International Journal of Health Sciences and Research
www.ijhsr.org ISSN: 2249-9571

Case Report

Primary Unilateral Pulmonary Hypoplasia of Lung in Adult: Case Report

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Received: 20/09//2014 *Revised: 07/10/2014* *Accepted: 13/10/2014*

ABSTRACT

Pulmonary hypoplasia is a bronchopulmonary foregut anomaly characterized by a decrease in the number or size of airways, vessels and alveoli. Unilateral pulmonary hypoplasia is a rare congenital anomaly. A young pregnant woman with 30 weeks gestation presented with severe breathlessness and died soon after admission. Autopsy reveals left pulmonary hypoplasia with interstitial pneumonitis.
Key Words: Lung, Hypoplasia, Autopsy, Pulmonary.

INTRODUCTION

Congenital malformations of the lung are extremely rare. Pulmonary hypoplasia is a bronchopulmonary foregut anomaly characterized by a decrease in the number or size of airways, vessels and alveoli. [1] Under development of the alveolar tissue results in a small fibrotic and non functioning lung.

Unilateral pulmonary hypoplasia is a rare congenital anomaly. Although its exact prevalence is not well known, it is estimated to be seen in about 1 - 2 out of every 12,000 births. [2]

The incidence of unilateral pulmonary agenesis has been estimated approximately one in 15,000 live births, [3] or between 0.0034% and 0.0097%. [4,5]

At autopsy, pulmonary hypoplasia is one of the important causes of death in newborns and infants. Pulmonary hypoplasia is rare in adults. [6] Severity of the clinical symptoms and timing of their occurrence is related to the degree of hypoplasia. [2]

This condition is frequently associated with other congenital anomalies. [7] Common developmental anomalies associated with pulmonary hypoplasia involve urinary system, [8] diaphragm, [9] cardiovascular system (Tetralogy of fallot), [10] and central nervous system (anencephaly and hydroencephaly). [11]

Hypoplasia of the lung is classified as primary (idiopathic) and secondary: [1]

Primary Pulmonary Hypoplasia is an intrinsic defect in the process of lung development. The etiology is not completely known but vitamin A deficiency, viral agents, or genetic factors are described.

In ***Secondary Pulmonary Hypoplasia***, several mechanisms like decreased hemi thoracic volume, decreased pulmonary vascular perfusion, decreased fetal respiratory movement and decreased lung fields are implicated. Most common cause is

a space occupying mass like congenital diaphragmatic hernia. Thoracic neuroblastoma, sequestrated lung also contribute to the deformity.

In the literature, the most described cases were neonates and infants.

In this article, a case report of a patient with this anomaly is presented who remained asymptomatic until young adult age, which is a rare clinical presentation of pulmonary hypoplasia.

CASE HISTORY

A 26 year primigravida with 30 weeks amenorrhea, presented with breathlessness. It was not associated with any other complaints. She was admitted to casualty. Breathlessness was grade IV. Patient died within 2 hours of admission.

Clinical autopsy was performed to know the cause of death. On autopsy, surprisingly found the left sided lung was hypoplastic with few areas of consolidation [Fig. 1 - 4].

On cut surface, Left lung showed areas of fibrosis and consolidation [Fig. 4].

No other anomaly / abnormality were found. Case was further evaluated by histopathological examination.

Histopathological examination of left lung showed the features of hypoplasia with interstitial pneumonitis [Fig. 5 & 6].

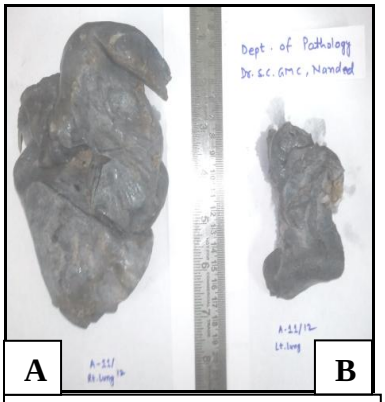

Fig. 1. Gross of Right Lung [A] & Left Lung [B]. Left lung appears to be small & hypoplastic.

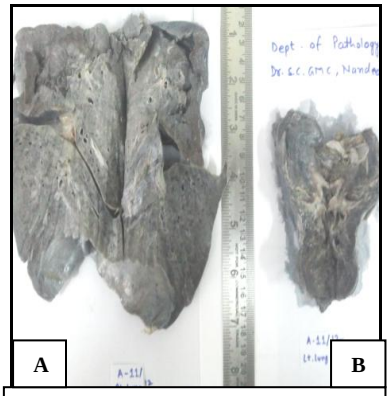

Fig. 2. Cut surfaces of Right Lung [A] & Left Lung [B]. Right lung appears pale & spongy. Left lung shows areas of fibrosis & consolidation.

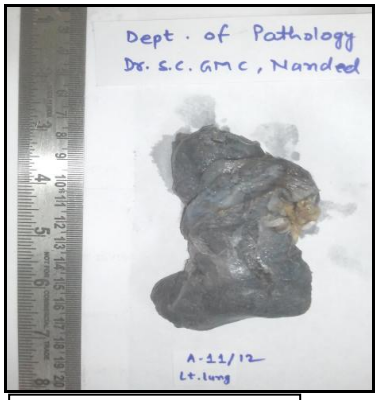

Fig. 3. Gross of Left Lung .

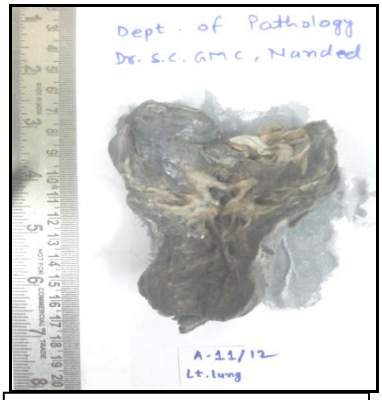

Fig. 4.Cut surface of Left Lung.

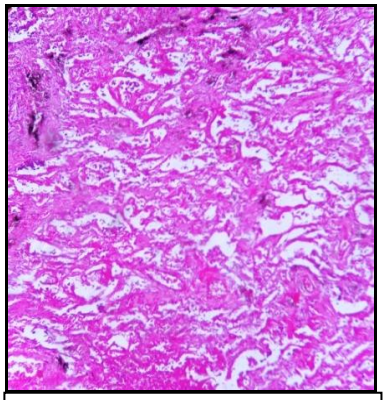

Fig. 5. Shows small & decreased number of aveoli along with decreased number of blood vessels. H &E, 10X.

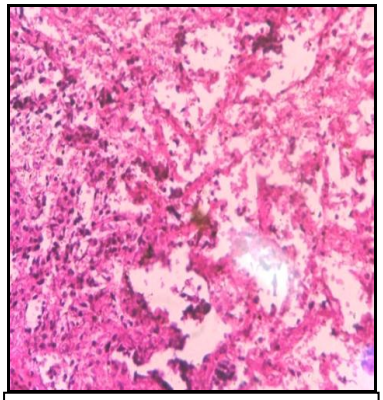

Fig. 6. Shows interstitium infiltrated with inflammatory infiltrate with fibrotic areas. H &E, 10X.

DISCUSSION

Development of bronchial tree takes place at about 26th to 31st day of intrauterine life.[2] Pulmonary hypoplasia is characterized by reduced and/or less developed lung tissue to varying degrees.

Pulmonary hypoplasia can be identified pathologically by the radial alveolar count and the ratio of lung to body weight. [6]

When pathologic specimens of the patients with pulmonary hypoplasia were examined, it was found that ratio of lung weight/body weight of the patients was less than 0.012. [12]

Monaldi categorizes developmental disorders of the lung to four categories. [13] Group I: No bifurcation of trachea; Group II: Only rudimentary main bronchus; Group III: Uncompleted development after bifurcation of the main bronchus; Group IV: Incomplete development of small segment and subsegmental bronchi of the corresponding lobe.

According to Boyden, developmental disorders are seen in three different extents [14] : 1) Agenesis (complete absence of the lung tissue), 2) Aplasia (no lung tissue, but there is a rudimentary bronchus), 3) Hypoplasia (all lung tissues exist but under-developed).

The patient presented in this article is in group IV according to Monaldi classification and is of third degree according to Boyden.

During pregnancy, the cardiac output is increased to meet the additional oxygen required due to increased metabolic activity. In order to increased oxygenation demand of body, lungs has to perform more work than in non-pregnant state. In our case, lungs can't satisfy the demands of increased oxygenation due to unilateral pulmonary hypoplasia. Hence, the patient presented with the features of respiratory failure.

CONCLUSION

Unilateral hypoplasia is a congenital anomaly causing a wide spectrum of clinical manifestations from mild, non-specific findings to severe respiratory symptoms. Our case remains asymptomatic until the age of young adult, which is a rare clinical presentation of pulmonary hypoplasia. Although it is seen rarely, pulmonary hypoplasia should always be kept in mind.

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How to cite this article: Hanmante RD, Suklikar SD, Deshpande SA et. al. Primary unilateral pulmonary hypoplasia of lung in adult: case report. Int J Health Sci Res. 2014;4(11):278-281.

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