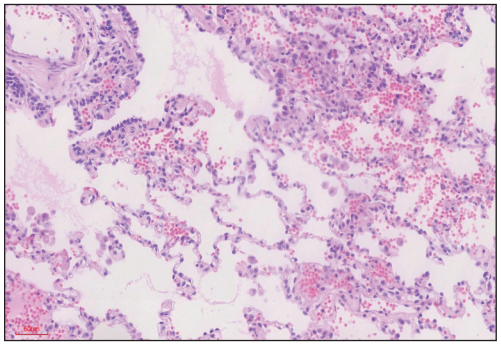

FIGURE 4: Microscopic image of left lung pathology sections compatible with CPAM Type 2.
200x, H&E cysts partially lined with respiratory epithelium. Lung tissue with small cystic structures lined with bronchial epithelium, accompanied by congested vascular structures.

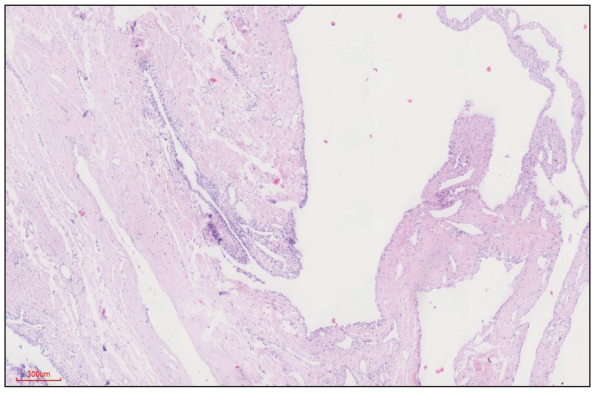

FIGURE 5: Microscopic image of left lung pathology sections compatible with CPAM Type 1.
40x, H&E, cystic alveoli lined by a single layer of flattened epithelium. Lung tissue that has lost its normal appearance with large cystic areas lined by a single layer of cuboidal epithelium.

provement, was discharged with recovery. 'Informed consent' was obtained fort the case report.

DISCUSSION

CPAMs are rare developmental lung airway malformations that lead to cystic and/or adenomatous pulmonary spaces. It is the most common congenital lung disease with an incidence of 1 in 8,300-35,000 live births.[3] They are characterized by an abnormal airway pattern that occurs during the development of lung morphogenesis and may possibly lead to cystic and/or adenomatous pulmonary spaces.[4] They can cause respiratory distress in newborns or remain undiagnosed and asymptomatic for years. The most common presenting cause is reported to be cough and lower respiratory tract infection.[5] CPAM presents clinically with respiratory distress and cyanosis, which begins shortly after birth or later, depending on the size and extent of the cystic lung tissue. Respiratory distress increases in parallel with the expansion of the air-filled cysts. There are no cystic changes in other organs, but it may be associated with lung hypoplasia, diaphragmatic hernia, hemivertebra, Pierre-Robin syndrome, Prune Belly syndrome.[1] Hydrops fetalis is seen in 25% of cases due to cardiac compression and vena cava obstruction due to the compression of cystic lung tissue in the mediastinum in intrauterine life.[1] Treatment varies according to whether the patients are symptomatic or not. Surgical treatment, preferably lobectomy, is applied in symptomatic patients. In our case, since the disease was widespread, pneumonectomy was performed. Since surgical margins could not be clearly distinguished, wedge resection is not recommended. Surgery is performed in the neonatal period in patients with severe respiratory distress. However, in less symptomatic cases, it can be performed electively in older children.[6] Resection in older children is usually performed to prevent recurrent infection and to eliminate concerns about malignancy.[7,8] The most common complication is recurrent lung infection. Although the development of malignancy in previously unnoticed asymptomatic patients seems unlikely, it is a valid reason for resection of the lesions.[7] According to Stocker's criteria, there are overlapping features between CPAM type I and type II, for example, both subtypes may be small (2 cm) and type II may also have mucosal cells. The size of the cysts is not spe-

cific to the CPAM subtype. Therefore, in some cases, the precise subclassification of CPAM is challenging.[9] The definitive diagnosis is made after pathological examination.[10] Although some of these patients develop symptoms at an early stage, some patients may not experience symptoms until adulthood. In conclusion, CPAM is a disease that should be considered in the differential diagnosis of respiratory distress. With early diagnosis and appropriate treatment, it is possible for patients to have trouble-free lives in terms of their respiratory system.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Refik Ülkü, Metin Çelik;* ***Design:*** *Metin Çelik;* ***Control/Supervision:*** *Refik Ülkü;* ***Data Collection and/or Processing:*** *Metin Çelik;* ***Analysis and/or Interpretation:*** *Refik Ülkü, Metin Çelik;* ***Literature Review:*** *Metin Çelik;* ***Writing the Article:*** *Metin Çelik;* ***Critical Review:*** *Refik Ülkü;* ***References and Fundings:*** *Metin Çelik;* ***Materials:*** *Refik Ülkü, Metin Çelik.*

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