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Case report (Pages: 16319-16323)

Unilateral Lung Aplasia with Absent Right Pulmonary Artery in an Infant: A Case Study

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Abstract

Unilateral Pulmonary aplasia with absent right pulmonary artery is a rare disorder, caused by the developmental arrest of primitive lung during embryonic life. It's a complete absence of lung parenchyma vasculature and its main bronchus. The most common variant consists of carina and main stump bronchial stump with the absence of distal lung. It is frequently associated with many other congenital disorders like VACTERL syndrome (V-vertebral anomalies, A-anal atresia, C-cardiovascular anomaly, T- tracheosophageal fistula, E- esophageal atresia R-renal anomalies L- limb defects) and cardiac anomaly. These patients usually present with respiratory distress. Here we are presenting a case of a 2-month-old male infant suffering from unilateral lung aplasia with absent right pulmonary artery.

Key Words: Congenital Malformation, Infant Lung Aplasia, Pulmonary Artery.

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1- INTRODUCTION

Pulmonary aplasia is a rare and a potentially lethal condition. It could be unilateral or bilateral, of which bilateral is lethal. Schinder recognised 3 major types of pulmonary defects.

(i) True agenesis, with no trace of bronchus and vascular supply on the affected side.

(ii) Rudimentary bronchus with no pulmonary tissue.

(iii) Bronchus with varying amounts of pulmonary alveolar tissue.

In fifty percent of the population, death occurs during the first 5 years of life generally from infection. Unilateral pulmonary aplasia with absent right pulmonary artery is a rare disorder. They usually present at early life with complaints of increased work of breathing and recurrent infection.

2- CASE REPORT

A 2-month-old male infant was brought with complaints of fever, cough and cold for 3 days. There was no history of suck-rest-suck cycle, cyanosis or any similar history in the past. He was born to a primigravida with at term gestation with no perinatal complications. Developmental milestones were normal. The child had tachypnea (respiratory rate- 56 cycles per minute) tachycardia (160 beats per minute), saturation of 95% on oxygen by prongs with subcostal retraction. On inspection, he had decreased movements on the right side of the chest. There was a dull percussion note on the whole right side chest, with absent air entry on right side and crepitations over left side of the chest. Other system examinations were in normal limits. Chest xray showed haziness over the right side (**Fig. 1**).

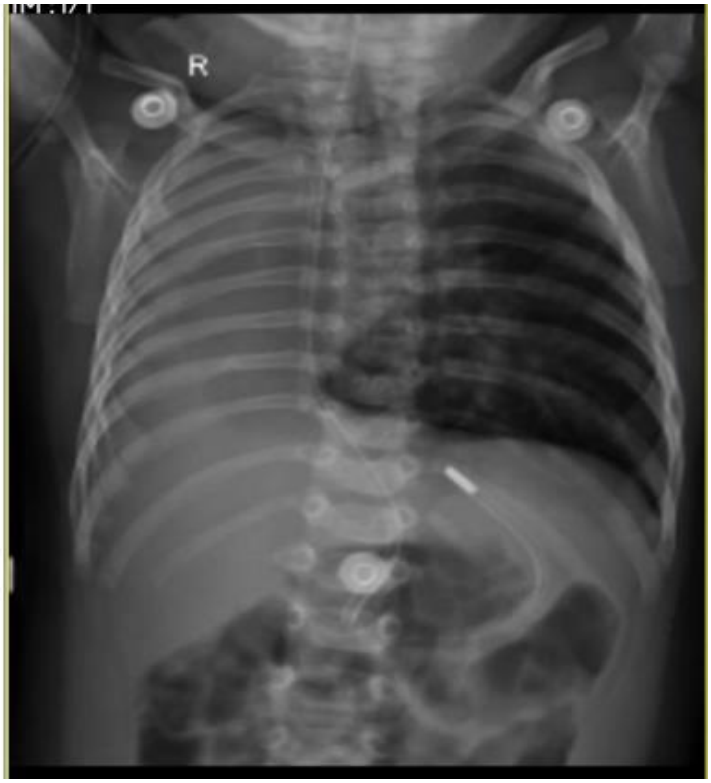

Fig. 1: chest x-ray suggestive of homogenous opacity in right hemithorax with non-visualization of bronchovascular marking in peripherals. Hyperlucent left hemithorax with bronchovascular marking with hilum noted with midline shift.

On investigation CBC revealed hemoglobin – 12gm%, total leucocyte count – 10,000 WBCs per microliter, platelet – 4.59 lakhs and hematocrit – 36.4. Liver and kidney function were in normal limits and CRP of 1.21 which was in normal range. ECG was normal. Echocardiography was suggestive of absent right sided pulmonary artery, and ejection fraction of 65% with normal biventricular function. CT pulmonary angiogram was suggestive of hypoplastic right lung with absent right pulmonary artery (**Fig. 2** and **3**).

The patient was given antibiotics for 10 days, and was discharged.

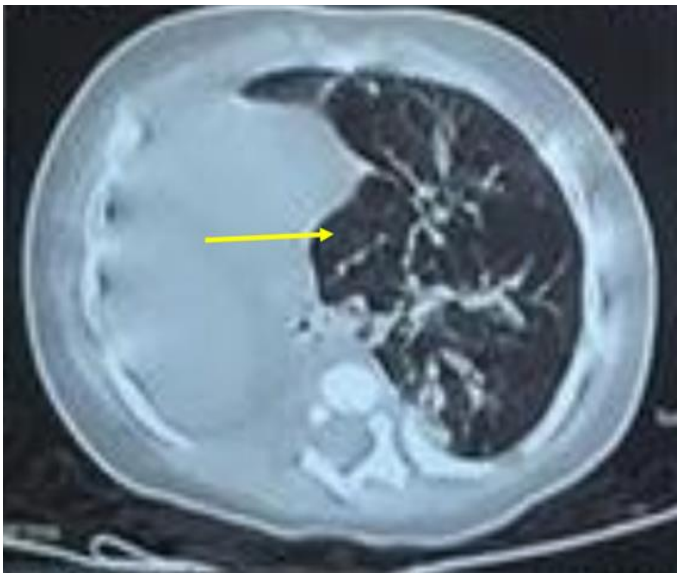

Fig. 2: Lung window showing left side hypertrophic lung with right hypoplastic (arrow) just anterior to vertebra. Mediastinal shift to right side with crowding of ribs.

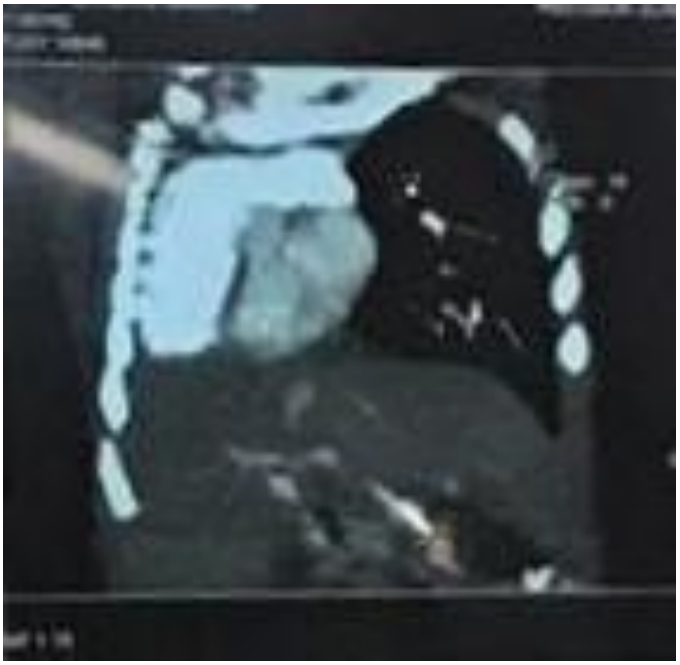

Fig. 3: CT pulm angiography showing absent right lung

3- DISCUSSION

This case of unilateral aplasia of lung was first described by De pozze (4). In India the first case was reported by Mummhad et al. pulmonary aplasia is a rare disorder with the incidence of 0.0034% to 0.0097% (2). Genetic and environmental and mechanical factors have been bearing etiology. They are generally sporadic with only few reports as an autosomal recessive. They are seen equally in both sexes. There is a high incidence of association with cardiac and VACTRL defects (V-vertebral anomalies, A-anal atresia, C- cardiovascular anomaly, T- tracheosophageal fistula, E- esophageal atresia R-renal anomalies L – limb defects). Pulmonary aplasia or agenesis is a result of embryological defect in 5^{th} week of intrauterine life. Diagnosis should be suspected when respiratory difficulty occurs with tracheal deviation, in the presence of a clinically symmetric chest and chest X-ray suggestive of massive atelectasis with mediastinal shift. No treatment is required for asymptomatic patients. Patients with left out stump might need surgical removal as it may act as the source of infection. Those with right lung agenesis have higher mortality compared to left lung due to the compression of the tracheobronchial tree, shifting the normally mid-thoracic structures into the right chest. If a patient survives in his first years of life without any major infection, a normal span is expected. Most common anomalies include unilateral agenesis, hypoplasia and aplasia with a non-specific finding and only one third of them are diagnosed (6).

Absent right pulmonary artery is a rare disorder. The diagnosis of absent pulmonary artery is confirmed by history, clinical evaluation and imaging. Most patients present with congestive heart failure and pulmonary hypertension, but few may be asymptomatic. The incidence of pulmonary hypertension in absent pulmonary arteries was about 19 to 44percent (4). Chest xray is suggestive of absent hilar shadow, reduced pulmonary markings, mediastinal shift to the affected side and contralateral hyperinflation. ECG is likely to be normal in patients with absent pulmonary arteries unless it is associated with pulmonary hypertension where we can see right ventricular dominance. Any infant with an unexplained pulmonary hypertension should be evaluated for the absent pulmonary artery. Infants with isolated absent pulmonary arteries usually present with pulmonary hypertension. Pool et al. documented medial hypertrophy of the pulmonary artery of the normal lung (opposite to the affected side) in 53% of the 17 patients (3). The reason for pulmonary hypertension was thought to be because of insufficient elasticity of the normal pulmonary artery receiving cardiac output. Most patients might not require treatment and have to be kept in for regular follow up. In patients with heart failure early surgical repair might help and be also effective in preserving the lung function if any is left. If intrapulmonary arteries are present, a primary anastomosis can be done to the central pulmonary artery. If the intrapulmonary artery is small, a modified Blalock- shunt can help. Krammoh et al. described their two-stage therapeutic approach in three patients (5). The first stage was PDA stenting, followed by surgical anastomosis in the second stage. The patients required anticoagulation therapy after the ductal stenting. They reported good outcomes in three cases during their midterm follow-up.

4- CONCLUSION

Unilateral lung aplasia with absent right pulmonary artery is a rare entity. Most of the children present with respiratory distress, cough and cold. A differential vascularity on chest x ray gives an index of suspicion. CT pulmonary angiography can confirm the

echocardiographic diagnosis and help in tracing the pulmonary artery. Most of them require symptomatic management for lower respiratory tract infection and some might need treatments for cardiac failure.

5- CONFLICT OF INTEREST

None.

6- FUNDING

None.

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