RESEARCH PAPER

Pulmonary agenesis: A case report with review of literature
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ABSTRACT
Congenital pulmonary agenesis is a rare condition which poses diagnostic difficulties. More than 50% of these patients die before the first five years of life. Few patients may remain asymptomatic throughout their life. We report a case of a two month old male infant with right pulmonary agenesis who presented with severe respiratory distress in whom a diagnosis was made following computed tomography of the chest.

Keywords: right lung agenesis, atrial septal defect, CT scan of chest

INTRODUCTION
Pulmonary agenesis is a rare condition with a reported incidence of 1:15,000 autopsies. It can be unilateral or bilateral. Bilateral pulmonary agenesis is extremely rare, and is incompatible with extra uterine life, first described by Morgagni in 1955.

We report a case of a two-month-old infant who was recently managed at Owaisi Hospital and Research Centre a part of Deccan College of Medical Sciences in India.

CASE REPORT
A two month old male infant presented to the hospital with a cough, severe respiratory distress, chest retractions and cyanosis.

The mother gave a history of breast feeding prior to admission and the baby had sudden respiratory difficulty and severe cough following the feed.

On examination, the infant had tachypnea with a respiratory rate of more than 70 breaths per minute, chest retractions, and cyanosis. Air entry was absent on the right side, while wheezing was noted on the left side of the chest.
Investigations revealed haemoglobin 9.2 gm/dl, total leucocyte count 13,000/mm³ (neutrophil 36%/lymphocyte 48%, eosinophil 2%, monocyte 4%). Laboratory parameters were within normal limits. ABG was done, pH 7.30, paO₂ 52, pCO₂ 30, HCO₃ 15 and saturation was 68% on RA.

Plain x-ray of the chest showed a homogeneously opaque right hemithorax with hyperlucency of left lung field with a slight shift of mediastinum to the right side (see Figure 1).

CT scan of the chest showed the absence of the right lung, right bronchus and pulmonary vasculature. The left lung was normal, hyperinflated with normal pulmonary vasculature (see Figure 2).

2D Echo was within the normal limits, except for a small ASD.

The condition of the child was too serious to permit any further investigation such as bronchoscopy and bronchogram. Treatment with intravenous fluids and antibiotic IV ceftriaxone (100 mg per kilo in two divided doses), aminoglycoside Amikacin (15 to 20 mg per kilo in two divided doses), and oxygen was given. Saturation was not maintained even after giving oxygen. The patient was intubated and put on ventilatory support for 48 hours.

The child continued to deteriorate and he expired inspite of ventilatory support.

DISCUSSION

Pulmonary agenesis was first reported by Depozze in 1673 as a finding from autopsy of a woman.¹

Lung agenesis is defined as complete absence of the lung and its bronchus.¹ In case of complete unilateral pulmonary agenesis, no pleural cavity can be found on the affected side. Both the lungs are equally affected, patients with agenesis of the left lung have better prognosis, where as agenesis of right lung carries a poorer prognosis.²

More than 50% of patients with pulmonary agenesis have other associated anomalies including PDA, VSD, ASD, narrowed trachea and tracheoesophageal fistula.³

In the absence of other malformations, unilateral pulmonary agenesis compatible with normal life,⁴ severe respiratory infection during infancy is common in these children and may lead to pneumonia and death.

Reasons attributed to signs of respiratory insufficiency in these babies may be due to kinking and compression of trachea, mediastinal shift and posterior curvature of trachea due to pressure by dislocation of aortic arch and truncus arteriosus.¹⁻³

Few cases have been reported in literature that were treated successfully by the technique of aortopexy⁶ and diaphragmatic translocation.⁷

Other surgical procedures such as subclavian artery reimplantation¹, tracheal stenting,⁵ slide tracheoplasty² and laser ablation of fibrotic tissue at the site of tracheal stenosis² are some of the procedures described in literature.

Figure 1. Plain x-ray of the chest showing right pulmonary agenesis with mediastinal shift and hyperinflated lung.

Figure 2. CT scan of the chest showing absence of the right lung, right bronchus and pulmonary vasculature with normal hyperinflated left lung.
In the present case the child was asymptomatic up to one and a half months of age, then presented with sudden severe respiratory distress most likely due to aspiration of the feed. Unlike other cases reported in literature, the present case is not associated with any major congenital abnormality except for a small ASD.

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