Case Report

Lobar Agenesis of the Lung in a Newborn: A Rare Case Report

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Abstract

Lobar agenesis (LA) of the lung is a very uncommon congenital anomaly. Typically, the presentation is with respiratory symptoms, but the onset is highly variable. LA may be an isolated defect, but is often associated with congenital VACTERL anomalies. Here, we describe a neonate with agenesis of the middle and upper lobes of the right lung.

Keywords: Agenesis of upper lobe, congenital malformation of lung, Lobar agenesis

Introduction

Pulmonary agenesis is a very rare congenital abnormality which represents the unsuccessful growth of the primitive lung bud that leads to a spectrum of defects. Lobar agenesis is a rare and typically affects the right upper and middle lobes. It may remain asymptomatic and undetected during childhood or may present as late complication. We present a neonate with agenesis of the middle and upper lobes of the right lung.

Case Report

At the 9th day of life, a full-term, 3.1-kg male baby born by cesarean delivery was brought in with a complaint of breathing difficulty. He was born out of a nonconsanguineous marriage to a 27-year-old primigravida mother with no perinatal problems. The baby cried immediately after birth. On admission, the heart rate was 146/min, respiratory rate 56/min, and blood oxygen saturation level (SpO_2) on room air was 87%. There were no subcostal and intercostal retractions or peripheral cyanosis. His chest was normally shaped. The trachea was central. The apex beat was palpable on the right side at the fifth intercostal space in the anterior axillary line. The upper 2/3rd of the right side of the chest had a dull percussion sound, and the left side had no cardiac dullness. On the right side, breath sounds were reduced. Abdominal examination was within normal limits. Chest radiography showed opacification of the upper 2/3rd of the right hemithorax with a mediastinal shift toward the right [Figure 1]. The chest computed tomography (CT) scan [Figure 2] revealed the absence of the bronchus and pulmonary artery of the right upper and middle lobe with a shift of mediastinum toward the right side. Echo demonstrated small patent ductus arteriosus (PDA), small atrial septal defect, dilated right atrium, and right ventricle. The child was started on nasal oxygen. The baby clinically improved and was discharged in stable condition after 17 days. At 6 months, the infant displayed normal growth and development, and the lobar agenesis (LA) did not hinder the child's quality of life. He had no episode of lower respiratory tract infection in the last 6 months.

Discussion

LA of the lung is a rare anomaly, with an incidence between 34 and 97/million births.[1] It usually affects the right upper and middle lobes. LA can be life-long asymptomatic or can unexpectedly present with serious respiratory distress. Embryologically, it is due to a spectrum of failure in the development of the respiratory system from the foregut during the 4th–6th week of gestation.

Monaldi divided the maldevelopment of the lung into four groups: Group I: no bifurcation of the trachea; Group II: only rudimentary main bronchus; Group III: incomplete

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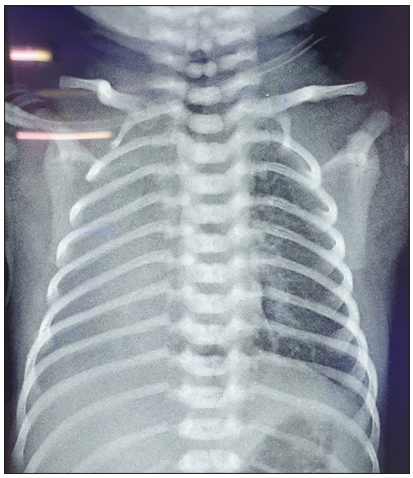

Figure 1: X-ray chest showing right-sided lobar aplasia

development after the division of the main bronchus; and Group IV: incomplete development of subsegmental bronchi and a small segment of the corresponding lobe.[1] Pulmonary agenesis was initially classified by Schneider and Schawatbe and later modified by Boyden into three variants based on the stage of development of lung bud (i) agenesis, in which there is complete absence of lung tissue; (ii) aplasia, in which the rudimentary bronchus is present; however no lung tissue is present; and (iii) hypoplasia, in which all the normal pulmonary tissues are present but are underdeveloped.[2]

The etiology is not entirely clear, but genetic, teratogenic, and mechanical factors have been attributed. There is a high incidence of associated cardiac, gastrointestinal, genitourinary, skeletal, central nervous system malformations, and VACTERL anomalies.[3] Antenatally, LA may be suspected on the total or partial absence of pulmonary parenchyma associated with an abnormal position of the heart within the thorax.[4] Postnatally, contrast-enhanced CT scan chest forms the standard investigation for the diagnosis of LA. Three-dimensional reconstructed images can be particularly helpful in delineating abnormalities of the bronchi and associated arterial and venous structures.

The presentation of LA is not limited to the neonatal period. Gowrinath *et al*.[5] recorded persistent, recurrent cough as an uncommon presentation of left upper lobe agenesis in a 34-year-old man with associated patent foramen ovale and PDA. Gupta *et al*.[6] reported a 29-year-old male who had no major disease, who had a chest X-ray as part of a routine health checkup, and who was diagnosed with isolated left upper lobe pulmonary agenesis. Kuo *et al*.[7] reported an 18-year-old female, diagnosed with allergic rhinitis and bronchial asthma since her early childhood. Following complete workup was found to have agenesis of the right upper lobe.

The most common complications in these patients are respiratory tract infections. Tracheal compression and kinking caused by heart rotation, mediastinal shift, and tracheal curvature cause respiratory insufficiency.[8] No treatment is required in asymptomatic cases. Treatment is needed for recurrent lower respiratory tract infections. Various surgical corrections have been attempted to correct

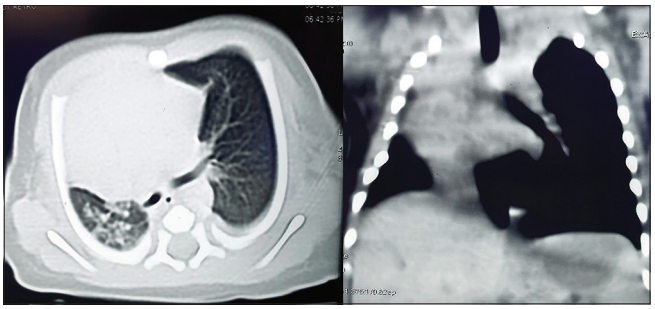

Figure 2: Computed tomography scan chest showing absence of upper and middle lobes of the right lung with shifting of the mediastinum toward the right side

respiratory distress due to air trapping in normal lung and tracheal abnormalities. Placement of inflatable prosthesis, aortopexy, and diaphragmatic translocation have been reported.[9] These procedures alleviate respiratory distress by reducing heart rotation, mediastinal shift, relieving kink, and tracheal compression.[9] It is not recommended to use as a prophylactic surgery.[3] The close follow-up for neonates suffering from unilateral pulmonary LA is of paramount importance. During the neonatal and childhood periods, they may be mildly symptomatic or asymptomatic, but later in life, they might experience obstructive pulmonary or reactive airway disease. Prognosis depends on the severity of the underlying congenital defects and the involvement of the normal lung in the phase of disease.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the parent has given his consent for images of his child and other clinical information to be reported in the journal.

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Conflicts of interest

There are no conflicts of interest.

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