



Left Upper Lobar Agensis Associated with Tracheal Trifurcation: A Case Report

기관의 삼분지와 동반된 좌상엽 무형성: 증례 보고

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Lobar agensis is a rare congenital anomaly that is characterized by the absence of the lobar pulmonary artery, pulmonary vein, bronchi, and parenchyma. We encountered a unique case of a young male patient with agensis of the left upper lobe with tracheal trifurcation into three bronchi, all arising at the carinal level. Complex tracheobronchial anatomy was explicitly demonstrated by three-dimensional CT reconstruction and virtual bronchoscopy. Left upper lobar agensis associated with tracheal trifurcation is an extremely rare anomaly that, to the best of our knowledge, has not been previously reported.

Index terms Lung; Trachea; Congenital Abnormality; Computed Tomography, X-Ray

INTRODUCTION

Pulmonary underdevelopment constitutes a group of congenital anomalies that include agensis, aplasia, and hypoplasia of the lung, on the basis of the presence or absence of lung parenchyma, bronchial tree, and/or pulmonary artery. In pulmonary agensis, the lung parenchyma, pulmonary artery, pulmonary vein, and bronchi of the associated lobe are absent. Mardini and Nyhan (1) reported an incidence of 0.0034%–0.0097% for unilateral pulmonary agensis. Agensis of one or two lobes of a single lung is an extremely rare condition, the exact incidence of which is unknown. Tracheal trifurcation is the most dramatic form of tracheal bronchus; trifurcation at the carina is an extremely rare anomaly. Herein, we report a unique case of a young male patient with agensis of the left upper lobe with tracheal trifurcation.

CASE REPORT

A 22-year-old man was admitted to our hospital due to fever. He had no past medical history except for several mild upper respiratory infections. He demonstrated mini-

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mal wheezing on the upper zone of the left hemi-thorax on physical examination. His C-reactive protein levels were slightly elevated to 40 mg/L; other routine laboratory findings were in the normal range.

Chest radiography showed a mild leftward shift of the mediastinal structures with a reduction in the size of the left hemi-thorax and a downward-displaced left hilum (Fig. 1A).

Multidetector CT scan was performed on a 64-slice CT system (Discovery CT 750HD; GE Healthcare, Milwaukee, WI, USA) to obtain precontrast and postcontrast CT scans with iodinated non-ionic contrast. Contrast-enhanced chest CT revealed a mediastinal shift towards

Fig. 1. A 22-year-old man exhibited agenesis of the left upper lobe, combined with tracheal trifurcation, on CT.

A. Chest X-ray shows volume loss on the left side with tracheal and mediastinal shift toward the left side, as well as slight compensatory hyperinflation of the right lung field. Note the caudally displaced left hilum.

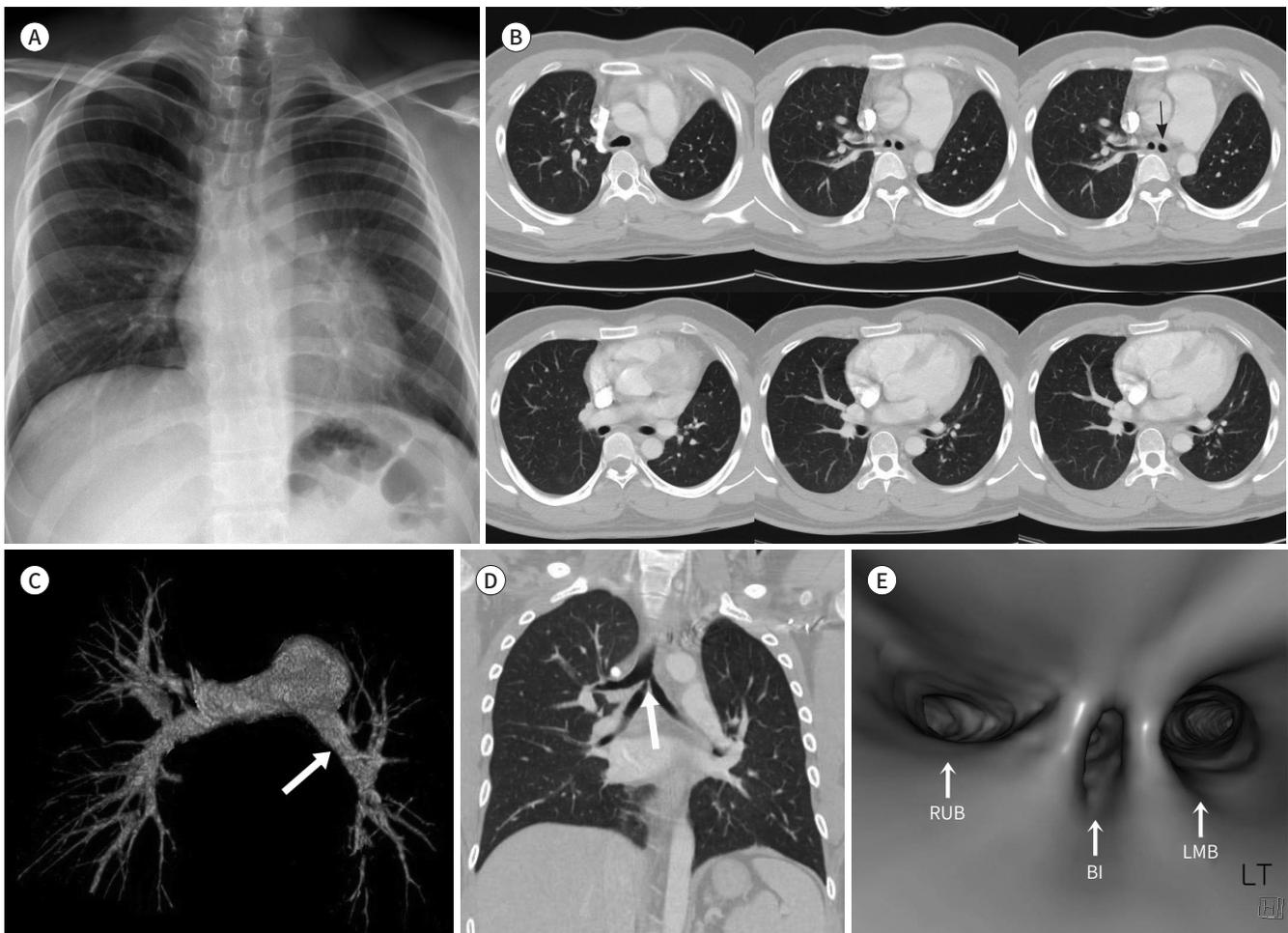
B. Chest CT (lung window) axial view at subcarinal level reveals the left main bronchus (arrow), which continues as the left lower lobe bronchus with mild mediastinal shift toward the ipsilateral side, with compensatory hyperinflation of the right lung field. Note the absence of the left upper lobe.

C. Surface shade display three-dimensional CT-reconstructed image reveals a hypoplastic left pulmonary (arrow), which continues as the left lower lobe pulmonary artery without any upper lobe branch.

D. Chest CT (lung window) coronal view demonstrates division of the trachea into three bronchi. The right upper lobar bronchus, bronchus intermedius, and left main bronchus originate from the carina. There is relative narrowing of the proximal portion of the bronchus intermedius (arrow).

E. Virtual bronchoscopic image reveals three origins of the RUB, BI, and LMB (arrows).

BI = bronchus intermedius, LMB = left main bronchus, RUB = right upper lobar bronchus



the left hemi-thorax, due to volume loss of the left lung (Fig. 1B). There was no left upper lobe parenchyma, bronchus, artery, or vein. Mild compensatory hyperinflation of the right lung was also observed. The main pulmonary artery trunk was enlarged in width and there was no left upper lobar artery (Fig. 1C). There was no vascular anomaly in the right hemithorax. The trachea divided into right upper lobar bronchus, bronchus intermedius, and left main bronchus at carina, which exhibited a unique trifurcation configuration (Fig. 1D). Bronchus intermedius showed focal narrowing at the site of origin. Tracheobronchial anomaly was explicitly displayed on virtual bronchoscopic image (Fig. 1E).

There was mild pulmonary hypertension (39 mm Hg) on transthoracic echocardiogram without any other congenital heart disease. He was diagnosed with a case of agenesis of the left upper lobe with tracheal trifurcation. After receiving conservative treatment at our hospital, the patient was discharged without further evaluation because of improved patient symptoms.

DISCUSSION

Pulmonary underdevelopment can range from mild pulmonary parenchymal hypoplasia to total bronchial tree and parenchymal agenesis. This developmental anomaly was first described by de Pozzis in 1673; the first proposed classification of pulmonary underdevelopment was introduced by Schneider in 1912 (Class I, Agenesis, total absence of bronchus and lung; Class II, Aplasia, rudimentary bronchus and lung; Class III, Hypoplasia, bronchial hypoplasia and variable but reduced amount of lung tissue) (2). Agenesis of one or two lobes of a single lung is an extremely rare condition; the exact incidence of this anomaly is unknown. According to Berrocal et al. (3), the most frequently affected lobe is the left upper lobe. Solitary lobar agenesis goes undetected or is incidentally detected in majority cases. However, patients with pulmonary agenesis may exhibit several accompanying congenital abnormalities, including cardiovascular diseases, as well as spinal, genitourinary, and musculoskeletal pathologies (4). Our patient exhibited left upper lobar agenesis with associated tracheobronchial anomaly - tracheal trifurcation.

Tracheal trifurcation is the most dramatic form of right tracheal bronchus, in which right upper lobe bronchus is displaced at the carina, with separate bronchus intermedius and left main bronchus originating at the same level (5). In our patient, tracheal trifurcation was distinct, revealing three even-sized bronchi at the subcarinal level, due to a reduction in the size of the left main bronchus that resulted from the absence of the left upper lobar bronchus. Patients with tracheal trifurcation typically are asymptomatic and the trifurcation is detected incidentally on imaging (6). Some patients may present with recurrent respiratory tract infection, which may progress to bronchiectasis and chronic bronchitis. Tracheal trifurcation can be diagnosed easily and rapidly by conventional CT and by three-dimensional (3D) reconstruction. It is important to identify this anatomical variant in patients requiring intubation (7). Because, the recognition of tracheal anomaly before induction of intubation can be helpful for determining optimal positioning of the endotracheal tube and it can provide accurate diagnosis especially when he/she suffers from complications due to surgery or tracheal intubation, such as lobar collapse.

Lobar agenesis, lobar collapse, and Swyer-James syndrome can be considered when the unilateral lung volume is reduced or mediastinal shifting on chest X-ray.

CT is a helpful diagnostic modality for identifying lobar agenesis. In lobar agenesis, the volume of the affected hemi-thorax is reduced; mediastinal shift towards the affected site and herniation of the contralateral lung can be observed (8). 3D reconstruction provides more and easier visualization of tracheobronchial anomaly than conventional CT and it can be particularly helpful in delineating abnormalities of the bronchi, as well as associated arterial and venous structures (4, 9). A virtual endoscopic image can be obtained through image reconstruction; this plays an important role in characterizing bronchial anatomy without invasive bronchoscopy.

In conclusion, lobar agenesis associated with tracheal trifurcation is an extremely rare anomaly. Chest CT and 3D reconstruction may be useful in diagnosing the extent of this anomaly and evaluating associated abnormalities.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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기관의 삼분지와 동반된 좌상엽 무형성: 증례 보고

김형섭 · 우정주* · 안진경

폐엽 무발생은 매우 드문 선천성 기형으로 폐엽 동맥, 폐정맥, 기관지 및 폐 실질이 없는 것이 특징이다. 저자들은 젊은 남자에서 좌측 상부 무형성과 동반되어 기관 용골 높이에서 기관이 세 개의 기관지로 나누어지는 독특한 증례를 접하였다. 복잡한 기관기관지 해부학은 삼차원 재구성 컴퓨터단층촬영과 가상 기관지 내시경 검사로 명확하게 증명하였다. 좌상엽 무형성과 동반된 기관의 삼분지는 매우 드문 선천성 기형이며 저자들이 아는 바로는 보고된 적이 없다.

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