Pulmonary Artery Agenesis and Lung Hypoplasia

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ABSTRACT

Unilateral pulmonary artery agenesis with lung hypoplasia is very rare and only a few cases have been reported in the literature. Some patients with hypoplasia may suffer from recurrent hemoptysis dyspnea, and lung infections. Other patients are asymptomatic. Chest X-Ray reveal a reduction in the volume of hemithorax, an elevation in the diaphragm, reduced intercostals spaces and mediastinal shift in the affected side. We present a 15 year old male patient who complained of shortness of breath and chest pain.

Key Words: Pulmonary artery agenesis, lung hypoplasia.

INTRODUCTION

Unilateral pulmonary artery agenesis with lung hypoplasia, one of the developmental anomalies of the lung, is not very common. It is very rare with the agenesis of pulmonary artery of the same side. While some of the congenital lung lesions are diagnosed in neonates or in later periods of the life, others are asymptomatic and may be diagnosed during the childhood or adult periods accidentally (1,2).

In this report, we describe a 15-year-old male patient who presented with complaints of dyspnea and a left chest pain. Detailed examinations revealed agenesis of left pulmonary artery and left lung hypoplasia.

CASE

A 15-year-old male patient was presented with exertional dyspnea and chest pain in our clinic for further investigations. The patient stated that he had been suffering from these complaints for 4-5 years, with increased chest pain during recent months. Physical examinations revealed a decreased left hemithorax volume, with

accompanying decreased respiratory sounds upon auscultation. Whole blood count and biochemistry parameters were within normal levels. Spirometric values were compatible with restrictive lung disase; FVC: 67% pred, FEV_1: 67% pred, FEV_1/FVC: 87% pred. Arterial blood gas tensions(FiO_2 0.21) at rest was mild hypoxemia (PaO_2, 76 mmHg) and hypercapnea (PCO_2, 45 mmHg) Chest X-Ray indicated an elevated left diaphragm, a mediastinal shift to the left hemithorax and ventilation in the left superior zone (Figure 1). A computed tomoghraphy scan revealed that the volume of the left hemithorax was reduced, hypoplastic left lung and bronchial tree and the mediastinal structures displaced towards the left. There was an emphysema in the rigth lung while the superior lobe of the right lung was partially displaced towards the left hemithorax. The left lung and the bronchial tree were hypoplastic (Figure 2). Magnetic resonance imaging (MRI) examination indicated that heart and mediastinal structures were displaced to the left hemithorax.

A nuclear ventilation-perfusion scan showed that the right lung was normal, the left hemithorax was slightly perfusion connected with the right lung while the left lung did not present any perfusion. The volume of the left lung was greatly reduced, indicating a left lung hypoplasia. Pulmonary angiography performed with MRI angiography examination revealed that left pulmonary artery agenesis-hypoplasia (Figure 3). The patient, with the above findings, has been under follow-up with a diagnosis of left pulmonary artery agenesis and left lung hypoplasia, without considering a surgical intervention.

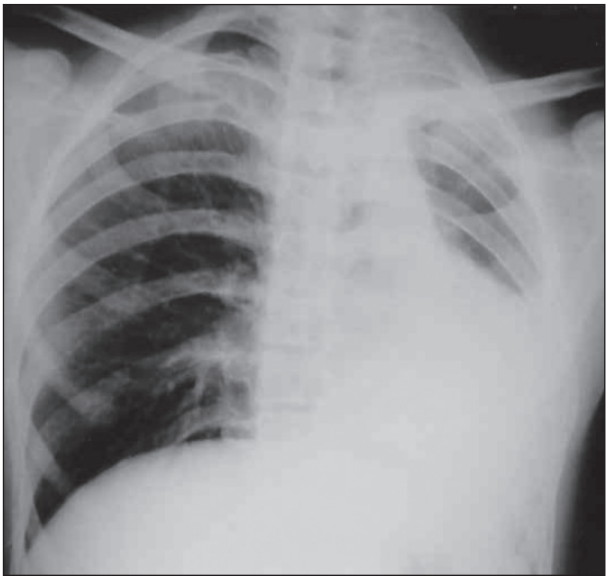

Figure 1. Chest X Ray revealed a significant air reduction of left hemithorax.

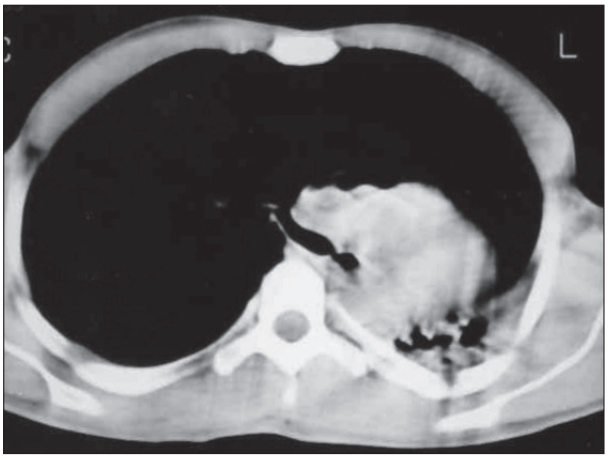

Figure 2. Computed tomography showed hypoplasic left lung.

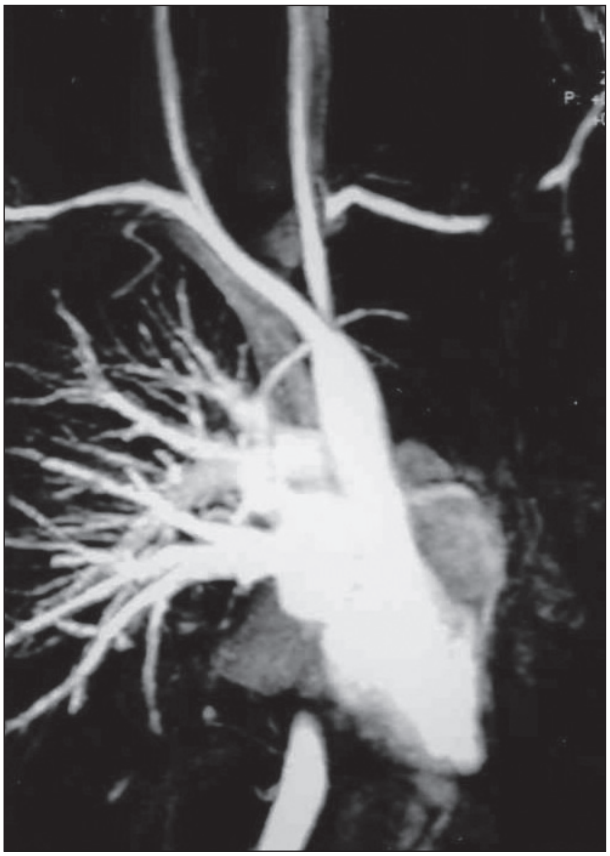

Figure 3. Pulmonary angiogram revealed left pulmonary artery agenesia.

DISCUSSION

Unilateral pulmonary artery agenesis is a rare condition often associated with other congenital abnormalities. In some cases, they occur together with anomalies, mostly with skeletal, heart and diaphragm anomalies. Pulmonary agenesis was first defined by De Pozzi in 1673 (3,4). Chromosomal origin, vitamin A deficiency, intrauterine infections, environmental factors have been held responsible for the etiology of congenital lung malformations. Pulmonary hypoplasia, however, is frequently associated with situations causing fetal lung compressions. These compressions may be either intrathoracic or extrathroacic. Intrathoracic compressions may be caused by diaphragmatic defects, excess pleural fluid, intrathoracic tumors or cysts while oligohidroamnios or chronic elevation of hemidiaphragm may be held responsible from extrathroacic compressions (5). Thus, the lungs become smaller at the 16th gestational week due to decreased branching of air passage pattern.

Pulmonary hypoplasia is divided into two groups as primary and secondary. A developmental lung anomaly is involved in primary type, and the cases die by respiratory distress at birth or soon after the birth. Diagnosis can be made only with a post mortem autopsy. In secondary hypoplasia, the bronchi and the lobes of the lung are rudimentary, and therefore they tend to be asymptomatic clinically. However, during the neonatal period, they may be lost due to mostly skeletal, heart and diaphragm anomalies (1,2,4).

Survivors with hypoplasia may suffer from lung infections and hemoptisia attacks, or they may be diagnosed during routine examinations performed for a non-specific symptom, as in our case.

Chest X-ray of agenesis hypoplasia complex reveal a reduction in the volume of hemithorax, an elevation in the diaphragm, reduced intercostal spaces and mediastinal shift in the affected side. This appearance in our case is very typical, which resembles atelactasis (4-6). It may mimic chronic pleural diseases. Contrast enhanced computed tomography (CT) is almost definitive for the diagnosis. Presence of bronchi and pulmonary artery is confirmed with CT. Pulmonary artery is normal in most hypoplasia cases, which can be confirmed with MRI angiogram and ventilation perfusion scan. But in our case, the MRI angiogram shows very clearly that pulmonary artery has poorly developed, while lung ventilation-perfusion scan reveals absence of perfusion in the left lung. This is a very rare and interesting case. Bronchography is not commonly used in the diagnosis. Therefore, we did not use bronchography. Since our case did not present any skeletal, heart and diaphragm anomalies, it was not necessary to attempt other diagnostic methods. Clinical situations such as trombotic embolus of pulmonary artery, extrinsic obstruction of the artery and anomalous venous drainage of the right lung into the inferior vena cava (the Scimitar syndrome)should be considered in the differentiating diagnosis.

In pulmonary agenesis-hypoplasia complexes, surgical treatment can only be attempted in accompanying cardiac and vascular anomalies or in recurrent hemoptisia attacks, or in persistent lung infections and bronchiectasia. Apart from this, follow-up and symptomatic treatment should be the basic approach. Survival is probable until maximum 6th decade. We therefore did not consider surgery during intervention, and have been keeping the patient under follow-up for 3 years.

In conclusion, we have concluded that the cause of the lung hypoplasia in our case is secondary hypoplasia, attributed to the pulmonary artery hypoplasia. Our view is supported by the literature indicating that reduction in pulmonary blood flow prevents the development of lung and causes lung hypoplasia (8,9).

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