Pulmonary Hypoplasia Wrongly Diagnosed and Treated as Pulmonary Tuberculosis

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Abstract
Pulmonary hypoplasia (PH), a rare congenital anomaly is observed with incomplete development of the lung, and may be associated with anomalies in other body systems. The clinical presentation varies with the extent of hypoplasia and the patient may be asymptomatic or may present with severe respiratory distress in the neonatal, infancy or childhood period. A six year old girl suffering from right sided hypoplasia was hospitalized with common presenting chest symptoms. She had taken antituberculosis treatment for past three years and was thought to be an MDR suspect. The child was thoroughly investigated was diagnosed to be a case of PH and is under followup.

Introduction
Pulmonary hypoplasia (PH), a rare congenital anomaly is observed with incomplete development of the lung, may be associated with anomalies in other body systems such as cardiovascular, renal, musculoskeletal, central nervous system and GI. The lung fails to attain the maturity, which is due to the decrease in number of lung cells, airways, vessels, and alveoli resulting in decrease in the size and weight.¹ However the gross morphology of the lung is essentially unremarkable.² The delay in alveolar tissue development causes underdeveloped lung which becomes small, fibrotic and non-functioning. The clinical presentation varies with the extent of hypoplasia, and the patient may be either asymptomatic or may present with severe respiratory distress in the neonatal, infancy or childhood period.² A case of PH wrongly diagnosed as suffering from pulmonary tuberculosis is presented.

Case Report
A six years old girl was treated by private medical practitioners/ hospitals and was referred to the department of Pulmonary Medicine, Rohilkhand Medical College and Hospital, Bareilly (UP) as a case of suspected MDR-TB for establishing the diagnosis and management with the complaints of breathlessness increased on exertion, cough with expectoration, mild chest pain, cold, fever off and on and loss of appetite for three years. There was no h/o haemoptysis. The h/o recurrent infection (LRTI) since 6 months of age was noticed. There was no h/o jaundice, seizure etc. The child had not been fully immunized except oral Polio vaccine. The child was given antituberculosis treatment for past three years. The family history was unremarkable. General physical examination was non-contributory except kyphoscoliosis. The respiratory system examination revealed impairment on right side. Air entry and movements of the chest were diminished on right side with restricted expansion of the chest. Trachea and heart were shifted to right side. The percussion note was dull and diffuse crepitations were present on right side. There was no remarkable finding in other body systems.

Direct smear microscopy of sputum specimen and culture was negative for acid-fast bacilli. The routine haematological examination such as blood counts, liver and renal function tests, blood sugar examination were within normal limits. Chest roentgenogram revealed right sided massive homogenous opacity, shifting of mediastinum, and chest retraction. The intercostal spaces on right side were narrowed. Left lung showed compensatory emphysema (Figure 1). X-ray whole spine revealed kyphoscoliosis and hemivertebra (at the level of T3) (Figure 2). CECT thorax showed right sided markedly reduced lung volume associated with hypoplastic right pulmonary artery with kyphoscoliosis. There was no evidence of any fibrocavitatory lesion (Figure 3). Bronchoscopy showed underdevelopment of right upper lobe bronchus and only two openings could be seen, middle lobe bronchus could not be seen and lower lobe bronchus was normally visualized. There was normal development of tracheobronchial tree on left side.

The child was discharged on symptomatic treatment. In the last two visits, she was observed to be doing well. She showed relief in
symptoms and is still under follow up.

Discussion

Pulmonary hypoplasia is the failure of the development of the lung in the utero, which is often unilateral and can range from hypoplasia to aplasia resulting in an abnormally low number and size of bronchopulmonary segments or alveoli. The lungs are abnormally small and the hypoplastic lung doesn’t have enough tissue and blood flow to allow the individual to breathe on his/her own. This condition can be grave and sometimes fatal.1,2

Monaldi categorizes developmental disorders of the lung in four groups.4 I. No bifurcation of trachea; II. Rudimentary main bronchus only; III. Uncompleted development after bifurcation of main bronchus; IV. Incomplete development of small segment and subsegmental bronchi of corresponding lobe. According to Boyden,5 the developmental disorders are seen in three different extents: (1) agenesis: complete absence of lung tissue, (2) aplasia: no lung tissue but rudimentary bronchus, (3) hypoplasia: all lung tissues exist but are underdeveloped. The present case belongs to fourth group of Monaldi classification.

The causes of PH are multiple and include (a) Thoracic: Congenital diaphragmatic hernia, extra lobar sequestration, agenesis of diaphragm, mediastinal mass(es), teratoma, decreased pulmonary vascular (arterial) perfusion from a congenital cardiovascular anomaly e.g. Fallot’s tetralogy or unilateral absence of pulmonary artery (b) Extrathoracic: oligohydramnios, Potter sequence, renal abnormalities, preterm premature rupture of membrane (PPROM); skeletal dysplasias causing narrow fetal thorax, Jeune syndrome – asphyxiating thoracic dystrophy, thanatophoric dysplasia, achondroplasia, osteogenesis imperfecta, short rib polydactyly syndrome, campomelic dysplasia, Klippel Feil syndrome, Down syndrome; large intra-abdominal mass compressing thorax; neuromuscular disorders interfering fetal breathing; CNS - anencephaly, hydroencephaly; urogenital and GI.1-3

The diagnosis of PH is made on the basis of clinical presentation. The pathological diagnosis requires formalin inflated routinely processed lung. The important tests are (1) lung weight (2) lung volume (3) lung volume: birth weight ratio of ≤1.2%. (The ratio ≤ 0.9% indicates PH) (4) Radial alveolar count (RAC), (5) Biometric indices eg. thoracic circumference to head or abdominal circumference ratio, thoracic area (TA) minus heart area (HA), HA/ TA ratio (6) Estimation of tissue maturity.1,3

Imaging eg. Chest X Ray, 3 D ultrasound, Doppler ultrasound...
velocimetry, CT thorax, MRI, CT/MRI angiography, bronchography are beneficial in the diagnosis of PH. The other tests are required to exclude tuberculosis and other cardiopulmonary diseases.

The management of PH includes medical and surgical care. In prenatal medical intervention, the patient is treated medically when repeated amnioinfusions with or without the use of tocolytics, antimicrobials, steroids and use of fibrin glue are advised. In postnatal period, the respiratory support is given ranging from supplemental oxygen to mechanical ventilation. In prophylactic measures, the immunization against influenza, pneumococcus, and respiratory syncytial virus has been recommended.

The prognosis depends on the size of the lungs and the underlying cause. The survivors will have chronic lung problems, decrease in lung capacity, recurrent chest infections and impaired growth.

References