



Congenital Primary Right Lung Agenesis in A Child – MDCT Imaging Features



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Summary

Pulmonary agenesis is a very uncommon congenital abnormality that represents the unsuccessful growth of the primitive lung bud. The cause may be a primary embryogenic defect or secondary to situations that restrict fetal lung growth. In the great majority of the cases, the diagnosis is usually made at or soon after birth, but some cases go unnoticed up to adulthood. It is generally associated with other congenital defects and must be suspected in the presence of a total radio-opaque hemithorax. In this article, we present a case of 8 years old female child with congenital pulmonary Agenesis who remained asymptomatic until childhood.

Keywords: Congenital Primary Pulmonary Agenesis; Pulmonary Hypoplasia; Pulmonary Aplasia; Pulmonary Artery Agenesis

Introduction

Lung agenesis is a rare developmental anomaly which affects both lungs. Long term survival with unilateral lung agenesis is possible in the absence of associated severe anomalies. However, the condition is frequently associated with other congenital abnormalities, particularly esophageal atresia and the VACTERL syndrome (vertebral, anal, cardiac, tracheo-esophageal, renal and radial and limb anomalies) [1]. The prevalence of this condition has been noted to be 0.0034 - 0.0097%. There appears to be no sexual predilection for this condition. Most cases present in the neonatal period with cyanosis, tachypnea, dyspnea, stridor or feeding difficulties. The condition is often associated with fetal distress at birth. Yet, it may also be asymptomatic and manifest itself in adulthood. The cause may be a primary embryogenic defect or secondary to situations that restrict fetal lung growth [2]. Left-sided agenesis (70% of cases) is more frequent than right-sided. Right-sided defects have a poorer prognosis due to often coexisting cardiac anomalies or greater mediastinal shift and pressure on other structures [3]. We herewith report an 8 years old female child with right congenital Primary Pulmonary Agenesis and describe the MDCT imaging features.

Case Report

An 8 years old female child presented with complaints of dyspnea and left chest pain to the hospital for further

investigations. General physical examination was non-contributory. Chest examination revealed absent air entry on right side, mediastinal shift to right side with hyper-resonance on left side. Cardiac examination revealed apex beat palpable in the right 4th intercostal space. Clinical examination of the abdomen revealed no abnormalities.

- i. Routine biochemical and arterial blood gas analyses were unremarkable. Electrocardiographic assessment demonstrated a normal sinus rhythm pattern.
- ii. The patient also had a cardiology consult to rule out any vascular, cardiac anomalies or effect of the agenesis on cardiac function.
- iii. Echocardiography was suggestive of dextroposed heart with normal intracardiac connections.

Radiograph of chest revealed symmetric bony cage, opaque right hemithorax, mediastinal shift with herniation of the normal lung towards the right side (Figure 1). Ultrasonography done revealed absence of right lung tissue and shifting of heart towards the affected side. To exclude the diagnosis of a pulmonary thrombo-embolic event, the patient underwent further evaluation with contrast-enhanced MDCT scanning (General Electric (GE) Light Speed Multi Slice CT Scanner) of the thorax with dynamic

bolus injection of contrast (100 ml Omnipaque 350 mg/mL, 4.5 mL/s). Multidetector Computed Tomography (MDCT) scan revealed that the volume of the right hemithorax was reduced, Agenesis of right lung and bronchial tree with the mediastinal structures displaced towards the right. There was compensatory hypertrophy of left lung, while the superior lobe of the left lung was partially displaced towards the right hemithorax (Figures

2-4). MDCT Angiography revealed complete agenesis of right lung with non-visualization of right pulmonary artery and vein (Figure 5). On Bronchoscopy the patient had agenesis of the Right bronchial tree and no endobronchial lesions of left lung. Based on these findings a diagnosis of Congenital Primary Right Lung Agenesis was made.

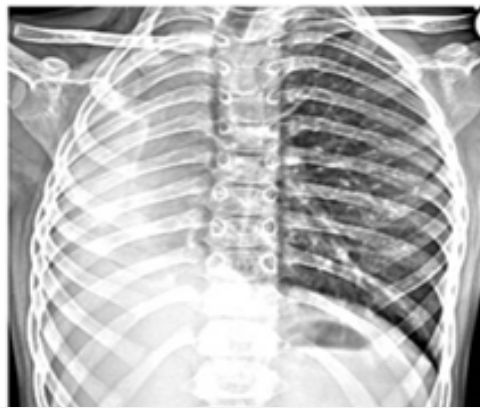


Figure 1: Postero-Anterior Radiograph of Chest revealed symmetric bony cage, opaque right hemithorax, mediastinal shift with herniation of the normal lung towards the right side.

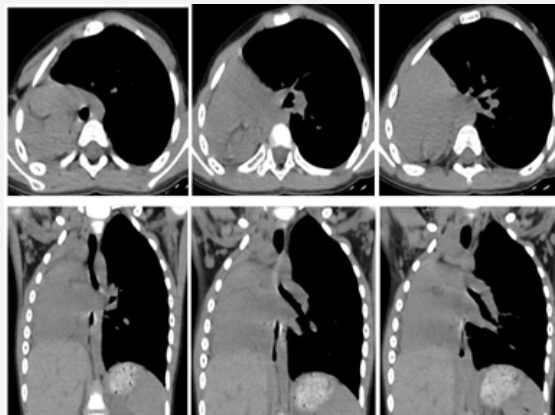


Figure 2: Non-contrast mediastinal window CT, Axial (A-C) and reconstructed coronal (D-F) sections shows reduced volume of the Right hemithorax, Agenesis of right lung and displaced mediastinal structures towards the right.

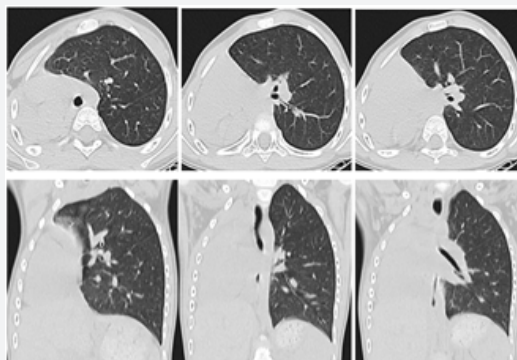


Figure 3: MDCT chest Lung window, Axial (A-C) and Reconstructed Coronal (D-F) sections shows Absent right lung and Bronchial tree, Compensatory hypertrophy of left lung and herniation of superior lobe of left lung towards the right hemithorax.

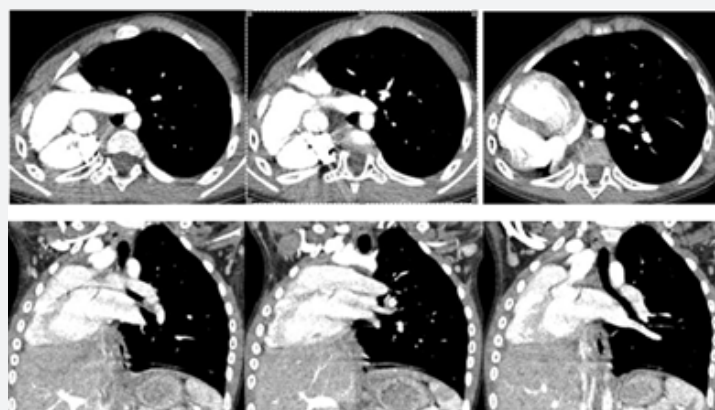


Figure 4: Contrast enhanced CT Arterial phase, Axial (A-C) and reconstructed coronal (D-F) sections shows Agnesis of right lung, Absent right pulmonary artery and displaced heart and mediastinum to right.



Figure 5: MDCT Angiography. 3D Volume rendered images (A,B) and MIP images (C) revealed Non-visualisation of Right pulmonary artery and vein and normal left pulmonary artery and its branches.

Discussion

Pulmonary Agnesis is a rare congenital anomaly characterized by complete absence of the bronchus, lung parenchyma, and pulmonary vasculature on that side. Unilateral lung agnesis has a birth prevalence of approximately 1 in 15,000 [4]. Normally the lung develops as a diverticulum from foregut in the 4th week of intrauterine life. Pulmonary agnesis results due to failure of the bronchial analogue to divide equally between the two lung buds, leading to normal development of one side, while failure of other side to develop either completely (agnesis/aplasia) or partially (dysplasia or hypoplasia) [5].

Genetic, teratogenic and mechanical factors have been implicated in the etiology of pulmonary agnesis [6,7]. A history of recurrent chest infections during the first year of life is the usual history elicited; however, the patient may be completely asymptomatic and diagnosed accidentally from the radiograph or detected during autopsy [8]. Associated congenital malformations of the cardiovascular, skeletal, gastrointestinal, or genitourinary systems are present in half of the cases and may modify clinical presentation, course and outcome of patients with pulmonary agnesis [9]. Schneider classified pulmonary agnesis into three groups, which were later modified by Boyden in 1955 [8].

Type 1 (Agnesis) - There is a complete absence of lung and bronchus and there is no vascular supply to the affected side.

Type 2 (Aplasia) - There is a rudimentary bronchus with the complete absence of pulmonary parenchyma.

Type 3 (Hypoplasia) - variable amounts of bronchial tree, pulmonary parenchyma, and supporting vasculature are present. Monaldi divided the maldevelopment of lung in four groups [10].

Group I: No bifurcation of trachea;

Group II: Only rudimentary main bronchus;

Group III: Incomplete development after division of main bronchus; and

Group IV: Incomplete development of subsegmental bronchi and small segment of the corresponding lobe.

Functionally, unilateral lung agnesis and lung aplasia are similar. Pathologically sole lung is larger than normal in pulmonary agnesis, and this enlargement is true hypertrophy and not emphysema [11]. Patients presenting late usually have readily detectable flattening and reduced movement of the chest wall on the affected side, with reduced air entry on auscultation. Breath sounds from the herniated, hypertrophied

lungs may be heard on the side of agenesis except in the axilla and base Chest wall deformity can be quite pronounced, with an associated secondary scoliosis [11]. Diagnosis of Pulmonary Agenesis has been made on chest skiagram, bronchoscopy, bronchography and angiography. With the advent of CT scan, these invasive procedures which entail significant risk have become unnecessary [12].

As might be expected, Chest radiographs in patients with unilateral absence of a lung show loss of aeration and marked volume loss on the affected side. Volume loss is shown as elevation of the ipsilateral hemidiaphragm, shift of the mediastinum toward the abnormal side, and anterior herniation of the contralateral lung. There is usually characteristic wedge-shaped opacity in the inferior and lateral aspect of the affected hemithorax. This opacity is due to herniation of heart and mediastinum into the lower thorax and proliferation of fat to partially compensate for the absence of lung tissue. The obvious Differential diagnoses are acquired total lung collapse or prior pneumonectomy [13]. Other conditions to consider in the differential include hyperlucent and hypoplastic lung syndromes, obstructive lung lesions mainly cancer, diaphragmatic hernia, adenomatoid cystic malformations and sequestrations and the Scimitar syndrome [14]. Pulmonary Angiography shows absence of ipsilateral pulmonary artery. Bronchoscopy demonstrates underdeveloped bronchus in agenesis. Other investigations e.g. echocardiogram, ultrasound abdomen etc. are required depending upon the presence of associated congenital abnormality [15]. The MDCT allows high-resolution isotropic imaging and is the ideal modality for diagnosing right lung agenesis and for diagnosing simultaneously any associated tracheobronchial anomaly. MDCT can very speedily acquire volumetric data sets, allowing for 2D, multiplanar, maximum intensity projection and volume rendering technique reconstructions, and has the capability to image both airway and vascular abnormalities. The characteristic MDCT findings include opaque hemithorax, Agenesis of lung with mediastinal shift towards the affected side and bony cage symmetry (Figures 2-4). Similar findings were seen in our case and a diagnosis of primary right lung Agenesis was made. However, no etiological diagnosis could be confirmed as genetic mutation analysis was not affordable in our case.

No treatment is required in asymptomatic cases. Treatment is necessary for chest infections. Patients having stumps (hypoplastic bud) may require surgical removal if postural drainage and antibiotics fail to resolve the infection. Corrective surgery of associated congenital anomalies, wherever feasible, may be undertaken [16]. Several methods have been proposed to reduce vascular compression and intrinsic stenosis such as mediastinal stabilization by an expansion prosthesis, aortopexy, aortic grafting and resection, detour of the left pulmonary artery and slide tracheoplasty [4]. Overall, prognosis depends on two factors. Firstly, the severity of associated congenital anomalies and secondly, involvement of the normal lung in any disease

process. While bilateral Pulmonary agenesis is incompatible with life, 50% of those with even unilateral lung agenesis die within 5 years of life [17]. Right lung Agenesis carries a poorer prognosis because of the larger shift of the heart and mediastinum with a consequent distortion of the great vessels, especially aortic arch resulting in extrinsic compression over the trachea anteriorly [18,19]. In our case the child is surviving beyond 5 years of life.

Conclusion

Pulmonary Agenesis is an uncommon pathology. Clinical findings of recurrent respiratory infections and radiologic evidence of opaque hemithorax, bony symmetry and herniation of normal lung to the affected side, along with associated congenital anomalies are suggestive of Pulmonary Agenesis. No treatment is required in asymptomatic cases. Management is mainly supportive, correcting associated malformations, prevention and treatment of repeated respiratory tract infections. MDCT imaging not only guides us to a proper diagnosis but also can identify the etiology, level of airway narrowing and associated congenital anomalies, thus helping in proper patient care.

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