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ANATOMIC VARIATIONS

Adult presentation of asymptomatic right lung agenesis: a rare anatomical variation

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Abstract
Pulmonary agenesis is a very rare congenital anomaly characterized by the absence of pulmonary parenchyma and its vasculature. The diagnosis is usually during childhood. Herein, we report a case of incidental discover right pulmonary agenesis in adulthood male. A 30-year-old male presented with pre-operative medical evaluation for varicocele. There was no complaining from respiratory symptoms. He underwent plain chest X-ray and post-contrast CT scanning was performed using 64 multi-detector CT scanner. Chest Plain X-ray revealed cardiac and mediastinal shift to right side with hyper-inflated left lung crosses to right side. Multi-detector computed tomography examination revealed total absence of right lung with compensatory hyperinflation and increase volume of left lung. Hyper-inflated left lung extended to right hemithorax. Mediastinal structures including heart and great vessels were displaced to middle and lower parts of right hemithorax. Descending aorta located to anterior to thoracic vertebrae. Right pulmonary artery was absent. Right main bronchus was rudimentary. No detected other organ anomalies. No detected bony thoracic cage abnormalities.

Keywords Lung agenesis · Pulmonary artery · Congenital

Introduction

Pulmonary agenesis is a very rare congenital anomaly characterized by the absence of pulmonary parenchyma and its vasculature. Bilateral pulmonary agenesis is incompatible with life. Unilateral agenesis is often associated with other organ malformations. Right lung agenesis has poor prognosis as compared to left lung agenesis [4]. Earlier diagnosis also allows for proper management and follow-up care. The diagnosis is usually during childhood but can be delayed, if the clinician is not aware about this entity [1]. Herein, we report a case of incidental discover right pulmonary agenesis in adulthood male.

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Case report

A 30-year-old male presented with pre-operative medical evaluation for varicocele. There was no complaining from respiratory symptoms. He underwent plain chest X-ray. Post-contrast CT scanning were performed using 64 multi-detector CT scanner (Brilliance 64; Philips Healthcare, Best, Netherlands). For contrast-enhanced CT, a total of 100 mL of ohexol (Omnipaque; GE Healthcare, Cork, Ireland) 350 mg/mL was given intravenously. The images were viewed on both lung (window width, 1200 HU; level, − 600 HU) and mediastinal (window width, 350 HU; level, 40 HU) setting. The images were then transferred to a workstation (Extended Brilliance Workspace V3.5.0.2254; Philips Healthcare, Cleveland, OH, USA). All CT images were reviewed on a Picture Archiving and Communication System (PACS, Paxeramed, Paxeramed Corp, Oslip, Austria). The reader recorded the following findings: ground-glass opacities (GGOs), fibrosis, consolidation, nodules, pleural effusion, cardiac chambers, pericardial effusion, mediastinal structures and great vessels. Great vessels evaluation included ascending, descending aorta, aortic arch and its branches, pulmonary arteries, and its branches. Trachea, main trachea, main bronchi, lobar & segmental bronchioles

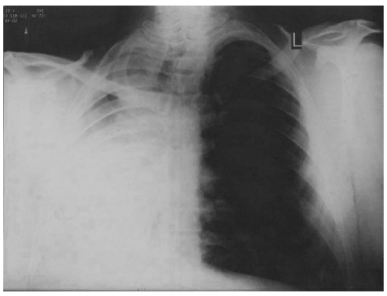

Fig. 1 Chest Plain X-ray (PA view) reveals: Homogeneous opacity of middle and lower zones of right hemithorax with deviation of heart to right side. Hyper-inflated left lung crosses to right side

were evaluated. Written informed consent was obtained from the patient.

Chest Plain X-ray revealed cardiac and mediastinal shift to right side with hyper-inflated left lung crosses to right side (Fig. 1). Multi-detector computed tomography (MDCT) examination revealed total absence of right lung with compensatory hyperinflation and increase volume of left lung. Hyper-inflated left lung extended to right hemithorax. Mediastinal structures including heart and great vessels were displaced to middle and lower parts of right hemithorax. Descending aorta located to anterior to thoracic vertebrae. Right pulmonary artery was absent. Right main bronchus was rudimentary (Figs. 2, 3). No detected other organ anomalies. No detected bony thoracic cage abnormalities.

Discussion

Unilateral lung agenesis is a relatively rare congenital anomaly with a reported incidence of 1 in 15,000 births. Due to its association with other congenital malformations; only few cases have been a normal life span. Many cases have associated organ malformation which increase their mortality [2]. This is not in agreement with our study. Our case is unilateral lung agenesis that incidental discovers at age of 30. Additionally; there are no other associated anomalies or pulmonary hypertension.

Diagnosis of unilateral lung agenesis can be suspected from plain chest X-ray films. On chest X-ray, there may be cardiac and mediastinal displacement, the absence of the pulmonary artery shadow on the affected side, an ipsilateral elevation of the diaphragm, contralateral compensatory

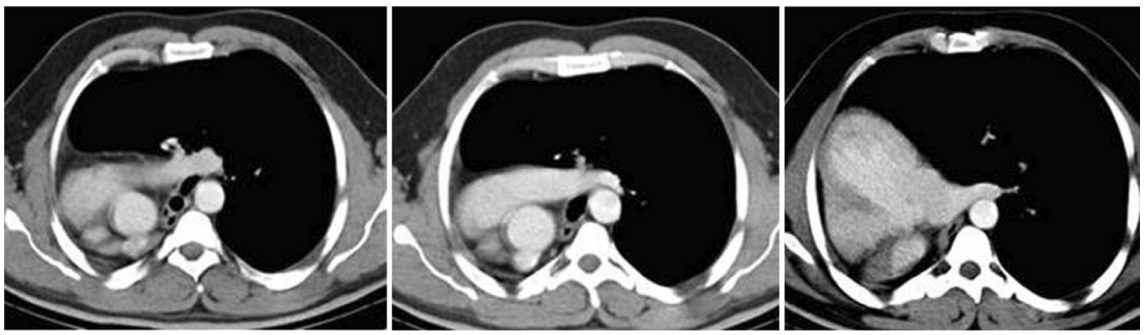

Fig. 2 Axial MDCT scan (mediastinal window) reveals absence right pulmonary artery. Descending aorta lies anterior to thoracic vertebrae. Cardiac shift to right side is detected

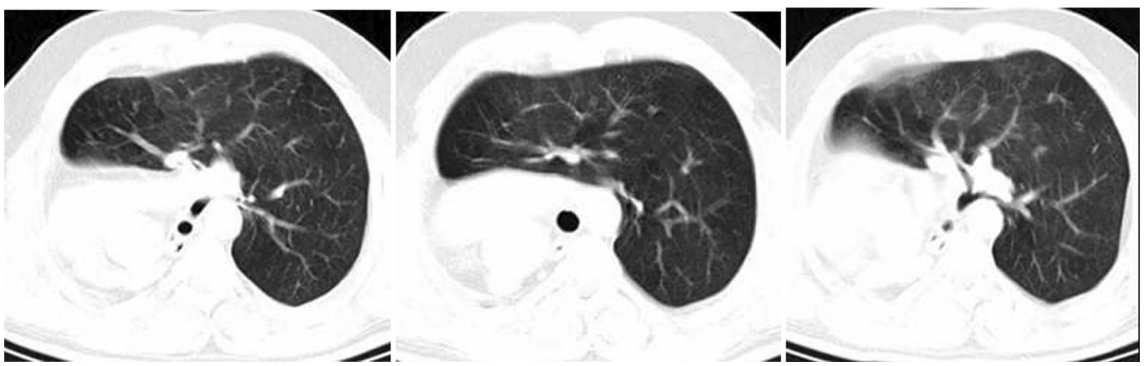

Fig. 3 Axial MDCT scan (pulmonary window) revealed evidence of tracheal bifurcation with rudimentary right main bronchus. Hyper-inflated left lung extends to right side. Left lobar bronchioles are normal

hyperinflation of the hemithorax, and herniation of lung across the midline [3]. All the features were present in our patient.

The characteristic radiographic findings include: absence of an ipsilateral pulmonary artery with enlargement of the contralateral artery, total volume loss of the affected lung and ipsilateral mediastinal shift. There is associated hyperinflation of the contralateral lung [1]. This coincides with our case.

Nguyen et al. [5] found that a chest computed tomography (CT) with intravenous contrast revealed complete absent right lung and complete shift of mediastinal structure in to right hemithorax. There is rudimentary blind-ending right bronchus and entire heart in right chest. Evaluation in the cardiology clinic revealed a well-appearing, well-developed adolescent with heart sounds that were able to be auscultated only in the right chest and absent lung sounds in the right lung fields. Additional airway reconstruction from chest CT images showed a rudimentary right main bronchus which confirmed the diagnosis of right lung aplasia [5]. This is in agreement with our findings.

Right lung agenesis was confirmed by CT-scan. 3D CT construction also clearly delineates lung parenchyma with pulmonary and bronchial tree. The early intervention to remove the distress is the key to save the patient. So, it is concluded that congenital lung agenesis without any other major congenital abnormality is an extremely rare anomaly and may be asymptomatic throughout life. It is recommended that CT scan is the key diagnostic approach for lung agenesis and invasive diagnostic procedures and prophylactic surgery should not be done in asymptomatic cases [6]. Our patient underwent follow up without surgical interference.

In the present case, there is adulthood incidental discover right lung agenesis without associated other anomalies. It is may be considered in differential diagnosis with other condition as bronchial obstruction from mucus plugging or malignancy causing endobronchial obstruction, prior lobectomy, or congenital pulmonary hypoplasia.

Author contributions AE and ME conceived the study concept and design and drafted the manuscript. AE, performed the acquisition of data and critical revision of the manuscript for important intellectual content. All authors read and approved the final manuscript.

Compliance with ethical standards

Conflict of interest The authors declare no conflict of interest.

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