

Case Report

Unilateral lung hypoplasia presenting in seventh decade of life: a rare case report

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Received: 20 August 2018

Accepted: 26 September 2018

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ABSTRACT

Pulmonary hypoplasia is a bronchopulmonary foregut anomaly in which gross morphology of the lung is preserved but there is decrease in the number or size of airways, vessels, and alveoli. Unilateral pulmonary hypoplasia is a rare clinical condition and most patients reported in the literature are newborns and infants, but patients may remain asymptomatic until late adolescent or till adulthood 65 years old male with parkinsonism was referred to our department with complaints of recent chest symptoms. Clinical examination revealed loss of lung volume in left hemithorax. CT scan thorax and bronchoscopy confirmed the diagnosis of left sided pulmonary hypoplasia. This case report highlights the possibility of pulmonary hypoplasia as one of the differential diagnoses in adult patients presenting with loss of lung volume and to the best of our knowledge he is the eldest patient so far reported.

Keywords: Asymptomatic, Elderly, Hypoplasia

INTRODUCTION

Pulmonary hypoplasia is a rare congenital foregut anomaly characterized by decreased number of airways, pulmonary parenchyma and vessels. It's commonly identified in infants and children.¹ Although rare even in adults several case reports are available.²⁻⁵

The developmental anomalies of the lung at the 4th and 24th weeks of gestation may cause bronchopulmonary foregut abnormalities. According to Boyden, there are three degrees of maldevelopment.⁶

- Agenesis- complete absence of lung tissue (absence of airways, parenchyma and vessels)
- Aplasia- where rudimentary bronchus is present without any lung tissue, and
- Hypoplasia - in which all the normal pulmonary tissues are present but are under-developed.

Hypoplastic region of the lung becomes small, fibrotic and dysfunctional as a consequence of developmental delay in the alveolar tissue.

Monaldi divided the mal-development of lung in four groups:⁷

- Group I: No bifurcation of trachea;
- Group II: Only rudimentary main bronchus;
- Group III: Incomplete development after division of main bronchus; and
- Group IV: Incomplete development of subsegmental bronchi and small segment of the corresponding lobe.

Unilateral pulmonary hypoplasia is a rare congenital anomaly. Although its exact prevalence is not well known, it is estimated to be seen in approximately one in 15,000 live births, or between 0.0034% and 0.0097%

without sex predilection.^{6,8,9} The right and left lung are affected with equal frequency.¹⁰⁻¹² It is very commonly associated with other congenital anomalies of the diaphragm, urinary system, cardiovascular system (tetralogy of fallot), central nervous system (anencephaly, hydrocephaly), and musculoskeletal anomalies of thoracic cage, Klippel Feil syndrome (KFS) and Downs syndrome.¹³

Ours is a case of isolated left lung hypoplasia belongs to the third group of mal-development according to Boyden classification, and group III of Monaldi classification. And this case is unique in view of its very late age of presentation and probably unmasked by the repeated aspirations caused by Parkinson's disease

CASE REPORT

A 65-years elderly male patient with known parkinsonism was referred to us from neurology department for productive cough and breathlessness for last 6 months with presumptive diagnoses of pulmonary Koch's. He had productive cough that aggravated with water intake and gradually progressive exertional breathlessness without any associated fever, wheeze, chest pain, hemoptysis, orthopnea or paroxysmal nocturnal dyspnea. Anti TB medications were started empirically from outside for 2 months. Patient had no hospital admission history for respiratory complaint till date.

On general physical examination mild pallor, grade 3 clubbing and pill rolling movement of bilateral fingers was seen without any jaundice, cyanosis, pedal edema or peripheral lymphadenopathy.

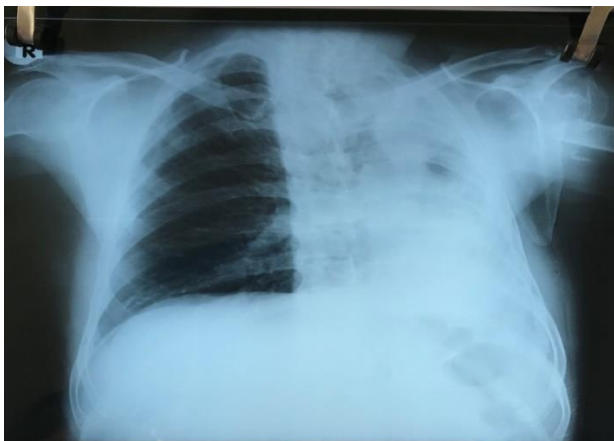


Figure 1: Hypoplasia of the left lung with compensatory increase aeration of the right, mediastinal shift to the left and elevated left diaphragm on postero-anterior chest radiograph.

Chest examination showed left shoulder drooping, scoliosis with tracheal deviation to left, rib crowding on left side, apical impulse shifted to left. Reduced respiratory sounds on the left hemi-thorax, dull on

percussion with normal vesicular breath sounds on right. Postero-anterior chest radiograph showed volume loss in the left lung with compensatory hyper aeration of the right lung, mediastinal shift to the left and elevation on the left diaphragm (Figure 1).

Routine hemogram, biochemical analyses were normal and Mantoux test was nonreactive. Sputum analysis for AFB was negative for 3 consecutive samples. CECT chest (Figure 2 and Figure 3) showed hypoplastic left main pulmonary artery, left lung parenchyma and left main bronchus. Trachea and mediastinal shift toward left with hyperinflated right lung was observed. Esophagus was dilated.



Figure 2: Hypoplasia of the left pulmonary artery and left lung on thoracic computed tomographic angiography.



Figure 3: Right lung herniation, hypoplastic left lung tissue, main pulmonary artery dividing into right and left main pulmonary arteries, mediastinal shift to left.

Bronchoscopy showed partial narrowing of left main bronchus with thick mucopurulent secretions, scope could not be negotiated into left upper and lower lobe bronchi. Right bronchial tree appeared normal. Bronchial wash was negative for mycobacteria on ZN stain and culture. Spirometry test showed restrictive pattern with reversible airway obstruction. With all these findings

patient was diagnosed to have asymptomatic left pulmonary hypoplasia previously and repeated aspirations due to Parkinson's disease have led to unmasking of the lung condition. Patient showed improvement with empirical treatment with antibiotics and with bronchodilators. Anti TB medications were stopped. We advised further evaluation of aspiration and preventive measures, but patient denied.

DISCUSSION

Hypoplasia of the lung may be regarded as primary (idiopathic) or secondary. Absence of any predisposition suggests primary pulmonary hypoplasia,¹ whereas secondary pulmonary hypoplasia results from small foetal thoracic volume, prolonged oligohydramnios, early rupture of membranes at 15-28 weeks 'gestation longer latent period before delivery, decreased foetal breathing, decreased pulmonary perfusion, congenital heart diseases and trisomies 18, 13, 21.^{3,6,10}

Unilateral pulmonary hypoplasia can present in late adolescent or in adult life as an infective pathology when pulmonary function is decompensated. Clinical presentation of lung hypoplasia in adult is highly variable like asymptomatic presentation or repeated respiratory infections. Patients with long survival into adulthood with minimal or almost no respiratory symptoms can be explained by contralateral lung hyperinflation compensating for the hypoplastic hemithorax as in cases of pneumectomy. Left lung hypoplasia shows best survival because of the good compensatory hyperinflation of the larger right lung.⁵

Symptomatic treatment is the mode of management for adults and older children in form of expectorants, bronchodilators, empirical antibiotics, control of recurrent infections, and management of other complications. Prophylaxis with pneumococcal and influenza vaccines are recommended. Asymptomatic cases do not require any treatment if there are no additional anomalies. Recurrent pneumonia due to blind ending bronchi and a susceptibility to infection affect the prognosis. Surgery is seldom required for agenesis or aplasia. Limited surgical intervention with the resection of blind-ending bronchi can be performed in such patients.¹ Prognosis depends on the size of the lesion, the degree of functional impairment, and the associated congenital malformations.

CONCLUSION

Although it is seen rarely, pulmonary hypoplasia should always be included in differential diagnosis in adolescents and adult patients presenting with loss of lung volume. Protection of the other lung is of immediate

priority and treatment measures should be directed towards that.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Chandra B, Bhaskar K, Paramjyothi GK. Unilateral lung hypoplasia presenting in seventh decade of life: a rare case report. *Int J Adv Med* 2018;5:1532-4.