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CASE REPORT

Congenital agenesis of the lung: A case report and literature review

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ABSTRACT

Developmental malformations of the lung are common but complete agenesis of the lung is quite rare and usually the patients presented are in early childhood; though according to literature, a few cases have come to light even in the adult age group. A 10-month-old male child presented with a sudden onset of respiratory distress with opacity of the right hemithorax on X-ray, raising suspicion of foreign body bronchus. Subsequent CT scans and bronchoscopy revealed agenesis of the right lung. Patient was managed conservatively and his parents were counseled about the anomaly. The child is doing well and is in a regular follow-up with us. Agenesis of the lung should be suspected in children with recurrent respiratory distress with opacity of the hemithorax on X-ray and herniation of the opposite lung across the mediastinum.

***Keywords**: agenesis; lung; congenital; collapse; foreign body*

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Introduction

Agenesis of the lung is an extremely rare congenital anomaly representing developmental failure of the primitive lung bud. The incidence is 34 per 1,000,000 births[1]. De Pozze accidentally found agenesis during an autopsy in 1963, which was similarly reported by Muhamedin from India in 1923, also during an autopsy[2,3]. A history of recurrent chest infection is usually present during the first year of life. About 50% of the cases have associated congenital malformations of the cardiovascular, skeletal, gastrointestinal or genitourinary system[4].

Case report

A 10-month-old child was presented to the emergency room with sudden severe respiratory distress. There was mild fever with tachypnea and tachycardia with an oxygen saturation of 80%. No history suggesting accidental foreign body aspiration could be clearly established. There was a history of a few episodes of chest infections in the past few months for which treatment from a local physician was prescribed and the child stabilized with treatment. The present respiratory symptoms were of a sudden onset of a few hours' duration. On auscultation, breath sounds were absent on the right side. The chest X-ray revealed opacity in the right hemithorax with slight shifting of the trachea towards the right (**Figure 1**). A right lung collapse due to foreign body was suspected. The child was kept on oxygen, IV antibiotics and nebulization. High Resolution Computed Tomography (HRCT) of thorax revealed non-visualization of the right main bronchus, segmental bronchi and pulmonary vessels, with right lung agenesis and compensatory hypertrophy of the left lung with herniation of the left upper lobe across the midline. The left lung was otherwise normal (**Figures 2** and **3**). A fiber-optic bronchoscopy was done once the distress settled, which revealed a blind-ending right main bronchus, confirming the diagnosis of lung agenesis. The parents were counseled and the child was kept on conservative treatment for five days, discharged when respiration settled, with advice to follow-up regularly. He is currently doing well and is in a regular follow-up with us.

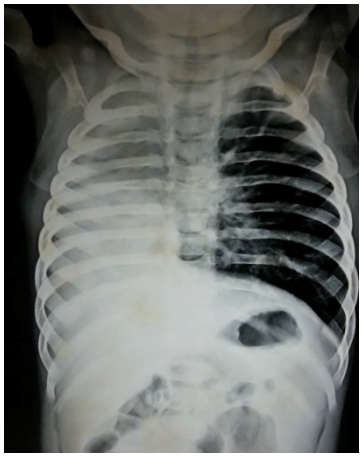

Figure 1. Plain X-ray of the chest showing opacity in the right lung field with shift of mediastinum to the right

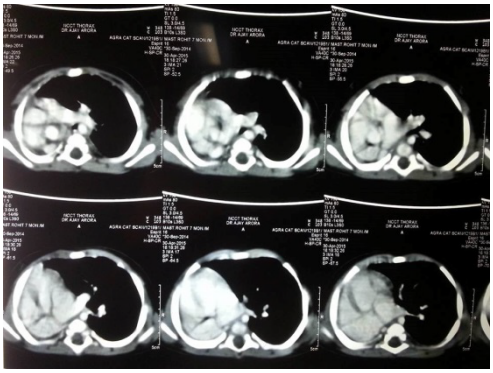

Figure 2. CT scan of the thorax showing absence of the right lung with hypertrophied left upper lobe crossing the midline

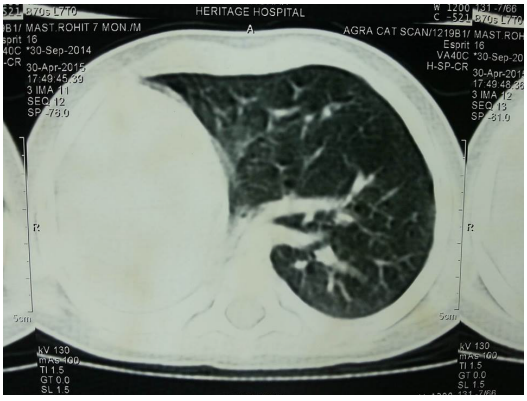

Figure 3. HRCT showing the absence of right-sided bronchi and vessels

Discussion

In fetal life, the lungs develop as an outpouching of the foregut called lung bud which divides into two: the right and left primary bronchial buds. During the fifth week, the right bud branches into three secondary bronchial buds while the left bud branches into two secondary bronchial buds. These give rise to the lobes of the lungs, three on the right and two on the left. Developmental anomalies in the primary lung bud division give rise to complete agenesis of the lung.

Mainly, three types of developmental anomalies are seen: agenesis, aplasia and hypoplasia. In aplasia, there is complete absence of both bronchus and lung, along with pulmonary vessels. In hypoplasia, rudimentary bronchus is present but the lung tissue and vessels fail to develop; all three structures are present but are hypoplastic[5].

The more severe the lesion, the earlier the presentation, hence agenesis usually manifests in infancy and early childhood. Clinically, the features of agenesis are similar to that of collapse of the lung, therefore it should be suspected if any child presented with respiratory distress and the chest X-ray suggests collapse and ipsilateral mediastinal shift and opposite lung herniation, especially if associated with other congenital anomalies[6]. Almost half of the cases are associated with other birth defects, mainly involving the urogenital, musculoskeletal and cardiac systems[7]. Fortunately, this child did not have any associated congenital defects.

Genetic predisposition, certain viral infections and deficiency of vitamin A during pregnancy are postulated as some of the causes behind this malformation[7]. Lung agenesis is more common on the left side, but our case had right-sided agenesis. Patients with left-sided agenesis have a better survival rate and longer life expectancy as excessive mediastinal shift and rotation of the carina in right agenesis causes less effective drainage of the left lung and increases the chances of recurrent respiratory infections[5].

There is no definitive treatment for lung agenesis other than regular follow-up to prevent and treat respiratory infections. Rarely, patients having longer bronchial stump may require surgical removal if it causes recurrent infections with no response to antibiotics. Associated malformations needed to be corrected as per protocol[5]. Overall, prognosis is guarded and depends on the severity of associated anomalies and status of the opposite lung.

Conclusion

The possibility of pulmonary agenesis must be considered strongly if clinical findings of recurrent respiratory infections and radiologic evidence of opaque hemithorax, bony and rib anomalies, along

with herniation of the normal lung to the affected side with no history of foreign body aspiration, especially if associated with congenital anomalies.

Author contributions

Rajesh Gupta is the primary treating surgeon and author of the manuscript. Anjali Gupta performed the CT scans and X-rays diagnosing the anomaly while Aradhana Singh helped in the preparation of the manuscript and literature research.

Conflict of interest

The authors declared no potential conflict of interest with respect to the research, authorship, and/or publication of this article.

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