UNILATERAL PULMONARY HYPOPLASIA – A CASE REPORT

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ABSTRACT
A case of right sided pulmonary hypoplasia is described. A 20 years male was admitted in our department with complaints of recurrent chest infections since childhood. Clinical examination revealed smaller right hemithorax. Serial chest X-ray showed nonprogressive lesions over several years. Bronchoscopy and CT scan thorax confirmed the diagnosis of right sided pulmonary hypoplasia. It was not associated with any other developmental anomaly.

Key words : Hypoplasia, Bronchoscopy, CT scan thorax.

INTRODUCTION
Hypoplasia of lung is a rare congenital anomaly in which gross morphology of the lung is essentially unremarkable but in which there is decrease in number or size of airways, vessels and alveoli. Under development of alveolar tissue results in small fibrotic and non functioning lung. Bronchiectatic changes have also been reported in the hypoplastic lung. This condition is frequently associated with other congenital anomalies. Common developmental anomalies associated with pulmonary hypoplasia involve urinary system, diaphragm, cardiovascular system (Tetralogy of Fallot), central nervous system (anencephaly and hydroencephaly), as also musculoskeletal anomalies of thoracic cage, Klippel Feil syndrome and Down syndrome.

CASE REPORT
A 20 years male was admitted in Department of Pulmonary Medicine, King George’s Medical University, Lucknow with chief complaints of cough and expectoration, off and on fever, breathlessness increased on exertion since childhood. Patient also complained of hemoptysis several episodes, varying in amount from streaking of blood in sputum to 200-300 ml/episode since six years. There was history of dull aching pain over right side of chest for four years and loss of appetite for two months.

General physical examination was non-contributory. Examination of respiratory system revealed smaller right hemithorax. Trachea and heart were shifted to right side. Movements were diminished and percussion note was dull over the right hemithorax. On auscultation, air entry was very poor on right side with diffuse crepitations. Rest of systems were within normal limits.

Routine haematological investigations (Blood counts, Liver function tests, renal function tests) were within normal limits. Sputum smear for Acid Fast Bacilli was negative on direct smear examination and later on culture. Chest roentgenogram showed a right sided massive homogeneous opacity, shifting of the mediastinum, and chest retraction. The intercostal spaces on right side were narrowed. Left lung was overinflated (Photograph-I). The lesions were non progressive on serial chest X-rays.

Bronchoscopy revealed the under development of right bronchial tree. Right upper lobe bronchus was seen arising just near the carina and only two small openings were seen. Middle and lower lobe bronchus were under developed. However, normal development of tracheobronchial tree of left side was seen. CT Scan Thorax revealed marked asymmetry in thorax. The left lung showed herniation through anterior recess with evidence...
of oligaeemia suggesting compensatory over inflation. The right sided lung showed severe decrease in volume with only minimal residual lung tissue. Bronchiectatic changes were seen in this lung tissue. The mediastinum was seen to be shifted to right side (Photograph-II). Patient is still under our follow up and treatment. Patient develops recurrent chest infections which respond to broad spectrum antibiotics and symptomatic treatment.

DISCUSSION

Development of the bronchial tree takes place at about 26th to 31st day of intrauterine life. Monaldi divided the mal-development of lung in 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. The present case belongs to the fourth group of Monaldi classification.

According to Boyden there are three degrees of mal-development: (i) agenesis, in which there is complete absence of lung tissue, (ii) aplasia, in which rudimentary bronchus is present but no lung tissue is present, and (iii) hypoplasia, in which all the normal pulmonary tissues are present but are under-developed.

Hypoplasia of the lung may be regarded as 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 secondary form is difficult to determine; however, because of its association with a variety of other abnormalities and the difficulty of pathologic diagnosis in some cases, it is likely to be more common than generally recognized. The incidence of primary hypoplasia has been estimated to be 1 to 2 per 12,000 births. Several mechanisms have been implicated in secondary pulmonary hypoplasia including decreased hemithoracic volume, decreased pulmonary vascular perfusion, decreased fetal respiratory movement and decreased lung fluid.

Although the pathologic diagnosis of pulmonary hypoplasia can be made on formalin-inflated, routinely processed lungs on the basis of a combination of fresh lung weight, fixed lung volume, radial alveolar count and estimates of tissue maturity, precise characterization of the morphologic changes is best performed by morphometric measurement after inflation of the lungs to a known transpulmonary pressure.

Hypoplastic lungs are typically smaller and weigh less than normally expected for their age. Although there is variation in the severity and type of changes between different cases, the most consistent finding is a decrease in number of airway generation, ranging from about 50% to 75%, of normal. In addition, there is frequently a decrease in the number of alveoli, estimated by one group of investigators to be about one-third normal. This is often associated with a decrease in alveolar size. Some investigators have shown normal airway and alveolar maturation for gestational age; others have found an immature appearance. Abnormalities of pulmonary arterial system have also been identified consisting of decreased elastic tissue in the larger arteries, increased muscle in normally muscular arteries, and extension of muscle into normally non-muscular arteries.

The basis of variation in morphologic findings may be related to severity and cause of hypoplasia as well as to the timing of the etiologic events that led to anomaly.

Radiographic findings in cases of hypoplasia are similar and characterised principally by almost total absence of aerated lung in one hemithorax. The markedly reduced volume is indicated by approximation of ribs, elevation of ipsilateral diaphragm, and shift of the mediastinum. In most cases the contralateral lung is greatly over inflated and displaced along with anterior mediastinum into the involved hemithorax; this displacement of air containing lung to the side of involved lung may lead to some confusion in diagnosis. CT scan may be required to establish the degree of under development and to differentiate hypoplasia from other conditions that may closely mimic it radiographically: atelectasis from other causes, severe bronchiectasis with collapse and advanced fibrothorax. Main differential diagnosis of hypoplastic lung is Swyer James syndrome. Swyer James syndrome is an uncommon abnormality characterized radiologically by hyperlucent lobe or lung and functionally by air trapping during expiration. There is substantial evidence that the syndrome is initiated by viral bronchiolitis. Although both conditions are associated with unilateral very low volume, patients with Swyer James.
syndrome characteristically demonstrate an air trapping on radiographs or HRCT scans performed at the end of maximal expiration16.

Clinical findings depend on degree of pulmonary abnormality and presence of other congenital malformations. Usually, however the patient is symptomatic. Physical examination characteristically reveals asymmetry of two sides of thorax, reduction in respiratory movements and absence of air entry in the affected side. This may be diagnosed incidentally during childhood when complicated by pulmonary infection19-21. Diagnosis may be established with help of chest x-ray, CT thorax17-18, fiber optic bronchoscopy, and if possible pulmonary angiography and bronchography. Still however there are no clear clinical diagnostic criteria to facilitate the identification and management of lung hypoplasia. Hence current research is based on identification of such clinical diagnostic criteria22.

Treatment20 of hypoplasia is in form of medical as well as surgical care, both before and after delivery. Before delivery patient is treated medically when repeated amniofusions with or without the use of tocolytics, antibiotics and steroids. After delivery respiratory support is given ranging from oxygen to mechanical ventilation including ECMO (Extracorporeal membrane oxygenation). Dialysis may be required for support of renal function.

Surfactant administration at 4 ml/kg improves survival rate. Surgical care consists of intrauterine vesicoamniotic shunts and endoscopic ablation of valves and PLUG (plug the lung until it grows) by fetoscopic tracheal occlusion with a clip. Post-delivery surgery can be done to correct diaphragmatic hernia, cystic adenomatoid malformations and decompresses pleural effusions.

Treatment in adults consists of control of recurrent infections, symptomatic treatment in form of expectorants and bronchodilators and management of other complications.

Prophylaxis for respiratory syncytial virus, pneumococcus, influenza infections are recommended.

REFERENCES