Incidental Case of Left Lung Hypoplasia in a Postabortal Young Female

Abstract

Congenital malformations of the lung are extremely rare with an incidence of pulmonary hypoplasia around 1-2/12,000 births. Boyden has categorized three degrees of malformation including (i) agenesis in which there is complete absence of the lung and bronchus and no vascular supply to the affected side, (ii) aplasia in which there is the presence of rudimentary bronchus with the absence of pulmonary parenchyma, and (iii) hypoplasia in which there are variable amounts of bronchial tree, pulmonary parenchyma, and supporting vasculature. Here, we present an incidental case of pulmonary hypoplasia in a young female.

Keywords: Computed tomography, fiberoptic bronchoscopy, pulmonary angiography, pulmonary hypoplasia

Introduction

Pulmonary hypoplasia is a rare congenital disorder in that the exact cause is not completely known. Usually, the entity is unilateral and is characterized by a decrease in the number or size of the airways, vessels, and alveoli resulting in a small fibrotic and nonfunctioning lung sometimes associated with other congenital anomalies and bronchiectasis.\(^\text{[1]}\) Computed tomography, pulmonary angiography, and fiberoptic bronchoscopy give clues and are helpful in reaching to the diagnosis; however, clinical diagnostic criteria are lacking regarding this entity. Computed tomography is useful in differentiating hypoplasia from mimicking conditions on chest X-ray. Incidence of pulmonary hypoplasia ranges from 1 to 2/10,000 live births. Both genders are affected almost equally. Boyden, in 1955, has classified the degree of lung involvement into three groups as pulmonary agenesis, pulmonary aplasia, and pulmonary hypoplasia.

Case Report

A 25 years female with bad obstetric history of two abortions with the last abortion (both of them male child) done around 5 days back with 4 months of gestational age, after that the patient developed breathlessness and fever probably due to some secondary infection related to the procedure. A chest X-ray [Figure 1] was done and accidentally found that the left lung field was whiteout and the patient was referred to our side for further evaluation. The patient detailed history was taken which suggested recurrent respiratory infections; she also had a history of taking antitubercular treatment at the age of 11-12 years; however, no documented evidence of pulmonary tuberculosis was there, she was admitted in the department for further workup. Trachea and heart were shifted to the left side. Movements were diminished and percussion note was dull over the left hemithorax. On auscultation, air entry was decreased on the left side. Other system examinations were within normal limits. Routine hematological investigations suggested anemia for which patient was given blood transfusion. Sputum smear for acid-fast bacilli and Gram stain was negative on direct smear examination. She was further advised a computed tomography of the chest [Figure 2] which was suggestive compensatory hyperinflation and herniation of the right lung with inadequate development of the lung on the left side including the airways and the vasculature which was confirmed with angiography [Figure 3], and the patient was subsequently subjected for fiberoptic bronchoscopy [Figure 4a and b], the right tracheobronchial tree was patent with opening of all segmental bronchi patent, mucosa was healthy, but on the left, there was only a small depression in place of
the left main bronchus with no opening to negotiate the bronchoscope (underdevelopment of the left bronchial tree). The patient otherwise responded to the antibiotics and other symptomatic treatment and was discharged in satisfactory condition with advice for regular follow-up.

Discussion

Lung malformations are very rare to occur with varying degree of severity and with incidence as low as 1–2/12,000 births, these are the results of insult to the embryo during the 4th to 5th week of intrauterine life.[2] Boyd has categorized them as pulmonary agenesis, aplasia, and hypoplasia. Monaldi on the other hand has divided the malformation into four groups, 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).[3] Agenesis of the lung was described first in 1673 by De pozze, and from India, the first case was reported by Muhammed in 1923. Causes of hypoplasia may be primary like deficiency of thyroid transcription factor-1, (guanine, adenine, thymine, guanine) transcription factor, hepatocyte nuclear factors, epidermal growth factor and its mitogen-activated protein kinase receptor, or secondary causes such as small fetal thoracic volume, prolonged oligohydramnios, early rupture of membranes, congenital heart diseases, and trisomies 13, 18, and 21.[4]

Around half of the cases of pulmonary hypoplasia or aplasia have associated congenital defects involving cardiovascular, skeletal gastrointestinal, and genitourinary systems.[5] In many cases, pulmonary hypoplasia presents with recurrent chest infections or symptoms of cardiopulmonary insufficiency, and there may be the presence of other congenital anomalies. Hypoplastic lungs are smaller and lighter with the most consistent finding of decrease in number of airway generation; frequently, there is also decrease in number of alveoli as well as decrease in their size. Pulmonary arterial abnormalities can also be often seen. Morphologic findings may be related to severity and cause of hypoplasia as well as to the timing of etiologic events that led to anomaly.[6]

Clinical diagnostic criteria are lacking regarding malformation of the lung. Computed tomography, pulmonary angiography, and fiberoptic bronchoscopy give clues...
and are helpful in reaching to the diagnosis.\textsuperscript{[7]} Computed tomography is useful in differentiating hypoplasia from mimicking conditions on chest X-ray. One of the important differential diagnoses is Swyer-James syndrome which is radiologically characterized by the presence of hyperlucent lung or lobe and functionally by the presence of air trapping during expiration which can be seen in expiratory roentgenograms and computed tomography.\textsuperscript{[8]} Few cases of lung malformation have also be seen in association with asthma.\textsuperscript{[9,10]} Extracorporeal membrane oxygenation, surfactant administration, intrauterine vesicoamniotic shunts, and endoscopic ablation of valves have been tried as treatment modalities, in adults, treatment comprises control of recurrent infections, symptomatic approach, and management of complications. Vaccination prophylaxis for respiratory syncytial virus, influenza, and pneumococcus is recommended.\textsuperscript{[11]}

\textbf{Declaration of patient consent}

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

\textbf{Acknowledgments}

The authors wish to thank the patient for her cooperation and following the instructions. The authors have no conflict of interest.

\textbf{Financial support and sponsorship}

Nil.

\textbf{Conflicts of interest}

There are no conflicts of interest.

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