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Case Report

Unilateral pulmonary hypoplasia*

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ABSTRACT

Pulmonary hypoplasia represents a broad range of malformations characterized by incomplete development of lung tissue. The severity of the lesion depends on the appearance time of the malformation during the timeline of lung development, and the presence of further anatomic anomalies. In this report, we present a case of pulmonary hypoplasia in a 27-year old woman. © **IJAV. 2008; 1: 23–25.**

Key words [lung] [congenital lesions] [anatomy] [computed tomography]

Introduction

The developmental anomalies of the lung at the 4th and 24th gestational weeks may cause bronchopulmonary foregut abnormalities. Most patients described in literature were newborns and infants [1]. Absence of a lung is a rare congenital anomaly that the exact cause is not completely known. However, it is usually associated with other anomalies [2]. The incidence of unilateral pulmonary agenesis has been estimated approximately one in 15,000 live births [3], or between 0.0034% and 0.0097% [4]. In 70% of cases, the left lung is absent [5]. Both genders are affected almost equally [2]. Boyden, in 1955, has classified the degree of lung involvement into three groups as pulmonary agenesis, pulmonary aplasia and pulmonary hypoplasia [1]. Here, we describe a case of left lung hypoplasia and anomalous formation of right lung lobes without any associated anomalies.

Case Report

A 27 year-old woman who complained coughing and dyspnea was admitted to hospital. No breath sound was heard at her lower left hemithorax. Chest radiogram revealed mediastinal shift to the left. Thus, computed tomography (CT) scan was performed. On CT scan, the heart was displaced posteriorly (Figure 1); complete opacification of left lower hemithorax visualized with shifting of mediastinum and trachea to the left; the left half of the diaphragm was also elevated (Figure 2). CT scan revealed reduction in volume of the left lower hemithorax; neither airways nor pulmonary tissue were present there. The left main bronchus was traced approximately for 3 cm and it had no branches (Figure 3). There was shifted pulmonary tissue in the left upper hemithorax with two bronchi; those were belonging to the upper anterior segmental bronchus and middle lobe's bronchus of the right lung (Figure 3). The hypoplastic left pulmonary tissue was present in the left hemithorax,

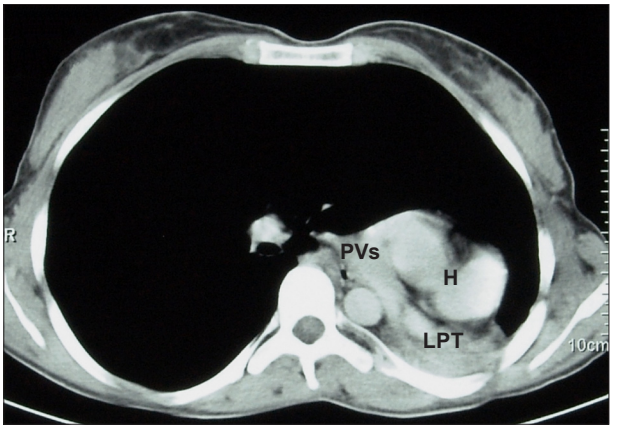

Figure 1. The heart is displaced posteriorly. The hypoplastic left pulmonary tissue is in the left hemithorax, posterior to the heart; no communicating bronchial tree is present. The left pulmonary artery and veins are thinner than normal. *(**H:** heart, **LPT:** hypoplastic left pulmonary tissue, **PVs:** pulmonary vessels).*

* This case was presented as a poster presentation in the 4th Asian-Pacific International Congress of Anatomists, APICA (2005, Kusadasi-Turkey).

posterior to the heart; it had no communicating bronchial tree. The left pulmonary artery and veins were thinner than normal (Figure 1). Rest of the examination was unremarkable.

Discussion

For a normal lung development, physical space in the fetal thorax must be adequate, and amniotic fluid must be brought into the lung by fetal breathing movements, leading to distension of the developing lung. Several factors affect the volume and composition of the amniotic fluid, including the volume and pressure, the composition of lung fluid, the role of the kidney in lung growth [6].

According the etiologic factors, pulmonary hypoplasia is classified as primary and secondary hypoplasia. In primary hypoplasia there is not an obvious cause of hypoplasia. Whereas in secondary hypoplasia, there is variable fetal and maternal abnormalities in 60% of cases; other causes are space-occupying lesions in the chest, developmental anomalies of the chest wall, and urogenital and neuromuscular diseases [1]. The

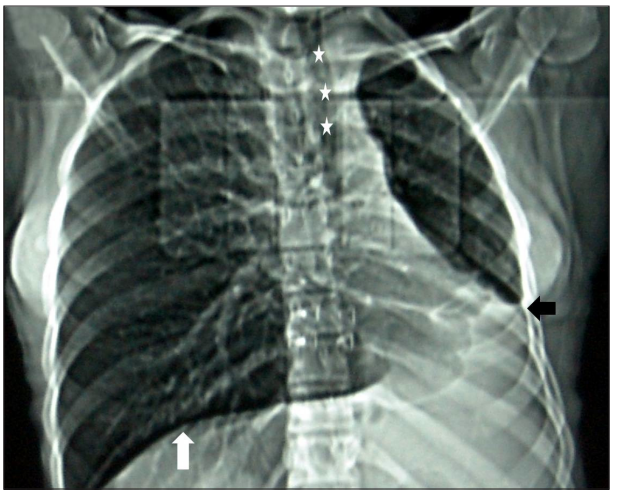

Figure 2. Reduction in volume of left lower hemithorax, complete opacification of left lower hemithorax with shifting of mediastinum and trachea to the left and elevated left half of diaphragm are shown. *(**Stars:** trachea, **white arrow:** right side of diaphragm, **black arrow:** elevated left half of diaphragm)*

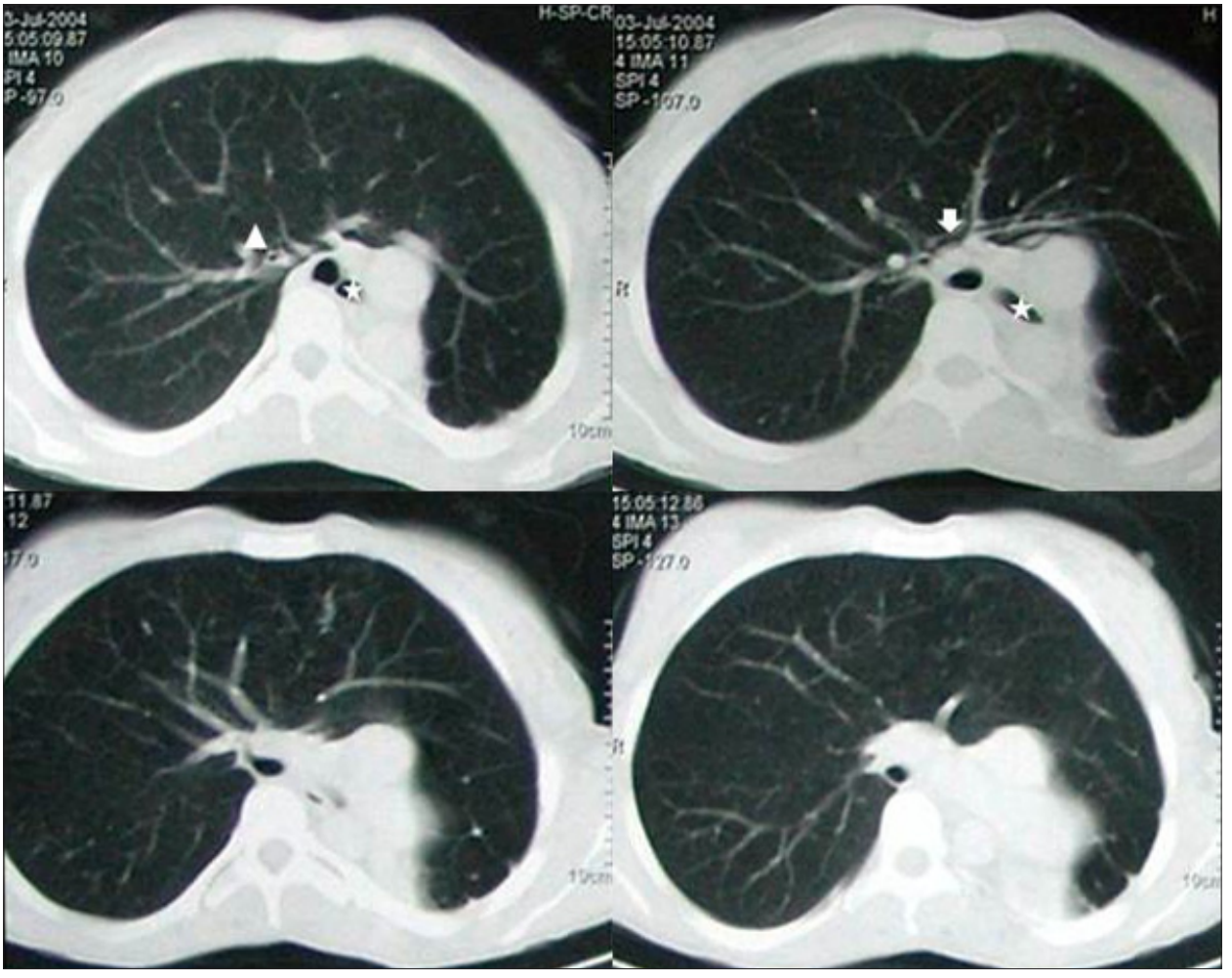

Figure 3. Reduction in volume of left lower hemithorax without any airways and pulmonary tissue. The left main bronchus is traced approximately 3 cm without any branches. Shifted pulmonary tissue in the left upper hemithorax has two bronchi: the right upper anterior segmental bronchus and the middle lobe's bronchus of the right lung are seen. *(**Star:** left main bronchus, **arrow:** middle lobe's bronchus of the right lung, **arrowhead:** the right upper anterior segmental bronchus).*

accompanying congenital anomalies include cardiac lesions, bronchogenic cysts, diaphragmatic hernias and skeletal anomalies (commonly vertebral or rib anomalies) [3]. However, patients with pulmonary agenesis may remain asymptomatic and the diagnosis may not be made until adulthood [7].

Several authors have categorized their cases according to the classification of Boyden in 1955. In group 1 or pulmonary agenesis; the entire lung, bronchus and its pulmonary vessels are absent. In group 2 or pulmonary aplasia, the lung and pulmonary artery are absent, but there is a rudimentary bronchus coming off the trachea. In group 3 or pulmonary hypoplasia, there is hypoplastic bronchus with hypoplastic lung parenchyma [2]. There is hypoplastic lung tissue only in group 3. In our case, there is hypoplastic left lung tissue, very thin pulmonary vessels and rudimentary left main bronchus. Consequently, our case can be classified as group 3 according to Boyden's criteria.

The difference between pulmonary agenesis and pulmonary hypoplasia needs to be distinguished. There is no main bronchus, pulmonary vessels and lung tissue in complete absence of lung. However, in pulmonary hypoplasia, there are underdeveloped lung parenchyma, pulmonary vessels and rudimentary bronchus.

There are many ways to diagnose hypoplasia of the lung: bronchoscopy, bronchography, CT, magnetic resonance imaging, pulmonary angiography, etc. Thomas et al. suggested that CT is more useful than bronchography in assessing the hypoplastic lung, especially for the purposes of its surgical removal [8]. In our case, left lung hypogenesis with displacing right upper anterior segment and middle lobe of the right lung to the left hemithorax is diagnosed via CT.

Agenesis of lung or its lobes must be considered in the differential diagnosis in the case of an opaque hemithorax. It is important to investigate the coexistence of this anomaly. Asymptomatic cases do not require any treatment if there are no additional anomalies, but these cases carry high-risk in any surgery because of low respiratory reserve.

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