Case Report

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ISOLATED LEFT LUNG HYPOPLASIA IN AN ADULT- A CASE REPORT

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ABSTRACT

Congenital anomalies of the lung occur due to various insults to the developing lung. Pulmonary hypoplasia is congenital anomaly that can present either in isolation or with other anomalies. However they are not routinely considered in the differential diagnosis in adults. We present a case of isolated left lung hypoplasia in an adult who had no significant respiratory symptoms. We believe this case report would help in better understanding of this condition and help to create further interest in other similar conditions.

Key words: Pulmonary hypoplasia, CT chest, Fiberoptic bronchoscopy

INTRODUCTION

Pulmonary hypoplasia is a developmental abnormality of the lung characterized by a decrease in the number of alveoli, cells, and airways with resultant decrease in size and weight of the lungs. Though predominantly a disease of infancy and childhood, its presentation in adults is not uncommon. Pulmonary hypoplasia has associated other congenital anomalies like renal agenesis,diaphragmatic hernia. Post natal diagnosis of this condition usually requires imaging studies and bronchoscopy. We present a case of isolated left lung hypoplasia in an adult who had few respiratory symptoms. The diagnosis was confirmed with fiberoptic bronchoscopy and computed tomography studies.

CASE SCENARIO

An asymptomatic 35 year old agricultural laborer presented with complaints of occasional left sided chest pain on moderate to severe exertion. He had no other respiratory complaints. He was not a smoker and had no co morbid illness. . He was first male child of a second degree consanguineously married parents. He is married since last 10 years and has two male children. He was initially evaluated outside for possible cardiac disease. His ECG showed no evidence of myocardial ischemia and he was referred to our respiratory medicine department for further evaluation. Upon presentation the patient was stable and maintained normal saturation at room air. There was no pallor or clubbing. Respiratory system showed trachea deviated to the left, diminished movements on the left side and a mild drooping of left shoulder. The apex beat was palpable in the left sixth intercostal space in mid axillary line. Breath sounds were absent in the lower left chest. Hematological investigations were within normal limits. Echocardiography showed a mild Tricuspid regurgitation. Spirometry revealed a mixed airway pattern. Chest x ray (fig. no 1) showed tracheal and mediastinal shift to left with crowding of ribs on the left upper zone with a hyper inflated right lung. A possibility of left lung collapse was suspected and he was planned for computed tomography(CT) of the thorax and fiber optic bronchoscopy. CT Scan Thorax (fig 2) revealed marked asymmetry in thorax. The right lung was hypertrophied and was observed to extend to the left hemithorax through anterior recess. The left lung showed a compressed left main bronchus with only minimal residual lung tissue. No endo-bronchial lesion was observed. Bronchiectatic changes were seen in the left lung tissue. The mediastinum was shifted to left side. The pulmonary arteries were normal. A possibility of left lung hypoplasia was considered and a fiber optic bronchoscopy(FOB) was performed. Bronchoscopy showed a normal trachea, carina and right side bronchial tree. The left main bronchus was narrowed and slit like in appearance. On negotiating the left main bronchus, left lobar bronchi were seen but

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ended in a blind pouch. (Fig 3, 4, 5). The diagnosis of left sided pulmonary hypoplasia was confirmed and patient evaluated for other congenital anomalies. He had no associated congenital anomalies. Retrospective questioning of the patient's antenatal and early post natal history revealed no significant events. Currently the patient is managed symptomatically and is on regular visit.

Figure 1: Chest x ray demonstrating a mediastinal shift to left with crowding of ribs on the left with hypo- lucent opacity on the left side

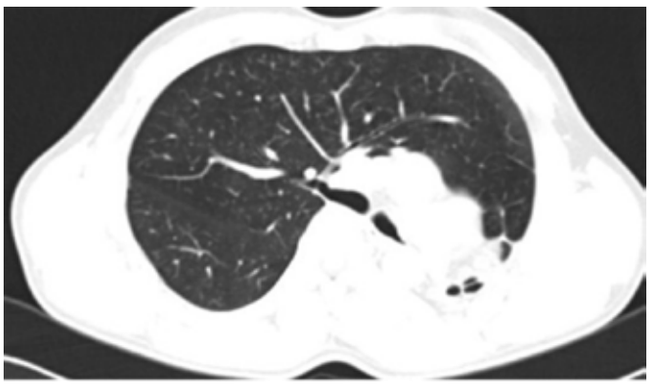

Figure 2: CT chest showing compressed left lung with hyper-inflated right lung herniating to left

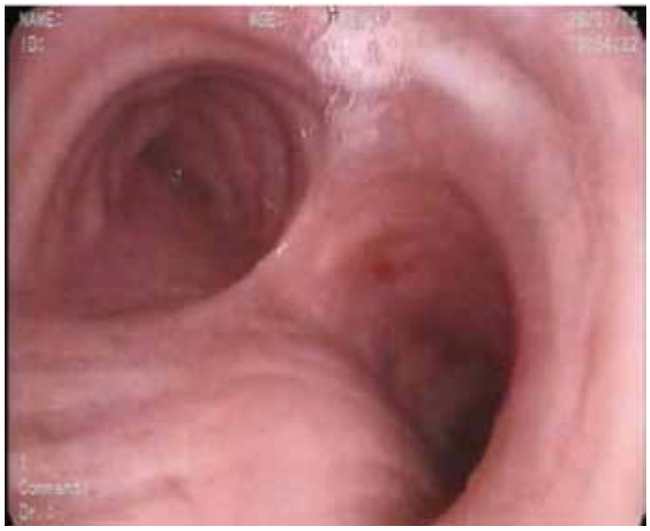

Figure 3: FOB showing a normal carina, right main bronchus and a narrowed left main bronchus

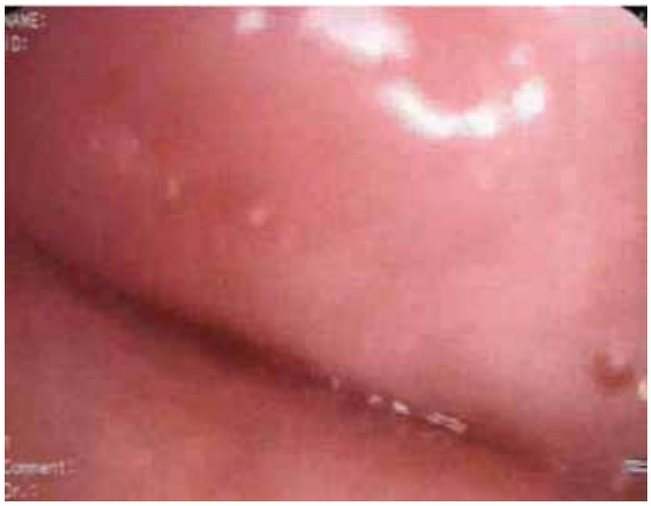

Figure 4: Distally narrowed and slit like left main bronchus

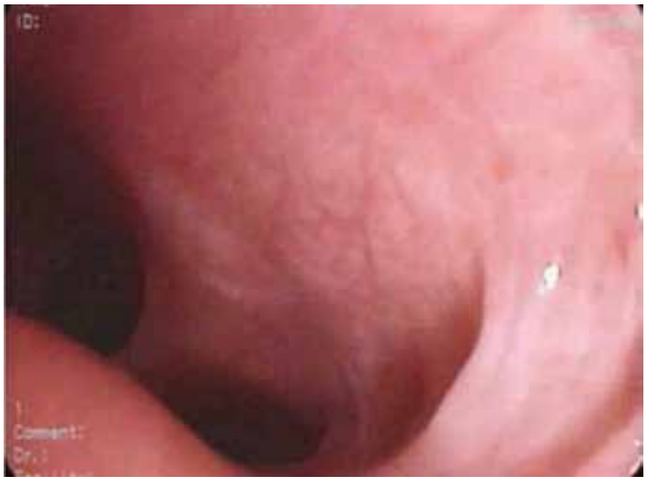

Figure 5: Left lobar bronchi ending in a distal blind pouch

DISCUSSION

Developmental anomalies of lung have been divided into different groups by Boyden and Monaldi. Boyden classified as (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. (1). Monaldi classified lung mal development as 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 sub segmental bronchi and small segment of the corresponding lobe. In pulmonary hypoplasia, the lung consists of incompletely developed lung parenchyma connected to bronchi that may be underdeveloped, depending on the timing of the insult. Besides disturbances of the Broncho-pulmonary vasculature, there is a high incidence, approximately 50-85%, of associated congenital anomalies such as cardiac, gastrointestinal, genitourinary, and skeletal malformations.(2) Pulmonary hypoplasia may be primary, but it is usually secondary, due to reductions in intra-thoracic space, fetal breathing movements, or amniotic fluid volume.

The cause of primary pulmonary hypoplasia has not been identified, although a few case reports suggest a possible familial autosomal recessive inheritance. Various animal studies have shown that mutations involving thyroid transcription factor TTF-1, GATA 4 factors, hepatocyte nuclear factor HNF310, epidermal growth factor and its receptor, EGFR, mitogen-activated protein [MAP] kinase result in congenital diaphragmatic hernia with resultant pulmonary hypoplasia.(3, 4, 5) Causes of secondary pulmonary hypoplasia include conditions that can result in small fetal thoracic volume, prolonged oligo-hydramnios, decreased fetal breathing movements, early rupture of membranes at 15-28 weeks gestation, longer latent period before delivery, decreased fetal breathing, decreased pulmonary perfusion, congenital heart diseases with poor pulmonary blood flow and trisomies 18, 13, 21(6, 7). Incidence of pulmonary hypoplasia ranges from 9–11 per 10,000 live births and 14 per 10,000 births (8) without sex predilection. 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. Pulmonary hypoplasia is usually unilateral but is occasionally seen bilaterally. The volume of the chest is reduced on the affected side, the affected lung is small, and the mediastinum is shifted toward the side of the hypoplastic lung (9). There is also an increased ipsilateral mediastinal shift during inspiration because of increased volume of the contralateral lung. CT scan usually show an abnormally shaped thorax, volume reduction on the affected side, compensatory over-inflation of the opposite side, ipsilateral mediastinal shift, hypoplastic airways, and rib abnormalities. CT angiogram may show hypoplastic pulmonary arteries. Antenatal diagnosis of pulmonary hypoplasia can be made reliably by a combination of ultra sound and Magnetic resonance imaging(MRI) . Factors used for prediction are a) the fetal lung volume, b) the relative lung volume, and c) the ratio of lung volume to body weight (which is the most widely used ratio). Ultrasonography can be used to assess factors contributing to pulmonary hypoplasia, such as oligo-hydraminos, renal agenesis, renal obstruction, renal cysts, renal dysplasia, diaphragmatic hernia, thoracic and abdominal masses, and pleural effusion. (10,11) Associated congenital malformations can also be seen. Perfusion scan is used to differentiate hypoplasia from Sweyer-James syndrome. (12)

Treatment of hypoplasia involves antenatal corticosteroids administration to enhance fetal lung maturation in pregnancies less than 34 weeks of gestation. Oligohydraminos secondary to renal agenesis should raise the possibility of pulmonary hypoplasia. Serial amnio infusions are increasingly used in cases of preterm rupture of membranes at less than 32 weeks gestation. Postnatally respiratory support is needed for the infant. Surfactant administration in pulmonary hypoplasia secondary to Congenital diaphragmatic hernia has been shown to be efficacious in improving oxygenation, decreasing the need for mechanical ventilation and improving the survival rate if prophylactically administered at birth. Intrauterine vesico-amniotic shunts and endoscopic ablation of posterior urethral valves are other techniques that are currently used in fetuses with urinary tract obstruction and pulmonary hypoplasia. Percutaneous fetal endo-luminal tracheal occlusion (FETO) with a balloon, inserted at 26-28 weeks' gestation, can be considered for infants with isolated CDH with poor prognosis. 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, and influenza infections are recommended. (2,13, 14,15).

CONCLUSION

Though pulmonary hypoplasia is usually congenital, isolated pulmonary hypoplasia may present in adults as highlighted in our case. Our patient had no significant respiratory illness and hence it should be remembered that pulmonary hypoplasia should strongly be consid-

ered in a differential diagnosis in a unilateral whiteout lung and unilateral lung bronchiectasis even when patient has no significant respiratory complaints. We also recommend that such patients be thoroughly evaluated to rule out other co-existing anomalies.

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