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Case report

Case of missing left lung

Ali Cheraghvandi, Saeid Fallah Tafti*, Firouzeh Talischi

Pulmonary Health Care and Nursing Research Center, National Research Institute of Tuberculosis and Lung Diseases, Shahid Beheshti University of Medical Sciences, Masih Daneshvari Hospital, DarAbad, Tehran 1955841452, Iran

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ABSTRACT

Here we present a young female with missing left lung and history of upper respiratory infections in childhood. The lungs have ability to grow and regenerate in children. She has had no major complications into adulthood. It is important to know diagnosis in recurrent pulmonary infections and here bronchoscopy was diagnostic.

1. About agenesis of the lung

Agenesis of the lung is a developmental defect that is rare. In this condition, one or both lungs are either completely absent or hypoplastic. This condition represents a spectrum of congenital anomalies in lung development. The prevelance of this condition has been noted to be 0.0034–0.0097%. There appears to be no sexual predilection for this condition. Most cases present in the neonatal period with cyanosis, tachypnea, dyspnea, stridor or feeding difficulties. The condition is often associated with fetal distress at birth.[1] Yet, it may also be asymptomatic and manifest itself in adulthood. A case was diagnosed at necropsy in a 72-year-old. Patients often have some pulmonary manifestations like cyanosis or respiratory difficulty. Left-sided agenesis (70% of cases) is more frequent than right-sided. Right-sided defects have a poorer prognosis due to often coexisting cardiac anomalies or greater mediastinal shift and pressure on other structures.[2]

Pulmonary agenesis is anatomically devided into three groups. First are patients who have absence of the entire lung and its pulmonary artery.

Coexistence of cardiac anomalies are consistent with embryologic developmental insult in the fourth week of life. Parental consanguinity and autosomal recessive pattern of inheritance has been noted in some cases. Although extrinsic insults such as drugs, infection during pregnancy, environmental substances and mechanical factors in the uterus or congenital small thoracic cage may also be causative factors.[3]

2. Presentation of case

The patient is a 23-year-old female without a significant past medical history except recurrent childhood upper respiratory infections, born in Tehran, who presents with a two-week history of a cold. After a week of cold symptoms, she visited her primary care physician who recommended to take a chest X-ray and started her on cefexime and salbutamol syrup. Her symptoms began one month prior to her presentation to a pulmonologist with cough, small amount of white sputum and a sore throat. The patient noted coughing up less than a teaspoon of phlegm on a given day during her cold. She was told that she has influenza and it had involved family members as well. She had some slight fevers and chills but did not measure her temperature. She had recurrent URI's as a child. Compared to people with her own age, she has less tolerance for physical activity. She had received all her vaccinations. Other than the cold medications, she is not regularly on any medications and has no drug allergies. She denies using any tobacco, or drugs of abuse. The patient is a housewife of eight years. She has an eight year education level which was disrupted due to her marriage. The patient denies family history of any medical illnesses and has two healthy children. On physical exam, the oral temperature was 37.2 °C, respiratory rate of 20, heart rate of 95, blood pressure of 130/80 and pulse oximetry of 98%. She was comfortable and at rest and alert and oriented to time and place. She had no surgical scars. The patient had assymetrical thorax with mild scoliosis with shift to the left. Cardiac exam showed heart sounds S1 and S2 best heard at the left anterior axillary line with no murmurs, rubs or gallops. The lung exam showed hyperresonant vesicular sounds on the right side. Abdomen was soft and

* Corresponding author. Tel.: +98 96 2126105050.
E-mail address: saeid_ftafti@yahoo.com (S.F. Tafti).

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nontender, extremities showed no clubbing, cyanosis, edema or anomalies. Neuro exam was normal.

On further questioning about her childhood medical history, she noted having had a chest X-ray when she was six years old. She was told by one physician that she might have cardiac or mediastinal shift but she did not investigate it further. The patient's mother was with her at the pulmonary clinic and denied consanguinity with her husband or taking any pills during her pregnancy and was 25 when she had her.

The patient's chest X-ray showed that she has mediastinal, and cardiac shadow displacement to the left side of the thorax. Collapse of the left lower lobe was considered.

A chest CT-scan with IV contrast was done for the patient which showed significant mediastinal shift toward the left side accompanied by compensatory hyperaeration in the right pulmonary parenchyma and total collapse of the left pulmonary parenchyma. The left main pulmonary artery was not present with normal pattern of the remaining bronchovasculature. The rest of the bronchovascular patterns of both lungs were normal.

3. Clinical course

On bronchoscopy the patient had agenesis of the left lung.

Spirometry and whole body plethysmography were done. The patient also had a cardiology consult to rule out any vascular, cardiac anomalies or effect of the agenesis on cardiac function. Transesophageal echo was normal and there was no dextrocardia on EKG (Table 1).

Table 1
Patient's laboratory values.

Labs	Patient's values	Normal range for adult
FBS	89 mg/dl	70–115
Urea	16 mg/dl	15–45
Creat	0.6 mg/dl	0.4–1.5
Uric acid	4.1 mg/dl	Male: 3.6–8.2 Female: 2.3–6.1
Bilirubin total	0.7 mg/dl	0.5–1.5
Bilirubin direct	0.1 mg/dl	Less than 0.5
SGOT	20 U/L	Male<37 Female<31
SGPT	23 U/L	Male<41 Female<31
ALP	152 U/L	Male upto 270 Female upto 240 Child 180–1200
Urinalysis	Yellow, cloudy, s.g. 1.037 Negative *Microscopic* WBC/HPF 4–6 RBC/HPF 1–2 Ep. cells/HPF 6–8 Mucus many	
WBC	6.4 × 1000/micl	3.5–11
RBC	4.88 mil/cu mm	3.3–5.8
Hgb	14.0 g/dl	11.5–16.5
Hct	42.4%	34–50
MCV	86.8 fl	78–96
MCH	28.7 pg	26–35
MCHC	33.1%	31–36
Plt	321 × 1000/cu mm	135–470
RDW	13.6%	11–14.5
Differential count	2	
Neut	55%	
Lymph	40	
Mono	3	
Eos	2	
ESR/1 h	19 mm/1 h	

4. Discussion

Differential diagnosis for the X-ray findings include total atelectasis from any cause, bronchiectasis with collapse and advanced fibrothorax which can be distinguished with the CT.[4]

Other conditions to consider in the differential include hyperlucent and hypoplastic lung syndromes, obstructive lung lesions mainly cancer, diaphragmatic hernia, adenomatoid cystic malformations and sequestrations and the Scimitar syndrome (which involves anomalous venous drainage of the right lung into the inferior vena cava associated with other vascular and cardiac anomalies).[3,5]

The lungs have ability to grow and regenerate in children. In cases of severe hypoplasia or dysplasia, lung regeneration can lead to survival in 40 percent of cases.[6]

Surgical management is best guided by pulmonary and left ventricular or aortic angiography. Indication for surgery is a hypoplastic lung prone to atelectasis and infection.[1]

Many patients due to coexistent anomalies are surgical candidates and preplanning for the intubation of the patients in the ICU or operation room can be done.[7] The intubation of the patients can cause prolonged atelectasis of the lung. Preplanning for correct intubation or avoiding it can be considered.

5. Future considerations

The organogenesis of the lung is influenced by genetic and epigenetic factors such as growth factors (e.g. EGF has stimulatory and TGF-β has inhibitory effect). Future development of gene therapy is the goal trying to prevent lung injury and promote lung repair.[6]

Furthermore lung organogenesis can be influenced by environmental factors in positive and negative ways. For example, hyperoxia occurring in treated premature infants adversely affects lung development and must be avoided if possible.[6]

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