

CASE REPORT

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# An unique case of isolated right upper lobe lung agenesis with abnormal middle lobe segmentation

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## Abstract

**Background:** Isolated right upper lobe pulmonary agenesis is a highly uncommon congenital anomaly, usually detected at adulthood when the patient is evaluated for an incidental abnormal chest radiograph. Chest radiography findings are non-specific. Chest computed tomography with pulmonary angiography is the modality of choice for diagnosing pulmonary agenesis.

**Case presentation:** We describe a case of isolated right upper lobe agenesis in a young man who presented with mild shortness of breath and an abnormal chest radiograph. High-resolution computed tomography chest showed right upper lobe pulmonary agenesis which was initially erroneously diagnosed as right lower lobe collapse on chest X-ray. Abnormal segmentation of the middle lobe was also seen which has not been described earlier according to our literature search.

**Conclusions:** There should be a high index of suspicion for congenital anomalies on chest X-ray to recommend further imaging studies. This case highlights the importance of computed tomography with pulmonary angiography to adequately assess and characterize the congenital lung anomalies.

**Keywords:** Pulmonary agenesis, Pulmonary aplasia, Pulmonary hypoplasia, Middle lobe Abnormal segmentation, Congenital lung anomaly, Computed tomography

## Background

Embryonic anomalies of the bronchi are one of the most common types of congenital malformations of the foregut. Bronchial anomalies occur in approximately 2% of the adult population, and the most common abnormality of the bronchial tree involves the right upper bronchus [1]. Pulmonary agenesis is a very rare congenital condition that represents the arrested growth of the primitive lung bud. The majority of reported cases have associated congenital anomalies involving the cardiovascular, musculoskeletal, or gastrointestinal systems. Infrequently,

genitourinary or central nervous systems were involved [2]. Here, we present a case of a young man with isolated right upper lobar pulmonary agenesis and abnormal segmentation of the middle lobe.

## Case presentation

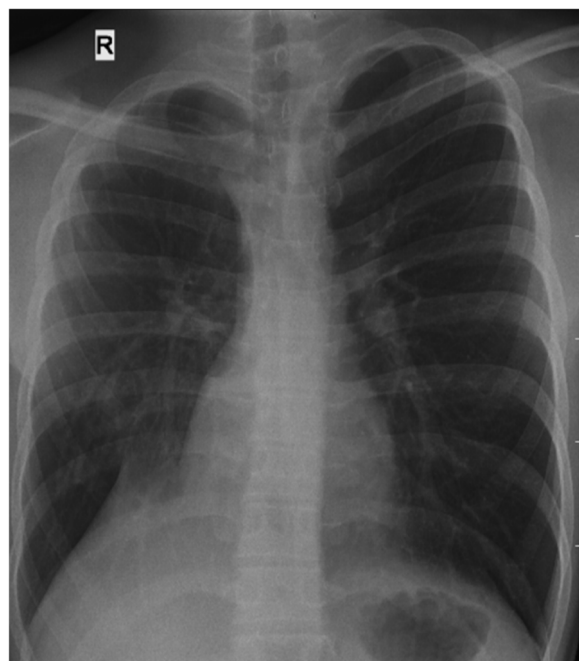
Eighteen-year-old young gentleman, non-smoker, without a familial lung cancer history, presented to the respiratory clinic with mild shortness of breath. His birth history was unremarkable. There was no history of recurrent respiratory infections or bronchial asthma since childhood. There was no history of chest pain, orthopnea, palpitation, loss of appetite, or weight. His medical history was unremarkable. Physical examination showed no abnormalities. Routine blood counts and urine analysis were normal. Sputum examinations were negative for mycobacteria and bacteria. The chest radiograph showed

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a mild reduction of the right lung volume with an ipsilateral shifting of the mediastinum and elevation of the right hemidiaphragm (Fig. 1). Atelectasis of right lower lobe was suspected.

High-resolution computed tomography (HRCT) chest was done for further evaluation of abnormal chest radiograph. CT showed a reduction in the volume of the right lung with elevated right hemi diaphragm and mild ipsilateral mediastinal shift (Fig. 2a). The right pulmonary artery caliber was relatively smaller than the left



**Fig. 1** Chest radiograph shows reduction of lung volume on the right side with mild ipsilateral mediastinal shift with an elevation of the right hemidiaphragm

pulmonary artery (Fig. 2b). There were bilobed bilateral lungs. The right upper lobe, bronchus, pulmonary artery, and veins of the right upper lobe were absent (Figs. 2c and 3b). Horizontal fissure was absent. A right oblique fissure was discontinuous in the right perihilar region. Superior segment of the right lower lobe was seen extending up to the apex (Fig. 3a). Two bronchi originated from the bronchus intermedius at the mid bronchus level (Fig. 3b). The first artery originating from the right main pulmonary artery was present at the level of the major fissure. Pulmonary vessels were crossing across a defect in the discontinuous oblique fissure. There was mild compensatory hyperinflation of the left lung. Left lung bronchial anatomy appeared normal. Ultrasonography of the abdomen and CT upper abdomen showed no significant anomaly in the liver, spleen, kidney, and other abdominal organs. He was diagnosed as a case of right upper lobe pulmonary agenesis.

The middle lobe showed abnormal segmentation. A right oblique fissure was seen dividing the middle lobe with the medial segmental bronchus of the middle lobe seen supplying the lung anterior to the fissure. Lateral segmental bronchus of the middle lobe and its corresponding segmental pulmonary artery was seen coursing posterior to the distal oblique fissure (Fig. 3c).

Transthoracic echocardiogram revealed no evidence of structural abnormalities. Pulmonary function tests were normal.

## Discussion

Lung agenesis is a rare developmental anomaly where there is a complete absence or hypoplasia of one or both lungs. It may be present as isolated lung agenesis or may be associated with congenital anomalies [3].

Pulmonary agenesis has been classified into three types as per Boyden classification. It is morphologically



**Fig. 2** Non-contrast computed tomography scan of the chest. **a** CT Coronal image in mediastinal window shows right side volume loss with ipsilateral mediastinal shift and elevation of the right hemidiaphragm. **b** CT Axial image in the mediastinal window shows the relatively small caliber right pulmonary artery (**c**) CT Coronal image in lung window shows complete absence of the right upper lobe pulmonary parenchyma, right upper lobe bronchi, and its vasculature. Also noted is the abnormal right oblique fissure



**Fig. 3** CT chest in lung windows. **a** Sagittal image shows discontinuous right oblique fissure. Superior segment of right lower lobe was seen extending up to the apex. **b** Axial image shows completely absent right upper lobe bronchi and pulmonary parenchyma. Discontinuous right oblique fissure was seen in the right perihilar region. Two bronchi were seen arising from the bronchus intermedius. **c** Axial image shows abnormal segmentation of the middle lobe. Medial segment and lateral segment bronchi of middle lobe seen supplying the lung anterior and posterior to the discontinuous oblique fissure respectively

based on the extent to which bronchopulmonary tissue is absent. Type I is called pulmonary agenesis where there is a complete absence of unilateral lung parenchyma, bronchus, and its pulmonary vessels. Type II is named pulmonary aplasia where there is a complete absence of lung and its pulmonary vessels with a small rudimentary blind ending bronchus. Type III is pulmonary hypoplasia characterized by bronchial hypoplasia with variable reduction of lung tissue and its vessels [4].

The lungs normally develop from the foregut during the 4th and 5th weeks of gestation and pulmonary agenesis occurs during this period. The etiology of lung agenesis is unknown. Lack of vitamin A during pregnancy, genetic, teratogenic, and mechanical factors have been mentioned as possible causes [2, 5]. The incidence of pulmonary agenesis is not exactly documented [6]. About 200 cases of unilateral pulmonary agenesis have been documented in the current literature [7]. The condition is usually unilateral. Bilateral pulmonary agenesis is incompatible with life with respiratory distress and failure. Occasionally, the agenesis may be confined to one lobe, most frequently the left upper lobe. Pulmonary artery hypoplasia is commonly accompanied with such lobar agenesis. Cases of isolated lobar agenesis have been uncommonly reported [1, 3, 8–10].

Pulmonary agenesis may be associated with cardiovascular (tetralogy of Fallot, patent ductus arteriosus and patent foramen ovale), gastrointestinal, skeletal, and genitourinary anomalies in more than 50% of children [5]. Right lung agenesis has a relatively poor prognosis than the left side owing to complications related to mediastinal shift and more frequent association with congenital heart disease [5, 7].

Isolated lobar agenesis of the lung may be asymptomatic and may go unnoticed in childhood until some complications develop. Patients may also present with

dyspnea or recurrent chest infections in childhood or in adult life [2, 11]. Patients are evaluated as an adult with incidental chest radiographic abnormality [11]. Chest X-ray shows loss of volume of ipsilateral hemithorax with a displacement of the mediastinal structures and diaphragm, compensatory hyperinflation, and herniation across the midline [5]. Isolated right upper lobe agenesis has been earlier reported in a few studies [1, 3, 12].

Chest radiograph is not diagnostic for congenital abnormalities of the lung. CT scan of the chest is the most definitive modality to diagnose such anomalies and the associated vascular anomalies. CT, magnetic resonance imaging (MRI), and CT/MR angiography are currently the imaging modalities of choice and provide important diagnostic information. Ideally, in our case, CT scan should have been combined with CT pulmonary angiography for a more confident diagnosis. CT pulmonary angiography was not done in our case as the patient did not give his consent for an intravenous contrast study. Affected lobe shows absent pulmonary parenchyma, bronchial tree, and pulmonary vessels on CT and MRI [5]. Abnormalities of the bronchi and associated vascular structures are even more clearly delineated on three-dimensional reconstruction. Patients requiring embolization or revascularization surgery may need angiography [3].

The differential diagnosis of our patient's chest X-ray was right lower lobe atelectasis, right hemidiaphragm paralysis, right hemidiaphragmatic eventration, pulmonary hypoplasia/agenesis, subpulmonic effusion, and intra-abdominal pathology causing right hemidiaphragm elevation [3].

Bronchial tree pattern in our case was similar to left isomerism and hence heterotaxy syndrome has to be excluded. Left isomerism is characterized by abnormal visceral situs (midline/transverse liver, intestinal

malrotation), polysplenia, congenital heart disease, azygos, or hemiazygos continuation of the inferior vena cava and bilateral bilobed lungs with the bronchial tree forming a mirror image. Our patient did not have any of these abnormalities except for bilobed lungs bilaterally. Absence of the right upper lobe, corresponding right upper lobe bronchus, and its pulmonary vessels were confirmed by CT. Anomalies in other systems were also excluded for a definitive diagnosis of right upper lobar agenesis. The middle lobe showed abnormal segmentation with medial and lateral segments of the middle lobe present medial and lateral to the discontinuous oblique fissure respectively [1].

Medical treatment is most often recommended with treatment for recurrent chest infections. Chest physiotherapy may be of some benefit. Surgical intervention is suggested in cases where there are other associated congenital anomalies. Asymptomatic cases when not associated with any other congenital anomalies do not require any treatment. The prognosis is good if there are no other malformations [2, 7].

## Conclusions

Congenital anomalies of the lung such as agenesis, hypoplasia, and aplasia should be considered with an abnormal chest radiograph findings mimicking collapse, in a symptomatic or an asymptomatic patient. CT with pulmonary angiography is very useful to assess and characterize the exact pathology and also its associated anomalies. Abnormal middle lobe segmentation should also be sought for, in such cases.

## Abbreviations

HRCT: High-resolution computed tomography; CT: Computed tomography; MRI: Magnetic resonance imaging.

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## Authors' contributions

All authors had substantial contributions to the conception of the case report. All authors were active participants in the drafting and revising of the case report. All authors approved the final version of manuscript. All authors agree to be accountable for all aspects of the work. All authors were directly involved in this patient's care and contributed to the writing and editing of this manuscript. RM researched and wrote the patient background and pulmonary history. SP, HV researched and wrote the abstract and conclusion of the manuscript, captured and arranged the images. SP conceptualized the process; researched, compiled, and wrote the procedure, conclusion, and introduction; and served as the overall editor.

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## Availability of data and materials

Provided in this case report.

## Declarations

### Ethics approval and consent to participate

Ethics has approved this study.

### Consent for publication

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his written informed consent for his images and other clinical information to be reported in the journal. The patient understands that his names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

### Competing interests

The authors declare that they have no competing interests.

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