Case Study

Right Upper Lobe Pulmonary Agenesis Type III - A Rare Case Report

Sakina S. Rajagara [1], Bela Verma [1], Amin R. Kaba [1]

Author's Affiliation:
1- Grant Government Medical College and J. J. Group of Hospitals, Mumbai.

Correspondence:
Sakina S. Rajagara, Email: sakmur118@gmail.com

Received on: 17-Jan-2021 Accepted for Publication: 15-Aug-2022

Right pulmonary lobar agenesis is a very rare anomaly with an incidence of 0.0034%. It is most often diagnosed as an incidentally on an abnormal chest radiograph. It is a congenital defect caused by unsuccessful growth of primitive lung bud. A 5-month-old, female child, with persistent pneumonia was referred to us with cough and respiratory distress. A contrast enhanced CT scan revealed hypoplasia of the right upper lobe of the lung. CT pulmonary angiography showed blind ending of the right upper lobe bronchus with hypoplastic upper lobe pulmonary artery i.e., type 3 pulmonary agenesis. She was treated with ionotropic and oxygen support and an adequate course of antibiotics for pneumonia. This case highlights the significance of a strong suspicion and ample amount of imaging for diagnosis of congenital lung anomalies. Early diagnosis of pulmonary agenesis is vital to lessen the incidence of fibrosis that can occur as a result of recurrent chest infections.

Key words: pulmonary agenesis, lobar hypoplasia, hypoplastic pulmonary artery

INTRODUCTION

Congenital lobar agenesis of lung is caused due to defective growth of primitive lung buds. There are only a handful of cases reported in India. Typically, agenesis of pulmonary lobe is almost always asymptomatic and may remain undiagnosed in the paediatric age group. The incidence of lobar agenesis is 0.0034% with no gender predisposition. There are only two Indian publications till date. A pulmonary computed tomography is regarded as the investigation of choice for diagnosis of lung agenesis. Here we report a rare case of lobar agenesis diagnosed at an earlier than usual age emphasizing the importance of a strong suspicion and ample imaging for the diagnosis of congenital lung anomalies.

CASE REPORT

A 5month old, female child was referred for further management of unresolving right sided pneumonia after visiting multiple doctors and hospitals. She presented to us with complaints of cough without expectoration and increased respiratory activity over a period of 2 months. There was no history of cardiac involvement. Birth was via normal vaginal delivery with a birth weight of 2.75kgs. Child was immunized for age. There was no developmental delay. On physical examination, the child had tachycardia, tachypnoea with subcostal retractions, bilateral crepitations on auscultation and hepatosplenomegaly. Child was given oxygen by nasal prongs, intravenous antibiotics and inotropic support.

Complete blood count was suggestive of leucocytosis with a positive C- reactive protein. The chest radiograph demonstrated right upper zone opacity with no air bronchogram and pull of trachea towards the right side

suggestive of reduced right lung volume with left upper lobar consolidation. Virtual bronchoscopy or a radiologically reconstructed imaging was done to look for an obstruction causing collapse but showed no luminal narrowing or foreign body. A contrast enhanced computed tomography (CT) demonstrated hypoplasia of right upper lobe along with right upper lobe bronchus with hyperinflation of right middle lobe with patchy consolidation in left upper lobe and a small blind ending of right upper lobar bronchus. CT Pulmonary angiography was suggestive of a hypoplastic upper lobe pulmonary artery. She was diagnosed as Type III pulmonary agenesis (hypoplasia).

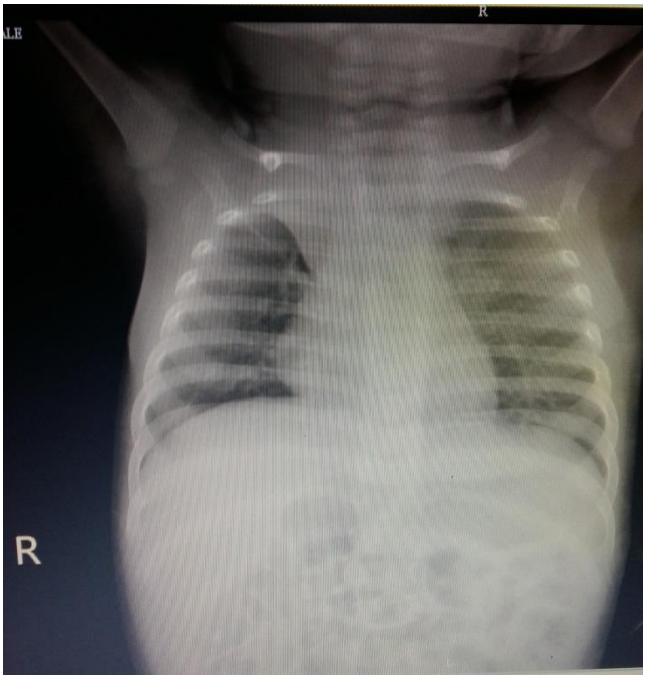

Fig 1: Chest radiograph showing right upper zone opacity with no evidence of air bronchogram within

Tests were done for tuberculosis, gastric lavage acid fast bacilli smear and GeneXpert, Mantoux test were negative. Her blood and sputum cultures were negative. Ultrasound of the abdomen was normal. 2D echocardiography was normal. Left sided pneumonia was treated with intravenous antibiotics. Gradually the tachypnoea and distress settled and the child was discharged with advice for strict follow up.

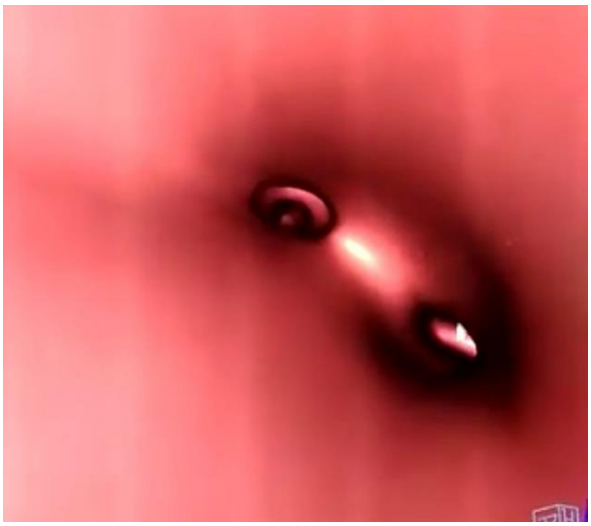

Fig 2: Bronchoscopic view of the blind end of the upper lobe bronchus

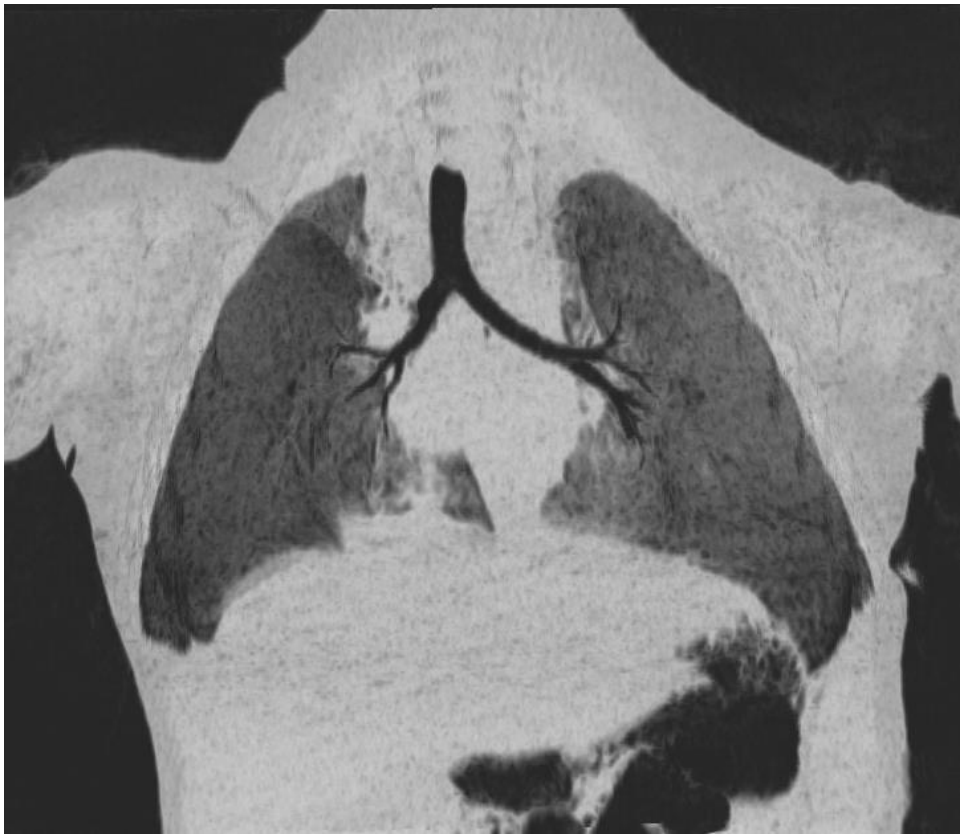

Fig 3: CT Pulmonary angiography showing blind ending of right upper lobar bronchus

DISCUSSION

Schneider classified pulmonary agenesis [1] which was then modified by Boyden [2] into three groups according to development of their primitive lung bud; (1) Type I which is called pulmonary agenesis including complete absence of unilateral lung parenchyma, its bronchus and vasculature, (2) Type II is called pulmonary aplasia with complete absence of unilateral lung with rudimentary bronchus and (3) Type III called pulmonary hypoplasia identified by partial existence of bronchial tree with few parts of unilateral pulmonary parenchyma and its vessels [3]. It is during the 4th and 5th weeks of foetal life that the pulmonary system develops. It has been observed that hypoplasia, aplasia or agenesis of pulmonary parenchyma is due to the failure of the bronchial analogue to divide proportionately between the two lungs with possible unusual blood flow in dorsal aortic arch. Almost twice the alveoli are seen in the contralateral lung in compensation [4]. Albeit, the exact aetiology of pulmonary agenesis is not known, but it's been implied that vitamin A deficiency during pregnancy, genetics, viral factors or iatrogenic agents might be involved [5]. This condition may coexist with other foetal anomalies of the heart, muscle, bones or gastrointestinal system. Bronchial anomalies are seen in less than 5% of general community with a preponderance to right upper bronchus abnormality [6].

Typically, agenesis of pulmonary lobe is almost always asymptomatic and may remain undiagnosed in the paediatric age group. In the right upper lobe agenesis of the lung, a chest radiograph shows the following features: a diminished right lung volume, shift of mediastinum to the right side and elevation of the right hemidiaphragm [7]. The differential diagnosis in our case comprised of right upper lobe atelectasis, lung

hypoplasia/agenesis, post-surgical i.e., lobectomy and paralysis of right hemidiaphragm. CT thorax is considered to be the best radiological study to determine lung anomalies and it's associated vascular abnormalities. The abnormalities of the bronchi and associated vascular structure can be delineated with three-dimensional reconstruction imaging. Pulmonary angiography is set aside for surgical procedures like embolization and revascularization.

Occasionally, the disease can be detected in antenatal life with the assistance of foetal ultrasound showing hyperechoic hemithorax, nonetheless, a definitive diagnosis may be tough, which can be confirmed on doing a magnetic resonance imaging [8]. Treatment is necessary for recurrent chest infections. Medical treatment includes bronchodilators and treatment of complications. The functionality of single existing lung along with other related anomalies determines the prognosis in such cases [3].

CONCLUSION

Considering that this abnormality can occur at any age, the possibility of pulmonary agenesis should be a differential diagnosis of all patients having reduced or completely absent breath sounds with decreased or no movement of unilateral chest wall and opaque hemithorax in plain chest x-ray. Confirmatory diagnostic imaging should be considered in such cases. An early detection of lung agenesis plays a key role in preventing development of lung fibrosis due to recurrent chest infections in the existing unilateral lung.

REFERENCES:

1. Schneider P, Schawatbe E. E. Die Morphologie der Missbildungen Des Menschen Under Thiere. Jena: Gustav Fischar. 1912;3 Part.2:817–822.
2. Boyden E. Developmental anomalies. Am J Surg, 1955;89:79–88. [PubMed].
3. Kisku KH, Panigrahi MK, Sudhakar R. Agenesis of lung-a report of two cases. Lung India: official organ of Indian chest. Society. 2008;25(1):28.
4. Yetim TD, Bayaroğullari H, Yalçin HP, Arıca V, Arıca SG. Congenital agenesis of the left lung: a rare case. J clin. imag scie. 2011:1.
5. Agarwal A, Maria A, Yadav D, Bagri N. Pulmonary agenesis with Dextrocardia and hypertrophic cardiomyopathy: first case report. J Neonatal Biol. 2014; 3(141):2167–0897.
6. Stovin PGI. 1985. Early lung development. Thorax 40:401–405.
7. Zack MS, and Eber E. 2001. Adult outcome of congenital lower respiratory tract malformations. Thorax 56:65–72.
8. Dembinski J, Kroll M, Lewin M, Winkler P. Unilaterale pulmonale Agenesie. Aplasie und Dysplasie Zeitschrift für Geburtshilfe und Neonatologie. 2009; 213(02):56–61.