Case Report

Congenital Agenesis of Left Lung: A Rare Case

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Abstract
Agenesis of lung is a rare congenital disorder, the etiology of which is not clearly known. Here we report a case of 2 years 10 months old male child who presented with repeated attack of respiratory tract infection since birth & at first the child was diagnosed, as a case of collapse-consolidation of left lung. The child was, later diagnosed as having left lung aplasia with congenital heart diseases.

Key Words: Opaque hemithorax, Aplasia of lung, Agenesis of lung, Congenital anomaly.

Introduction
Agenesis of the lung is an extremely rare congenital anomaly representing failure of development of the primitive lung bud. The condition was first discovered accidentally at the autopsy of an adult female in 1673, by De Pozze¹. This condition is generally diagnosed in childhood. However, patients without any co morbid anomalies or patients with a mild form of this disease can reach adulthood².

Case report
A 2 years, 10 months old male baby referred to Radiology & Imaging Department of National Heart Foundation Hospital & Research Institute for CT scan of chest with contrast. According to patient father the baby is suffering from repeated attack of respiratory tract infection since birth. No history of consanguineous marriage among the parents & other siblings are normal. Their socio-economic status is poor. Haematological report revealed Hb: 11.4 gm/dl; ESR: 28mm, WBC: 11,200/Cu mm, Polymorphs: 46%; lymphocytes: 50%; Monocytes: 02%; Eosinophils: 02; Basophils: 00. Serum creatinine is 0.6 mg/dl. Chest X-ray A/P view revealed opaque left hemithorax & hyper-inflated right lung (Fig-1). Echocardiogram revealed secundum type of ASD (about 9 mm’12 mm’), PDA (about 4mm’4mm), Mild TR & PAH good LV & RV systolic function. With the above history & investigation findings CT scan of chest with contrast was done for further evaluation. CT scan of chest shows complete absence of left lung parenchyma, right lung is hyper-inflated & herniated toward left. Mediastinum shifted towards left (Fig-2). Absence of pulmonary vessels (Fig-3) with rudimentary bronchus which appear as blunted tube (Fig--4). Secundum type atrial septal defect is seen which is measuring about 16.0mm’11.3mm (Fig-5).

Fig-1 (Chest A/P View)

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Fig-2 (Complete absence of left lung)
Introduction

Congenital agenesis of the lung is an extremely rare congenital anomaly representing failure of development of the primitive lung bud. The condition was first discovered accidentally at the stage of development of the primitive lung bud, pulmonary agenesis is classified into three categories.

Type – 1 (Agenesis): Complete absence of lung & bronchus & no vascular supply to the affected side.

Type – 2 (Aplasia): Rudimentary bronchus with complete absence of pulmonary parenchyma.

Type – 3 (Hypo-plasia): Presence of variable amounts of bronchial tree, pulmonary parenchyma & supporting vasculature.

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Discussion

Pulmonary agenesis means undeveloped pulmonary vessels, bronchi & parenchyma. It can be unilateral or bilateral3. Schneider4 classified agenesis into three groups, which has been sub-sequently modified by Boyden5. Depending upon the stage of development of the primitive lung bud, pulmonary agenesis is classified into three categories.

Type – 1 (Agenesis): Complete absence of lung & bronchus & no vascular supply to the affected side.

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Type – 3 (Hypo-plasia): Presence of variable amounts of bronchial tree, pulmonary parenchyma & supporting vasculature.

Our patient would classify as type-2 pulmonary agenesis (pulmonary aplasia). As agenesis & aplasia are difficult to
distinguish from each other clinically & pathologically, they are often used in the same sense.1,6 The onset of symptoms in pulmonary agenesis is remarkable variable. In many cases, presence of this anomaly usually comes to light during infancy because of recurrent chest infections, cardio-pulmonary insufficiency or due to associated congenital anomalies. However, patients with one lung have been reported to survive well into adulthood without much complaint. The oldest patient cited by Oyamada et al7 was 72 years old. The present cases are few of the older cases of aplasia reported in the literature. The exact aetiology of this condition is unknown although genetic factors, viral agents & dietary deficiency of vitamin A during pregnancy have been implicated.8 Left sided agenesis is more common & these subjects have a longer life expectancy than those with right sided agenesis.9 This is probably due to excessive mediastinal shift & malrotation of carina in right sided agenesis which hinders proper drainage of the functioning & increases chances of respiratory infections. It’s incidence in males & females are about the same.9 Hypo-plasia & aplasia are often observed together with other malformations such as diaphragmatic defects, kidneys anomalies, extra-pulmonary sequestration, muscle or skeleton system defects. Nearly one third of the patient’s have congenital heart diseases. Although the most common one is the atrial septal defect, ventricular septal defect, patent ductus arteriosis or aorta coarctation can also be observed.10 In our case ASD, Small PDA, Mild TR & PAH are associated. Pulmonary aplasia (agenesis) can causes a predisposition to infections. In aplasia, secretions not cleared from the rudimentary bronchus and / or comorbid bronchiectasis may partially be responsible for the risk of infection.29. Thus in cases of repeated chest infections, with opacification of right hemi-thorax & herniation of lung to the affected side, this rare entity must be kept in mind. Asymptomatic cases do not require any treatment, if there are no additional anomalies. However this entity carries a high risk in any surgery because of low respiratory reserve in the patient. Bilateral lung agenesis is life threatening & about 50% patient’s with unilateral lung agenesis die within 5 years of life.11

Diagnosis of pulmonary agenesis is made on basis of chest skiagram, bronchoscopy, broncho-graphy, CT scan of chest with contrast, angiography & magnetic resonance imaging. With the advent of CT scan, other procedures having significant risk have become unnecessary.12 Radiographic features of this patient are – on chest X-ray it can present as an opaque hemi-thorax with ipsilateral mediastinal shift & contra-lateral lung hyper-inflation.13 CT scan will confirm the absence of lung parenchyma & mediastinal ipsi-lateral shift, ipsi-lateral absence of pulmonary artery & ipsilateral bronchus remnant. It may also show other cardiac congenital malformation.

References