Opaque Hemithorax. Can We Ignore Pulmonary Aplasia As A Cause? – Case Report

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Congenital malformations of the lungs are rare disorders occurring with variable degree of severity. They are categorized as pulmonary agenesis, aplasia and hypoplasia with distinct clinical implications. The thoracic imaging appearance of pulmonary agenesis or aplasia consists of complete opacification of one hemithorax with severe volume loss evidenced by extensive shift of the cardiomediastinal structures toward the affected side. The complete absence of lung tissue and a pulmonary artery on the affected side confirms the diagnosis and may be best appreciated on thoracic computed tomography. Aplasia has been diagnosed during antenatal sonography but attempts to surgically correct the condition have not met with significant success. This paper describes the case of a young patient who had clinico-radiological evidence of severe hemithoracic volume loss (gross mediastinal shift and opaque hemithorax on chest radiograph) and had been treated for possible tuberculosis for long period of time without relief. CT examination confirmed absence of lung tissue on this side in the presence of a rudimentary bronchus, thereby confirming the diagnosis of pulmonary hypoplasia. It is stressed not to overlook the rare possibility of pulmonary agenesis/aplasia or hypoplasia whenever confronted with a skiagram of chest showing complete hemithoracic opacification.

Key words: CT of pulmonary hypoplasia, pulmonary aplasia, opaque hemithorax, Congenital lung malformations.

Unilateral absence of a lung or a lobe is rare congenital anomaly that in itself may cause surprisingly few clinical problems[1]. It is usually seen in infancy or early childhood. Patients who have no or mild associated anomalies may survive into adulthood.[2,3] Indeed absence of a lung may be encountered de novo in an adult who has had no symptoms referable to chest. Interestingly agenesis of the right lung appears to be strongly associated with oesophageal atresia[4]. Associated congenital malformations are present in nearly half of patients with lung congenital anomalies which are a major cause of morbidity and mortality.[5,6]

Non invasive imaging techniques have now emerged as the diagnostic modality of choice, especially CT Thorax. With the advent of these techniques, the exact diagnosis can be established without opting for invasive procedures like bronchography and pulmonary angiography[7].

In our country, an opaque hemithorax is often mistaken for fibrotic lung disease subsequent to pulmonary tuberculosis. This often results in patients erroneously receiving anti tuberculous therapy as was seen in our patient. This report highlights the fact that a high index of suspicion is required to diagnose congenital lung anomalies whenever confronted with an opaque hemithorax on an X-Ray of the chest.

Case Report

A thirteen year old, short stature, thin and emaciated male who had complaints of dyspnoea since childhood and intermittent cough for six years was referred to the Department of Radiology for an X-Ray of the chest. The patient had already received a full course of anti tuberculous therapy in a local tertiary care hospital renowned for cure of diseases of the chest. Consultation from some other hospitals had already been sought without reaching a final diagnosis.

Physical examination conducted in Cardiology Department had revealed decreased breath sounds on auscultation on lateral and back side of left hemithorax and trachea was found to be shifted to the left side on palpation. Other systems were found to be normal.

On X-Ray chest, there was complete opacification of the left hemithorax with some crowding of the ribs. Neither left hemidiaphragm could be discriminated nor was a cardiac silhouette detected. The heart and mediastinal structures were shifted to the left and a substantial segment of the air containing right lung was found herniating across the mid line into the left hemithorax. (Figure 1).

Keeping in mind the broad differential diagnosis of unilateral opaque hemithorax, patient was subjected to Spiral CT scan examination which revealed a markedly displaced carina with a rudimentary left main bronchus beyond which no lung tissue could be identified (Figure 2). Heart and mediastinal structures were grossly shifted to the left side and mediastinal vasculature was greatly distorted (Figure 3). Mediastinal lymphadenopathy or other stigmata suggestive of granulomatous infection could not be detected.

Abdominal ultrasonography and skeletal survey failed to reveal any concomitant congenital anomaly. The patient was planned for bronchoscopy and echocardiography but failed to return and was lost to follow up.

Discussion

Pulmonary underdevelopment has been classified into three groups by Schneider and Schwalbe (8). In group 1, bronchus and lung are absent (agenesis); in group 2, a rudimentary bronchus is present and limited to a blind-end pouch without lung tissue (aplasia); and in group 3, there is bronchial hypoplasia with variable reduction of lung tissue (hypoplasia).

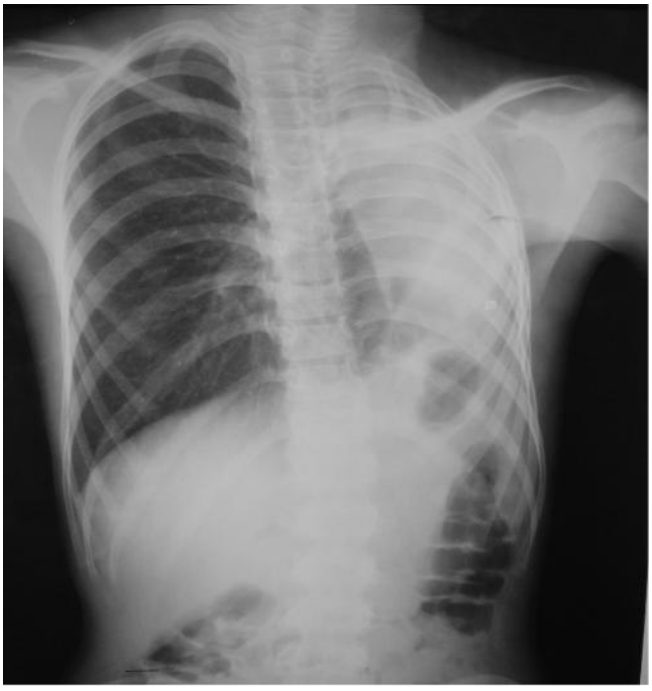

Fig. 1: *Pulmonary aplasia: X-Ray Chest showing complete opacification of the left hemithorax with marked volume loss. Over expanded right lung is seen herniating across the mid line into the left hemithorax. Marked chest asymmetry is quite evident.*

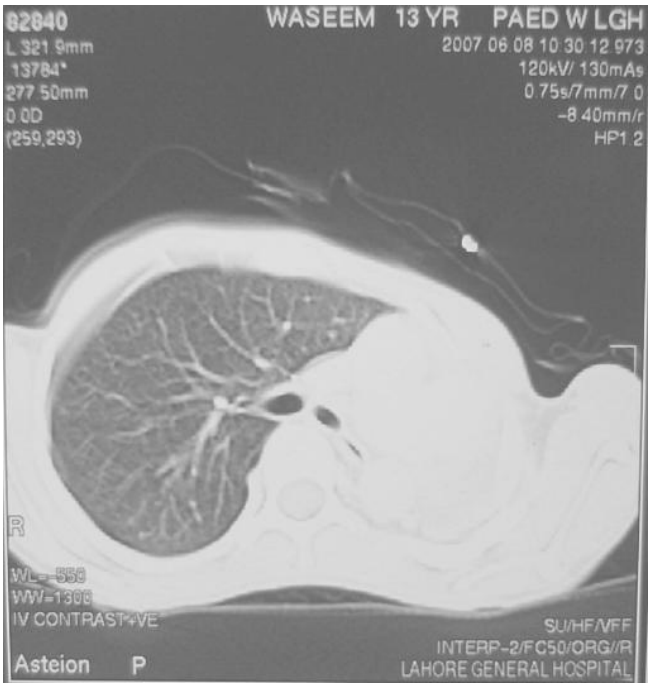

Fig. 2: *Pulmonary aplasia: CT Scan image on lung window shows hyper expanded right lung gaining access into the left hemithorax secondary to left pulmonary aplasia .Left main bronchus is clearly visualized differentiating it from pulmonary agenesis.*

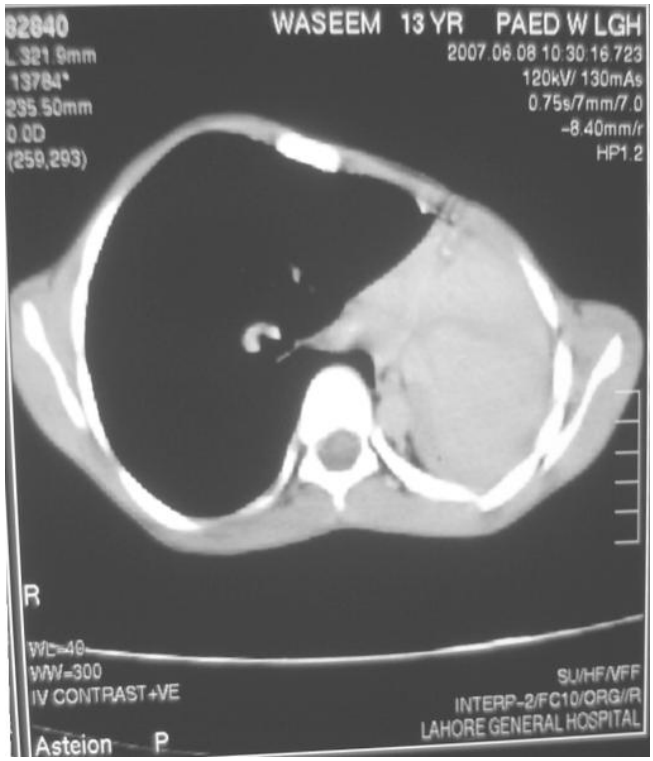

Fig. 3: *Pulmonary aplasia: Same patient. Mediastinal window CT slice reveals marked displacement of heart and major vessels into the left hemithorax due to absence of pulmonary parenchyma.*

Pulmonary agenesis is defined as a complete absence of carina, main bronchus, lung parenchyma and pulmonary vasculature. Pulmonary aplasia is similar except that a blind-ending rudimentary bronchus is present. The ipsilateral pulmonary artery develops but tends to be small or rudimentary[5]. Both pulmonary agenesis and hypoplasia maybe accompanied by other congenital anomalies of the vertebrae, anus, cardiovascular tree, trachea, esophagus, renal system and limb buds (VACTERL Syndrome) in up to 50% of the patients[9]. The survival rate is better with left sided agenesis because the right lung is larger of the two and excessive mediastinal shift and malrotation of carina in right sided agenesis hinders proper drainage of the functioning lung and increases chances of respiratory infections.[17] In practice, an etiologic, pathogenetic or clinical distinction between agenesis and aplasia is rare and the two conditions are usually considered together[10].

Pulmonary hypoplasia is defined as deficient or incomplete development of the lungs[11]. It is characterized by the presence of both bronchi and alveoli in an underdeveloped lobe, and it is caused by factors directly or indirectly compromising the thoracic space available for lung growth[8], such as a congenital diaphragmatic hernia, extra lobar sequestration, agenesis of the diaphragm, large pleural effusion, and Jeune syndrome (asphyxiating thoracic dystrophy. The extra thoracic causes include oligohydramnios and decreased pulmonary vascular perfusion (tetralogy of Fallot,

unilateral absence of the pulmonary artery). Intrathoracic causes, such as a congenital diaphragmatic hernia, are the most common.[11,12] The earlier the delivery of a child, the higher the incidence of lung hypoplasia which can approach 20% in babies delivered before 28 weeks of gestation. In lung hypoplasia, renal malformations, oligohydramnios, decreased fetal movements in neuromuscular disease, dysmorphisms in trisomies, and skeletal dysplasias may be identified[9].

There is no widely accepted embryological basis for lung anomalies and for this reason many theories have been presented. The major exception to these theories is agenesis of lungs. It is accepted that either a simple arrest of development occurs (bilateral agenesis) or there is failure to maintain the development balance of two lung buds[13].

Clinical presentation may vary depending on associated anomalies and/or existence of abnormalities in healthy lungs such as bronchiectasis. Some patients, those with no associated anomalies and/or no bronchiectasis or no recurrent respiratory infections might be symptom free or with minimal symptoms[10]. Patients who have symptoms could be misdialgnosed as bronchitis, recurrent pulmonary infections or cardiac disorders. These patients may have recurrent lung infections which are mainly due to bronchiectasis, rudimentary bronchus in aplasia or abnormal lung kinetics.

Diagnosis is usually made in neonatal period or in early childhood .There are some cases diagnosed antenatally. Chest X-ray is the key examination which leads a physician to further examination. The thoracic imaging appearance of pulmonary agenesis or aplasia consists of complete opacification of one hemithorax with severe volume loss evidenced by shift of cardio mediastinal structures towards the affected side. Such an appearance must be distinguished from a number of other conditions characterized by extensive pulmonary parenchymal loss such as pulmonary hypoplasia, main stem bronchial obstruction, extensive fibro thorax and pneumonechtomy[14].

Spiral CT, with its immense power of reformation, has revolutionized the visualization of respiratory system especially anatomy of bronchi and the lung parenchyma. Different reformation techniques include multiplanar reconstruction, shaded-surface display, MIP (Maximum intensity Projection), sliding thin slab imaging, volume rendering and bronchoscopy[15]. Contrast enhanced computed tomography is almost definitive for the diagnosis. It may show main vascular and bronchial structures as well as lung parenchyma. It also helps to determine existence of carina to distinguish agenesis from aplasia. Angiography or DSA might be necessary to show the vasculature. Angiography is a more accurate but a more invasive method than DSA. MRI and/or magnetic resonance angiography techniques may be used for the diagnosis and only MRI methods might be enough for reaching a diagnosis. Bronchography is seldom indicated any longer because CT scanning can demonstrate most (but not all) cases of bronchiectasis.

If pulmonary hypoplasia is diagnosed antenatally and judged to be incompatible with extrauterine life, some have suggested in utero intervention. This is done by occluding the fetal trachea with a balloon or clip. The accumulating fetal lung fluid seems to induce growth of the lung beyond normal. Surgical intervention may be necessary to manage airway narrowing. Tissue expanders have been used for this purpose. They offer the advantage that they can be slowly expanded over time by injecting saline through a subcutaneous port. Some prefer to use the old technique of placing ping-pong balls. This method creates a stable and long-lasting mass. As the patient grows, repeat operation to place more ping-pong balls is occasionally required[16].

The prognosis of patients with pulmonary hypoplasia depends on several factors, as follows:

- Associated anomalies.
- Pulmonary hypertension.
- Severe oligohydramnios, which increases the mortality rate.
- Preterm delivery or rupture of the membranes earlier than 28 weeks.
- Sidedness (Because the right lung is normally larger than the left, hypoplasia of the right lung is associated with a worsened outcome.)[16]

In a third world country like ours, most of these patients either remain undiagnosed or are treated for tuberculosis for long periods of time without a positive outcome. It is strongly believed and stressed that whenever a child or asymptomatic adult is found to have opaque hemithorax with signs of volume loss, suspicion should be raised about the possibility of lung aplasia-hypoplasia in addition to consideration of other pathological conditions falling into the differential diagnosis of opaque hemithorax.

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