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Case Study

LEFT LUNG APLASIA WITH OTHER CONGENITAL ABNORMALITIES: A CASE STUDY

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Congenital lung anomalies are categorized as pulmonary agenesis, aplasia and hypoplasia with distinct clinical implications. A baby came with repeated infection of thorax. A detailed evaluation including computed tomography of the thorax and fiberoptic bronchoscopy led to a diagnosis of left lung aplasia. He also had wheezing dyspnea, which was confirmed as bronchial asthma. Congenital lung defects with associated asthma was reported only twice till date. A high index of suspicion is required to recognize such a patient.

Keywords: Aplasia, Congenital anomolies, Skin Tags, Dermoids

INTRODUCTION

Depending upon the stage of development of the primitive lung bud, pulmonary agenesis is classified into three categories:

a. Agenesis—Complete absence of lung and bronchus and no vascular supply to the affected side.

b. Aplasia—Rudimentary bronchus with complete absence of pulmonary parenchyma.

c. Hypoplasia—Presence of variable amounts of bronchial tree, pulmonary parenchyma and supporting vasculature.

Agenesis of the lung is an extremely rare congenital anomaly representing failure of development of the primitive lung bud. Aplasia may be unilateral or bilateral. In aplasia there is rudimentary airway stump but absence of distal lung. Contralateral lung is enlarged with greater number of alveoli. Individual with aplasia may present with recurrent infection, dyspnea, pooling of bronchial secretion.

This condition was first described by De Pozze (1960), who discovered it accidently at the atopsy of an adult female in 1673. Munch Meyer (1944) first diagnosed unilateral agenesis of the lung clinically in 1885. From India, the first case was reported by Muhamed in 1923, of a left sided pulmonary agenesis in a medicolegal autopsy. Subsequently a few more case reports have appeared and by 1977, over 200 cases of under development of the lung have been reported.

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We are describing a case with respiratory complaints who had aplasia of left lung with associated congenital abnormalities.

CASE REPORT

A nine month old child admitted to Dept. of Paediatrics, KLES J N M C , Belgaum. Child presented with history of recurrent chest infection, cough with expectoration. On Examination skin tag seen in front of Right Ear, dermoid in right eye. Systemic examination reveals smaller left hemithorax, trachea shifted to left side, movements decreased and percursion dull on left chest, no air entry in left chest.

X-Ray chest shows total absence of aerated lung in left Hemithorax, Approximation of rib, elevation of ipsilateral diaphragm and shift of mediastrinum, right lung is over inflated. CT Scan shows Congenital Aplasia of the left lung with Ipsilateral Mediastinal (cardia) shift.

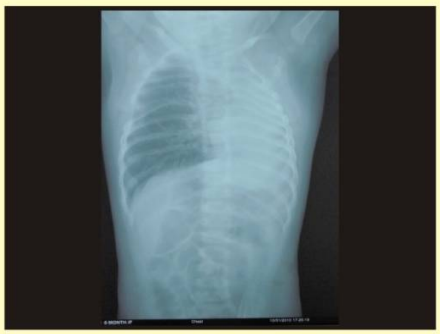
DISCUSSION

Development of lung takes place by 26 to 31 day of intra uterine life. Cause of aplasia can be idiopathic or environmental. Incidence is 1 in 12,000 live birth. Pathogenesis is decreased vascular perfusion, decreased lung fluid in intra uterine life.

Our patient present with respiratory complaints and diagnosis was suspected from routine radiological examination of chest, where possibilities of atelectasis or agenesis of the left lung were considered. The diagnosis was confirmed by CT scan of chest.

Figure 1: A. Dermoid in Right Eye B. Skin Tag in Front of Right Ear

A B



X-Ray Chest : Total Absence of aerated lung in left Hemithorax, Approximation of rib, elevation of ipsilateral diaphragm and shift of mediastinum, right lung is over inflated

Congenital Aplasia Of The Left Lung With Ipsilateral Mediastinal (cardia) Shift.

The onset of symptoms in pulmonary agenesis is remarkably variable. In many cases, presence of this anomaly usually comes to light during infancy because of recurrent chest infections, cardiopul-monary insufficiency or due to associated congenital anomalies. However, patients with one lung have been reported to survive well into adulthood without much complaints and oldest patient cited by Oyamada *et al*. (1953) was 72 years old.

Nearly 50% cases of pulmonary agenesis have associated congenital defects (Sbokos and McMillan, 1977), which usually involve cardiovascular, skeletal, gastrointestinal and genitourinary system. Here in this case patient presented with anomalies in eye and ear, which is found rarely. Finding in this case correlate with Treacher Collins syndrome.

The exact aetiology of this condition is unknown though genetic factors, viral agents and dietary deficiency of Vitamin A during pregnancy have been implicated Sbokos and McMillan (1977). Left sided agenesis is more common and the subjects have a longer life expectancy than those with right sided agenesis Oymada *et al*. (1953). This is probably due to excessive mediastinal shift and malrotation of carina in right sided agenesis which hinders proper drainage of the functioning lung and increases chances of respiratory infections.

In this case we have given symptomatic treatment to treat recurrent chest infection with antibiotics. Prophylactic treatment given in the form of pnemococcus and influenza vaccines. The prognosis in such cases depends upon the functional integrity of the remaining lung as well as upon the presence of associated anomalies.

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