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CASE REPORT

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Isolated agenesis of right upper lobe of lung: A case report

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ABSTRACT

Isolated agenesis of the right upper lobe of lung is very rare and can remain undiagnosed till adulthood. Patients may present with abnormal chest radiograph depicting features of volume loss, which is impossible to differentiate from other more common causes, like lobar collapse, producing this appearance. Contrast enhanced computed tomography (CECT) of the chest can delineate exact bronchial and vascular anatomy, thereby providing the diagnosis. However, given the rarity of this condition, it can pose a diagnostic challenge to the radiologist. We report a case of a young asthmatic girl presenting with recent onset of cough and shortness of breath. She was referred to the department of radiology for CECT of the chest following an abnormal chest radiograph. After a thorough imaging study, a diagnosis of agenesis of right upper lobe was made.

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KEYWORDS

Computed tomography; agenesis; right upper lobe.

Introduction

Developmental anomalies of the lung are a spectrum of congenital malformations ranging from complete agenesis to hypoplasia, depending upon the presence or absence of bronchus, lung parenchyma, and pulmonary artery. Among all, pulmonary agenesis is a very rare congenital abnormality resulting from impaired growth of the primitive lung bud [1,2] and is frequently associated with other anomalies of the cardiovascular and genitourinary systems. Spencer has further divided pulmonary agenesis into three categories, depending on the location of agenesis: (1) bilateral complete agenesis; (2) unilateral agenesis; and (3) lobar agenesis or lesser forms of congenital anomalies [3]. Isolated lobar agenesis is extremely rare with only few reported cases. The abnormality goes undetected or is incidentally detected in the majority of cases. Symptomatic patients present in early childhood with symptoms of recurrent respiratory tract infection. Chest x-ray can show elevated hemidiaphragm and ipsilateral mediastinal shift, indicating volume loss in effected hemithorax. Contrast enhanced computed tomography (CECT) of the chest with multiplanar reconstructions can provide a confident diagnosis by depicting the accurate anatomy of the bronchial tree, as well as pulmonary vasculature. We describe a case of a young woman who was referred with an abnormal chest x-ray and was found to have agenesis of the right upper lobe on chest CECT.

Case Report

A 22-year-old girl presented in the outpatient department of our hospital with complaints of cough and expectoration for the last 10 days with off and on episodes of breathlessness. There was no history of associated fever, orthopnea, palpitation, chest pain, or weight loss. She had been diagnosed with bronchial asthma since childhood.

On physical examination, she had a well-developed stature without any abnormalities. General physical examination was within normal limits. Her respiratory examination revealed relatively reduced breath sounds in the right upper chest as compared to the left side. Her cardiovascular examination was unremarkable. Transthoracic echocardiography did not reveal any abnormality.

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Chest x-ray showed volume loss in right hemithorax (Fig. 1).

On CECT of the chest, there was the presence of two lung lobes on right side with hyparterial bronchus supplying the right lung (Fig. 2). Normal eparterial right upper lobe bronchus was not visualized. The right upper lobe was absent with non-visualized horizontal fissure on the right side. Oblique fissure was seen on the right side between the right middle and lower lobe (Fig. 3A). The right lung was decreased in volume; however, no evidence of collapse was seen (Fig. 2). The right pulmonary artery was mildly reduced in caliber (measuring ~13 mm in diameter) as compared to the left pulmonary artery (measuring ~18 mm) (Fig. 3B). The right pulmonary artery continued as the right lower lobe pulmonary artery without any upper lobar branch. The heart was shifted toward the right side with the apex toward the left side (Fig. 3C). No other associated congenital anomaly was found.

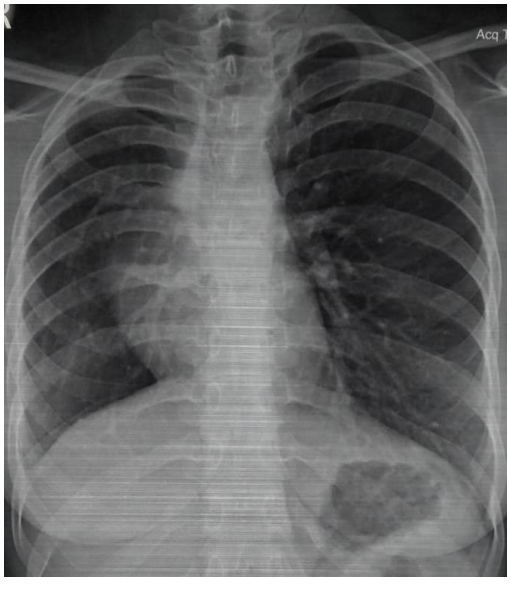


Figure 1. Chest x-ray posteroanterior view (PA view) showing a decreased volume of right lung with tracheomediastinal shift toward the right side. However, no radio-opacity is seen in bilateral lung fields. Also note the presence of stomach bubble on the left side and liver shadow on right side.

Based upon these computed tomography (CT) findings, a diagnosis of agenesis of the right upper lobe was made. Transthoracic echocardiography and ultrasound abdomen revealed no associated structural anomalies. The patient was treated on outpatient basis for acute exacerbation of bronchial asthma.

Discussion

Any perinatal insult during the phase of lung bud development is the most likely cause of the underdevelopment of the lung [4]. Apart from genetic causes, teratogen (allopurinol) [5] and vitamin A deficiency during pregnancy [6] have also been hypothesized as its causes. Abnormal development of the lung is classified into agenesis, aplasia, and hypoplasia. Agenesis shows complete absence of bronchus, lung parenchyma, and pulmonary artery. Aplasia has a rudimentary bronchus with absent lung parenchyma. Hypoplasia has hypoplastic bronchus and lung parenchyma [1,2,4].

Lobar agenesis of the lung is a very rare congenital anomaly that can occur in isolation or in conjunction with other congenital defects of the cardiovascular, musculoskeletal, or gastrointestinal

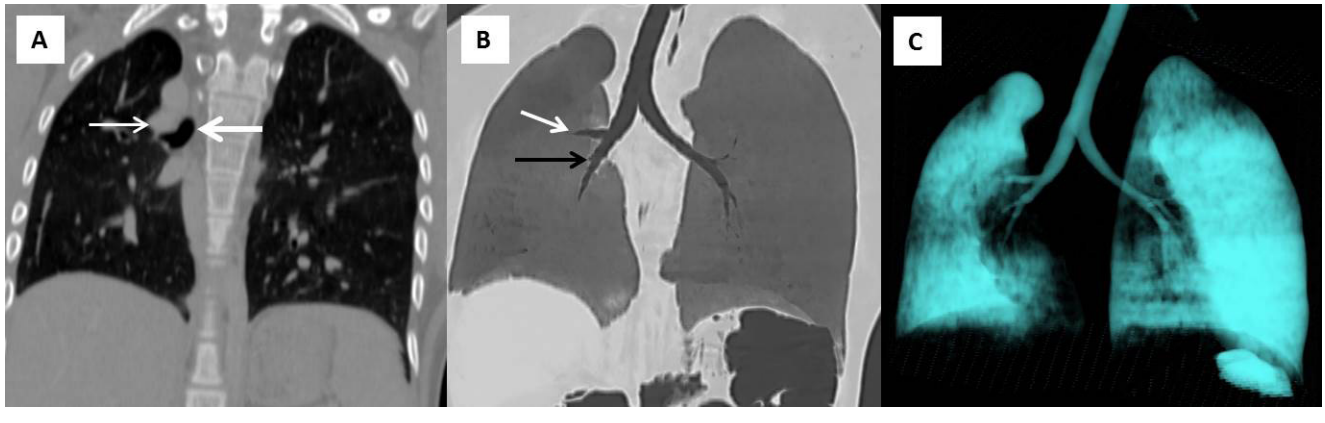


Figure 2. (A–C): A: coronal multiplanar reconstruction (MPR) image of CECT of the chest showing right main bronchus (large arrow) lying inferior to the right pulmonary artery (small arrow) suggestive of hyparterial bronchus. B,C: coronal minimum intensity projection (MinIP) image (B) and coronal volume rendered (VR) image showing division of right main bronchus into the right middle lobe (white arrow in B) and lower lobe bronchus (black arrow in B) with non-visualized upper lobe bronchus. Also note the decreased volume of the right lung as compared to the left lung.



Figure 3. (A–C): A: Oblique sagittal reformatted CT image showing oblique fissure between the right middle and lower lobe (straight arrow). Horizontal fissure was not visualized. B: Axial CECT image showing right main bronchus (straight arrow) lying posterior to right pulmonary artery with mildly reduced caliber of right pulmonary artery (*). C: More caudal axial CECT of the chest section showing cardiac shif

systems [5]. Pulmonary artery hypoplasia is commonly accompanied with such an anomaly of the lung lobe. Isolated lobar agenesis can remain asymptomatic till adulthood and the condition is diagnosed following an abnormal chest radiograph. However, it can also present as recurrent respiratory tract infections in childhood. It has been postulated that upward displacement of bronchus due to volume loss can lead to impaired drainage of respiratory secretions, thereby leading to recurrent infections of the respiratory tract [7].

Chest radiograph in the right upper lobe agenesis of the lung may show decreased right lung volume, mediastinal shift toward right side, and elevated right hemidiaphragm. The differential diagnosis includes right upper lobe collapse, lobectomy, and various causes of raised right hemidiaphragm (diaphragmatic palsy, eventration, subpulmonic effusion, and intra-abdominal volume increase) [8]. Taking a history of lobectomy is of utmost importance in these cases. A contrast enhanced chest CT is the investigation of choice to diagnose the developmental disorders of the lung. Multiplanar capability of CT is particularly helpful in delineating abnormalities of the bronchi and associated vascular anomalies.

On CECT of the chest isolated agenesis of the right upper lobe can mimic left-sided isomerism seen in heterotaxy syndromes [8]. In these cases, left-sided isomerism is associated with polysplenia syndrome, characterized by the liver located in midline, indeterminate location of stomach, multiple spleens, and interruption of inferior vena cava with azygous or hemiazygous continuation [9]. However, in our index case no associated anomalies were present (as seen on transthoracic echocardiography and ultrasound abdomen). Also, volume loss on right side and absence of upper lobar branch of right pulmonary artery confirmed our diagnosis.

Treatment depends on the patient's symptoms, with majority of the patients being either treated conservatively or left alone without any further intervention. Treatment is necessary for recurrent chest infections. No other associated congenital malformation favors a good prognosis [8].

To conclude, isolated agenesis of the right upper lobe of lung is extremely rare. CECT of the chest is the imaging modality of choice in these cases to demonstrate anatomy of bronchial tree. In an abnormal chest radiograph with volume loss in one hemithorax, possibility of lobar agenesis should also be considered particularly in absence of any radio-opacity. Radiologist should be familiar with this rare condition which can present as incidental finding on chest radiograph or chest CT.

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