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Case Report

Pulmonary hypoplasia with bronchiectasis diagnosed at adult age 22

22 yaşında erişkin yaşta teşhis edilen bronşektazili pulmoner hipoplazi

Fatih Altunyaprak[1]

[1]Ceylanpınar Devlet Hastanesi Göğüs Hastalıkları Kliniği, Ceylanpınar, Şanlıurfa. Türkiye

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Corresponding Author:
Fatih ALTUNYAPRAK, MD.
Ceylanpınar Devlet Hastanesi Göğüs Hastalıkları Kliniği
Ceylanpınar Şanlıurfa/Türkiye;
Orcid id:0000- 0002- 6638 -7944
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Abstract

Unilateral pulmonary hypoplasia can be defined as underdevelopment of the lung and pulmonary artery at different rates. Patients are generally diagnosed in childhood. Rarely, there are cases diagnosed in advanced age, and it manifests itself as case reports in the literature. The complaints of the patients are closely related to the degree of pulmonary hypoplasia. It can be seen as a part of systemic syndromes in some patients. It is important because the patient is diagnosed at an adult age and has accompanying bronchiectasis. It was learned that the patient had dyspnea that increased with exertion since childhood. Cough and sputum were thought to be intermittent and related to concomitant infections. The diagnosis of pulmonary hypoplasia is mostly made with thorax computed tomography (CT) angiography. First of all, diaphragmatic pathologies should be considered in the differential diagnosis. Pulmonary rehabilitation in treatment; treatment of additional diseases and surgical treatment is recommended.

INTRODUCTION

Pulmonary hypoplasia is a rare congenital anomaly characterized by incomplete development of lung tissue. There is impaired gas exchange and respiratory failure due to the decrease in the number of airways and alveoli.(1) It is divided into 3 types with the classification made by Schneider in 1912 and modified by Boyden in 1955 and still accepted today.

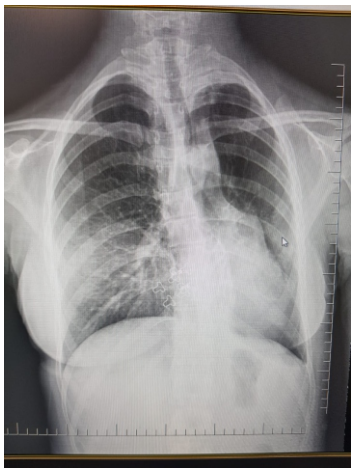
· Type 1 pulmonary agnesia: absence of all bronchi and pulmonary vessels of the pulmonary parenchyma
· Type 2 pulmonary aplasia: complete absence of the pulmonary parenchyma but presence of a rudimentary bronchus on the affected side
· Type3 pulmonary hypoplasia is a varying amount of reduction in lung parenchyma and airways (2)

Although its etiology is not fully understood, it is thought that environmental and genetic factors are effective in the fetal period. The exact incidence is unknown. The incidence of pulmonary hypoplasia in the general population is 1.4 per 1000 and 0.9 to 1.1 per 1000 live births in all births.(3,4) More than 50% of these cases are associated with other congenital anomalies involving the cardiovascular, gastrointestinal, and genitourinary systems and therefore result in early diagnosis in the neonatal period or early infancy.(5) Pulmonary function tests performed in late childhood or adulthood reveal a decrease in diffusing capacity and restrictive or obstructive lung defects.(6) The radiological presentations of lung aplasia and agenesis are similar – compensatory hyperinflation of the contralateral lung and herniation to the affected side are seen with volume loss, ipsilateral mediastinal shift, and elevation of the hemi-diaphragm on the affected side, and it is evaluated as hyperlucency extending along the mediastinum.(7)

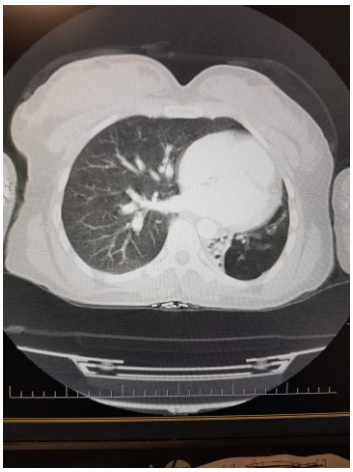
Differential diagnosis and detection of accompanying anomalies are important in patients. Scimitar syndrome, congenital pulmonary airway malformations, bronchopulmonary sequestration, congenital lobar emphysema, persistent pulmonary hypertension are important pathologies.

CASE REPORT

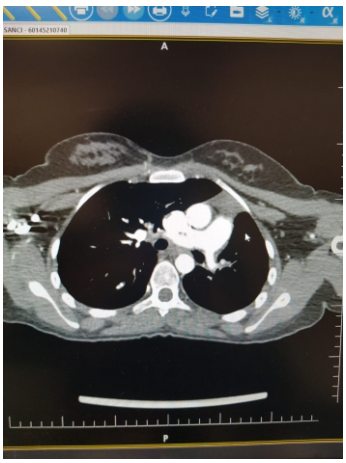
A 22-year-old female patient presented with shortness of breath, cough and sputum complaints. The dyspnea had been present for more than 10 years, increasing with intermittent exertion. Cough and sputum were intermittent. At the time of admission, sputum was yellow, sticky mucoid, up to 10-20 cc per day. On physical examination, respiratory sounds could not be heard in the left hemithorax basals. Thorax ct angio was requested due to increased radiolucency in the left hemithorax in the chest X-ray. Oxygen saturation (SaO2) by pulse oximetry was 96%. In the laboratory, liver function tests lower: 12 U / L, ast: 16 U / L, kidney function tests Blood urea nitrogen: 12 mg / dl, creatinine:0.61mg/dl, electrolytes sodium:138mm/L, potassium 4.7mm/L acute phase reactants; c reactive protein (crp):<2.00 md/L hematological parameters white blood cells(WBC):7,20x109 Hemoglobin: 12.0 g/dl platelet(PLT):285x109 arterial blood gas;
power of hydrogen(ph):7.38 carbon dioxide pressure in blood(paCO2):36mm Hg oxygen pressure in blood(paO2):84 SaO2:98% HCO3:23 mEq/L respiratory function tests forced expiratory 1 second volume (FEV1)/forced vital capacity volume ratio (FVC): %72.
The pulmonary artery and left lung were observed to be hypoplasic in the thorax tomography mediastinum was found to be displaced to the left. The bronchiectatic areas in the left lung were thought to be infective.



Arka-ön akciğer grafisi (görüntü 1) Sol akciğer hipoplazik ve bronşektazik alanlar (görüntü 2)



Toraks tomografisi (görüntü 2)



Toraks tomografisi (görüntü 3)

DISCUSSION

The complaints of the patients show a wide spectrum. Severe shortness of breath, respiratory failure and early death may occur, as well as lung graphs taken during general medical examinations. Severe dsypne, respiratory failure and early death may occur, as well as lung graphs taken during general medical examinations. However, accompanying congenital anomalies is another clinically important issue that needs attention. It is frequently associated with genitourinary, gastroenterological and neurological anomalies. The most common anomalies are the pathologies of the diaphragm, which should be paid attention to. Another expected situation in patients is hypertrophy of the lung on the healthy side, although it is thought as a compensatory mechanism, non-functional emphysematous lung areas, which are frequently encountered in the lung on the healthy side during this hypertrophy, aggravate the chart. Wong et al.(8) used different combinations of FGF10, FGF7 and BMP4 molecules to promote proximal lung differentiation. FGF10 is a critical growth factor expressed by the mesenchyme in early

lung development and is closely associated with lung development and organogenesis.FGF7, also derived from mesenchyme, not only promotes epithelial growth but also stimulates fluid formation by the lung. Human amniotic epithelial cells reduced lung injury and expressed lung epithelial markers in both in vitro and in vivo studies.(9) Studies have shown that pluripotent steamcell, which can be used in interstitial lung diseases and chronic obstructive lung diseases, can be used in pulmonary agnesic diseases.The main compelling area in the Steam cell draws attention as the difficulties experienced in side acquisition and storage.

REFERENCES

1- Laudy JA, Wladimiroff JW. The fetal lung. 2: Pulmonary hypoplasia. Ultrasound Obstet Gynecol. 2000 Oct;16(5):482-94
2- Boyden ea. Developmental anomalies of the lungs. Am J Surg. 1955 Jan;89(1):79-89
3- Schittny JC. Development of the lung. Cell Tissue Res. 2017 Mar;367(3):427-444
4- Porter HJ. Pulmonary hypoplasia. Arch Dis Child Fetal Neonatal Ed. 1999 Sep;81(2):F81-3.
5- Lee E.Y., Boiselle P.M., Cleveland R.H.: Multidetector CT evaluation of congenital lung anomalies. Radiology 2008; 247: pp. 632-648.
6- Bojanić K, Grizelj R, Dilber D, Šarić D, Vuković J, Pianosi PT, Driscoll DJ, Weingarten TN, Pritišanac E, Schroeder DR, Sprung J. Cardiopulmonary exercise performance is reduced in congenital diaphragmatic hernia survivors. Pediatr Pulmonol. 2016 Dec;51(12):1320-1329
7- Abbey P., Narula M.K., Anand R.: Congenital malformations and developmental anomalies of the lung. Curr Radiol Rep 2014; 2: pp. 71
8- Wong AP, Bear CE, Chin S . Directed differentiation of human pluripotent stem cells into mature airway epithelia expressing functional CFTR protein. Nat. Biotechnol. 2012; 30: 876– 882.
9- Moodley Y, Ilancheran S, Samuel C et al. Human amnion epithelial cell transplantation abrogates lung fibrosis and augments repair. Am. J. Respir. Crit. Care Med. 2010; 182: 643– 651.

Özet

Tek taraflı pulmoner hipoplazi akciğerin ve pulmoner arterin değişik oranlarda az gelişmesi olarak tanımlanabilir. Hastalara genel olarak çocukluk yaşında tanı konulmaktadır. Daha nadir olarak ileri yaşta da tanı konan olgular mevcut olup literatürde vaka sunumları şeklinde kendini göstermektedir. Hastaların şikâyetleri pulmoner hipoplazinin derecesi ile yakından ilişkilidir. Bazı hastalarda sistemik sendromların bir parçası olarak görülebilmektedir. Hastaya erişkin yaşta tanı konması ve eşlik eden bronşektazi olması nedeniyle önem arz etmektedir. Hastanın çocukluk çağından itibaren eforla artan nefes darlığı olduğu öğrenildi.Öksürük ve balgam aralıklı olduğu ve eşlik eden enfeksiyonlarla ilgili olduğu düşünüldü.Pulmoner hipoplazi tansı çoğunlukla toraks bilgisayarlı tomografi (CT) anjiyo ile konulmaktadır. Öncelikle ayırıcı tanısında diyaframa patolojileri düşünülmelidir. Tedavide pulmoner rehabilitasyon; ek hastalıkların tedavisi ve cerrahi tedavi önerilir.

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Address
Recep Tayyip Erdoğan Üniversitesi, Tıp Fakültesi, İslampaşa, 53020, Rize Türkiye
Faculty of Medicine, Recep Tayyip Erdogan University, Islampasa, 53020, Türkiye
Phone: (+90) 212 30 09, E-mail: rizetipdergi@erdogan.edu.tr
Web site: https://dergipark.org.tr/tr/pub/rizetip