A Rare Clinical Presentation of Primary Pulmonary Hypoplasia with Tuberculous Pleural Effusion

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Primary pulmonary hypoplasia is rare in adulthood. It is characterized by decreased number or size of bronchi, vessels and alveoli. We present a case of unilateral pulmonary hypoplasia in a 44-old-year male smoker who presented with right pleural effusion. His chest X-ray revealed an inhomogenous opacity on the left side with bronchiectatic changes and right minimal pleural effusion. Fiberoptic bronchoscopy revealed blind end bronchi in left upper lobe and computed tomography pulmonary angiography revealed hypoplastic lung with cystic bronchiectasis on the left side and hypoplastic left pulmonary artery. It was not associated with any other congenital anomalies. In addition to symptomatic management, he was started on anti-tuberculous treatment for tuberculous pleural effusion and kept under follow-up.

Keywords: Computed tomography pulmonary angiography, Congenital anamoly, Cystic bronchiectasis, Primary pulmonary hypoplasia

INTRODUCTION

Congenital malformation of the lung is extremely rare. Unilateral pulmonary hypoplasia is a rare congenital anomaly. Although its exact prevalence is not known, it is estimated to be 1-2/12,000 or 15,000 births. It is characterized by reduced number or size of the airways, vessels and alveoli; however gross morphology of the lung is mostly unremarkable.¹ Hypoplastic region of the lung becomes small, fibrotic and un functional with or without bronchiectatic changes. Other congenital anomalies may be seen especially in the cardiovascular system, gastrointestinal system, urinary and musculo skeletal system. Depending on the severity of hypoplasia, the patient may either be asymptomatic or present with severe respiratory distress. It is usually diagnosed in the neonatal period, infancy or childhood but rare in adulthood. It can be primary (idiopathic) or secondary associated with other congenital anomalies that are implicated in its pathogenesis.² Due to its rarity we hereby describe a case of middle aged male who was diagnosed as primary pulmonary hypoplasia with tuberculous effusion.

CASE REPORT

A 44-year-old male patient was admitted in our hospital with complaints of cough and expectoration and shortness of breath for 1 month and right sided chest pain for 20 days. Cough initially started as dry cough and progressed to wet cough with a mucoid expectoration. There was a low-grade fever for 10 days and there was no history of hemoptysis. Shortness of breath was initially exertional, but gradually progressed to grade. Medical and family history was unremarkable, and he was completely asymptomatic until his admission into hospital. There was no history of consanguineous marriage.

General examination was unremarkable, and vitals were stable. Examination of the respiratory system revealed asymmetric chest with flatness on the left side of the chest, narrowing of the intercostals spaces and decreased spinoccipital distance on the left side. Movements were decreased on the left side. Both upper and lower mediastinum were shifted to the left side. Apex beat was felt in the posterior axillary line on the left side. Percussion note was dull over left hemothorax and on auscultation bronchial quality breath sounds heard in upper and lower interscapular area on the left side, decreased breath sounds in right infra scapular area; coarse crackles were heard in upper and lower inter scapular areas on the left side.

Investigations

Routine hematological and biochemical tests were within the normal limits. Sputum smear microscopy for acid-fast bacilli (AFB) was negative for 2 consecutive days. Sputum culture showed growth of staph aureus that was sensitive to ciprofloxacin and clindamycin.

Chest X-ray (CXR) posterioranterior view: Revealed left inhomogeneous opacity with bronchiectatic changes and...
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Contrast-enhanced computed tomography (CT) showed features suggestive of volume loss in left hemi thorax; multiple thin and thick walled intercommunicating cystic cavities were seen in left lung parenchyma, which was connected to lobar bronchi with no identifiable normal lung parenchyma (Figure 3). Left main bronchus appeared normal in caliber; lobar bronchi appeared reduced in caliber. Hyperinflation of the right lung with herniation into left hemi thorax anteriorly was seen. CT pulmonary angiogram with three-dimensional (3D) volume rendering technique showed that right main pulmonary artery size was 23 mm, normal in caliber and enhancement pattern. Left main pulmonary artery was 15 mm at the level of bifurcation of main pulmonary artery smaller in caliber than that of the right side (Figure 4). The findings were suggestive of left lung hypoplasia/dysplasia with cystic bronchiectasis.

Pleural fluid analysis: Lymphocytic predominant exudative effusion with ADA 53 U/L, LDH 2250 IU/L.

Fiber optic bronchoscopy
Quadrupled division of right upper lobe bronchus seen. Left main bronchus appeared wide and spacious. Left upper lobe superior division showed blind pit like rudimentary subdivisions suggestive of blind-ended bronchi (Figure 2).

Bronchial washing for AFB was negative, gram stain showed a cluster of Gram-positive cocci, culture showed-growth of *Staphylococcus aureus*.

**Two-dimesional echocardiogram**
It showed mild pulmonary arterial hypertension with tricuspid regurgitation, TRJET 3 m/s, RVSP 46 mmHg with good left ventricular and right ventricular systolic function.

**Figure 1:** Chest X-ray showing reduced lung volume and bronchiectatic changes on left side and minimal pleural effusion on right side

**Figure 2:** Fiberoptic bronchoscopy showing blind pit like bronchial subdivisions in left upper lobe

**Figure 3:** Axial section of contrast enhanced computed tomography thorax in mediastinal window showing tubulocystic bronchiectasis in left lung with herniation of right lung anteriorly in to left hemithorax and hypoplastic left pulmonary artery

**Figure 4:** Computed tomography pulmonary angiography with three-dimensional volume rendered image showing right pulmonary artery (23 mm) and a much smaller left pulmonary artery (15 mm) at the level of bifurcation of main pulmonary artery
Initially, patient was managed symptomatically with bronchodilators and antibiotics. Later he was started on Cat 1 anti-tuberculous treatment under Revised National Tuberculosis Control Program and discharged. He came for review after 1 month and was found to be doing well. Repeat CXR showed clearance of right-sided pleural effusion. He is kept under regular follow-up.

**DISCUSSION**

Developmental anomalies of lung can be divided into three broad categories: (1) broncho pulmonary (lung bud) anomalies, (2) vascular anomalies, (3) combined lung and vascular anomalies. Majority of the developmental abnormalities that affect lung are manifested at or soon after birth. Very rarely they are encountered in adolescence or adulthood. Development of the bronchial tree takes place at about 26th-31st day of intrauterine life. Shneider classified lung maldevelopment into three groups, which was subsequently modified in 1955 by Boyd. They are: Type 1: Agenesis: In which there is complete absence of one or both lungs, with no traces of bronchial or vascular supply or of parenchymal tissue. Type 2: Aplasia: In which there is a suppression of all, but a rudimentary bronchus that ends in a blind pouch, with no evidence of pulmonary vasculature or parenchyma. Type 3: Hypoplasia: In which the gross morphology of the lung is essentially unremarkable, but there is a decrease in number or size of airways, vessels, and alveoli. Our patient classified as Type 3.

Monaldi divided mal development of lung into 4 groups. They are: Group 1: No bifurcation of the trachea. Group 2: Only rudimentary bronchus, Group 3: Incomplete development after division of the main bronchus. Group 4: Incomplete development of sub segmental bronchi and a small segment of corresponding lobe.

Hypoplasia may be primary (idiopathic) or secondary when it occurs in association with environmental factors or other congenital anomalies that may be implicated in its pathogenesis. The incidence of primary hypoplasia is estimated to be 1-2/12,000 or 15,000 births without sex predilection. By definition, primary hypoplasia is unassociated with other anomalies. It may represent an intrinsic defect in the lung development. Primary pulmonary hypoplasia may be caused by deficiency in thyroid transcription factor-1, GATA factors, hepatocyte nuclear factor 310, epidermal growth factor receptor alpha and mitogen activated protein kinase. The incidence of secondary hypoplasia is difficult to determine because of its association with a variety of other anomalies and the difficulty of pathologic diagnosis in some. Several mechanisms have been implicated in secondary pulmonary hypoplasia such as: a) decreased volume of ipsilateral hemithorax due to a space-occupying mass, b) decreased pulmonary vascular perfusion, c) neurological abnormalities resulting in decreased fetal respiratory movements, d) anomalies of kidney and urinary tract causing oligohydramnios.

Pathologically the most consistent finding is decreased in the number of airway generation ranging from 50% to 75% of normal. The number of alveoli is also reduced to one-third of normal. The basis of variation in morphologic findings may be related to severity and cause of hypoplasia as well as to the timing of etiologic events that may lead to anomaly.

CXR, fiberoptic bronchoscopy, CT thorax, CT pulmonary angiography, magnetic resonance imaging, bronchography, ventilation perfusion scans are useful modalities of diagnosis and assessment of hypoplasia lung. The radiographic findings of hypoplasia are characterized principally by total or almost total absence of aerated lung in one hemithorax. The reduced lung volume is indicated by approximation of the ribs, elevation of the ipsilateral hemidiaphragm and shift of the mediastinum. In most cases, the contralateral lung is greatly over inflated and placed along with the anterior mediastinum into the involved hemithorax. Radiological differential diagnoses include the total atelectasis of the lung, severe bronchiectasis with collapse, advanced fibrothorax, remote rupture of the main bronchus and pneumonectomy. Multidetector CT with advanced post processing techniques like multi-planar reconstruction and 3D volume rendering technique makes it an ideal noninvasive method for evaluation of congenital lung anomalies like hypoplasia of lung.

It is particularly helpful in evaluating the affected pulmonary artery, enlarged contralateral pulmonary artery and multiple collateral vessel formations. Clinical findings depend upon the degree of pulmonary abnormality and the presence of congenital malformation elsewhere. Patients with pulmonary hypoplasia are susceptible to recurrent respiratory infections due to distorted airways, defective mucociliary clearance and pooling and spilling of secretions from blind ended bronchus.

**Treatment**

Treatment is divided into medical and surgical care, both before and after delivery. Before delivery, patient is treated medically with repeated amnioinfusions with or without tocolytics, antibiotics and steroids. After delivery respiratory support is given in the form of oxygen, mechanical ventilation and extra corporeal membrane oxygenation. Surfactant administration at 4 ml/kg is said to improve survival rate. Surgical care consists of intrauterine vesicoamniotic shunts and endoscopic ablation of valves and plug the lung until it grows by fetoscopic tracheal occlusion with a clip. Post-delivery surgery can be done if there is any diaphragmatic hernia or cystic adenomatoid malformation or decompressive pleural effusion. Treatment in adults is mainly on conservative lines with antibiotics for
infections, symptomatic treatment with bronchodilators and prophylactic pneumococcal and influenza vaccinations. The present case was managed symptomatically along with anti-tuberculous treatment for right-sided tuberculous pleural effusion. In the follow-up months, the tuberculous pleural effusion resolved. However since the patient already developed pulmonary arterial hypertension, which is not due to congenital heart disease, but due to reduced vascular bed, he is regularly being monitored.

CONCLUSION

In cases of recent or long standing recurrent respiratory infections with homogenous or inhomogenous opacity of one hemithorax with herniation of the opposite lung, possibilities of hypoplasia/aplasia should be considered. We report a rare case of primary pulmonary hypoplasia in adults, who presented with complications of pulmonary hypertension and tuberculous pleural effusion. Regular follow-up of such cases with prompt management of complications like secondary infections, hemoptysis, and high altitude pulmonary edema is recommended to prevent progression to cor pulmonale or chronic respiratory failure.