

Cystic Lung Changes in a Thin Section CT in an Asymptomatic Young Adult with Unilateral Pulmonary Vein Atresia: A Case Report

폐동맥 폐쇄를 가진 무증상 젊은 성인: 고해상도 CT에서의 낭성 폐 변화

Seung Choul Lee, MD, Jeong Geun Yi, MD, Jeong Hee Park, MD

Department of Radiology, Konkuk University Medical Center, Seoul, Korea

Unilateral pulmonary vein atresia is a rare anomaly, usually associated with symptoms of recurrent hemoptysis and pneumonia in early childhood. Only one report of an asymptomatic adult patient can be found in the literature. We present the case of an asymptomatic patient with unilateral right pulmonary vein atresia in a 20-year-old man. Chest radiograph and multidetector computed tomography showed not only pulmonary vein atresia, pulmonary artery hypoplasia, but also cystic lung changes on thin section CT, along with septal and bronchovascular bundle thickening, and ground-glass opacity. Unilateral pulmonary vein atresia could be another disease which can show cystic lung changes on thin section chest CT.

Index terms

Pulmonary Vein Atresia
Computed Tomography
Cysts
Pulmonary Artery Hypoplasia

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Corresponding author: Jeong Geun Yi, MD
Department of Radiology, Konkuk University Medical Center, 120-1 Neungdong-ro, Gwangjin-gu, Seoul 143-729, Korea.
Tel. 82-2-2030-5544 Fax. 82-2-2030-5549
E-mail: jgyi92@gmail.com

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INTRODUCTION

Unilateral pulmonary vein atresia is a very rare congenital anomaly usually associated with high morbidity and mortality (1-4). Most of the patients reported were in infancy or early childhood with recurrent episodes of hemoptysis or pneumonia. Based on the gravity of symptoms, coil embolization and pneumonectomy are the usual forms of treatment (5, 6). There have been few radiologic reports about the findings of systemic collaterals to the pulmonary vessels such as interlobular septal thickening, ground-glass opacity and subpleural nodules (1, 2, 4, 7). We present the case of a 20-year-old male, who had been an asymptomatic pulmonary vein atresia and systemic-to-pulmonary collaterals patient. We observed cystic changes on right upper lobe of the lung, a rarely reported condition, and the septal thickenings, bronchovascular bundle thickening, and ground-glass opacity which represent peripheral collaterals on thin sec-

tion computed tomography (CT).

CASE REPORT

A 20-year-old previously asymptomatic male, visited our hospital for medical certificate. He has been told several years before, "your right thorax is small" when he took his chest radiograph.

A pulmonary function test was normal. A chest radiograph showed a small right hemithorax, small right pulmonary artery and, right sided mediastinal shift. The lung parenchyma showed diffuse ground-glass opacity, prominent reticulonodular densities and the pleura showed diffuse and irregular thickening. A large mass was suspected in the right lower lobe medial side (Fig. 1A). In the multi-detector CT, the right superior and inferior pulmonary veins were not visualized, and the right pulmonary artery was smaller than left one (Fig. 1B). There were

many hypertrophied bronchial and intercostal arteries from the ascending aorta that were connected with the pulmonary vessels at the peripheral subpleural area on coronal maximum intensity projection images (Fig. 1B). In the thin section CT, many cysts were on the right lung, more on upper lobe and peripheral side (Fig. 1D). Beaded interlobular septal thickening, bronchovascular bundle thickening, nodularities along the fissures and ground-glass opacity were noted in the right lung (Fig. 1E, F). The mass density on right lower lung zone was proved to be a herniated liver. No further evaluation was done.

DISCUSSION

Pulmonary vein atresia is divided into three categories ac-

ording to the extent of pulmonary vein involvement; the common, unilateral and individual pulmonary vein atresia, (3, 5). Among these three categories, unilateral pulmonary vein atresia can occur in either side of lung without right or left side predominancy (1-3). Unilateral pulmonary vein atresia without associated cardiac structural abnormalities is a rare occurrence and fewer than 40 cases have been reported in the literature (1-4). Most patients were diagnosed during infancy or early childhood due to episodes of recurrent pneumonia, hemoptysis and pulmonary hypertension due to the systemic collateral connection to pulmonary vessels (5). About 50% of patients had fatal results if they did not receive that did not receive treatment did not survive (6). Due to the serious pulmonary symptoms, such as hemoptysis, recurrent pneumonia, and progressive dyspnea

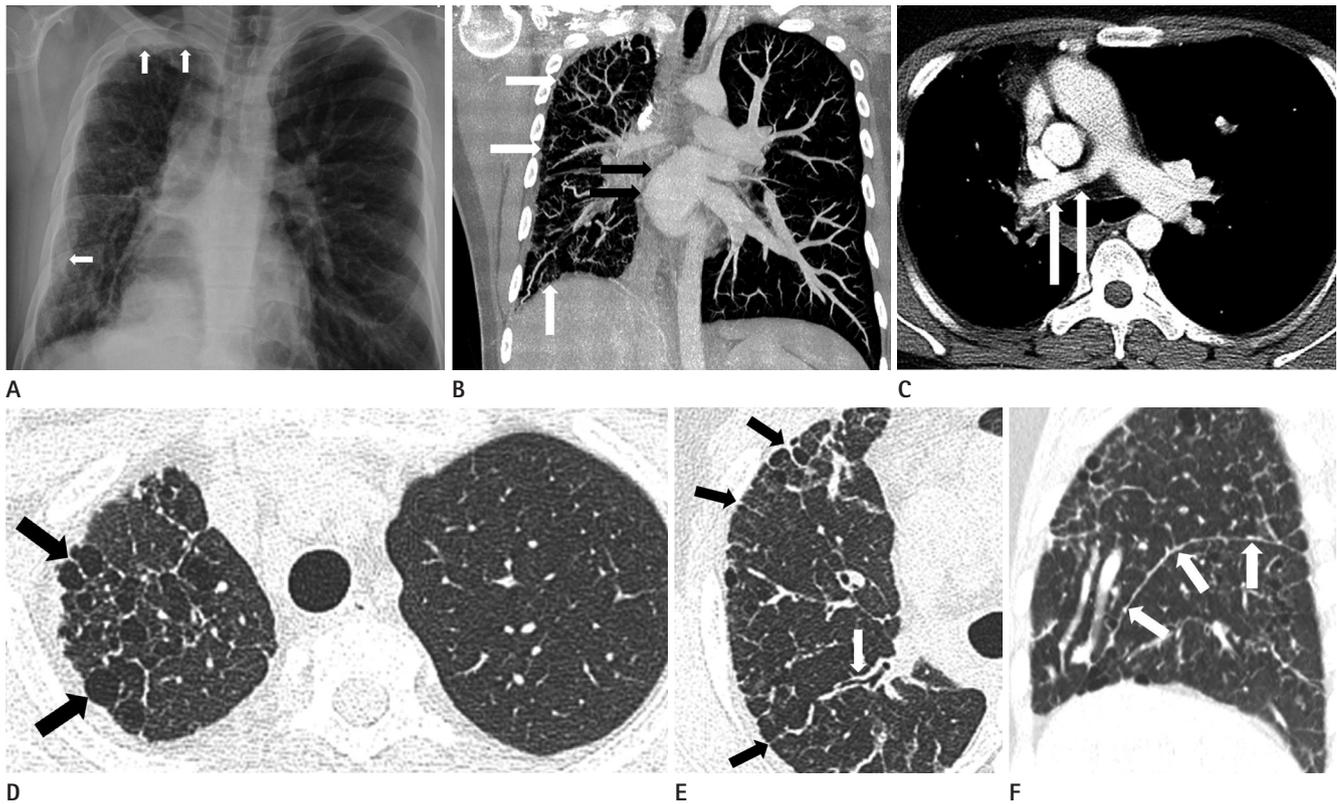


Fig. 1. The 20-year-old man with unilateral pulmonary vein atresia performed chest radiography and MDCT. **A.** The chest radiograph shows small right hemithorax and pulmonary artery. Right lung shows reticulonodular densities, thickened interlobular septa and pleural thickening (white arrows). **B.** Left atrium shows smooth margin and right pulmonary veins are absent (black arrows). Many hypertrophied systemic collaterals are subpleurally connected to pulmonary vessels (white arrows) in coronal MIP image of MDCT. **C.** Right pulmonary artery is hypoplastic (white arrows) in MDCT axial image. **D.** Thin section CT shows many variable sized cysts (arrows), beaded septal thickenings in right upper lobe. **E.** Thin section CT shows beaded septal thickenings in subpleural areas (black arrows) and bronchovascular bundle thickening (white arrow). Note the subpleural location of lung cysts and various thickness of cysts wall in lower level of the CT section. **F.** Thin section CT shows also nodular densities (arrows) along major fissure on sagittal reformatted image and subpleural location of lung cysts. Note.—MDCT = multi-detector CT, MIP = maximum intensity projection

or for the prevention of pulmonary hypertension, pneumonectomy is the usual curative treatment of choice (1, 3, 7, 8). Six adult patients and only one asymptomatic adult patient are described in the literature (1).

Due to the small pulmonary artery and absent pulmonary vein, systemic-to-pulmonary collaterals are prominent in the affected lung (Fig. 1B, C). Many hypertrophied bronchial and intercostal arteries and veins are connected to pulmonary vessels at the peripheral areas of the lung, such as subpleural and interfissural areas. In a thin section CT, beaded interlobular septal thickening, bronchovascular bundle thickening, ground-glass opacity, and fissural/pleural nodularity represent these systemic collaterals (engorged draining veins, lymphatics and hypertrophied systemic arteries) (Fig. 1E, F) (1, 2, 4, 7).

Interestingly, we observed the cysts in the affected lung dominantly on the upper lobe, and more on subpleural areas (Fig. 1D, E). Up until now, three lung cysts cases were described in the literature in pulmonary vein atresia patients; in an asymptomatic unilateral pulmonary vein atresia (1), in a child with cardiac anomaly (8) and, in a bilateral Scimitar syndrome (9). The current patient with lung cysts (Fig. 1D) is the second case to follow the one by Kim et al. (1), which focused on the asymptomatic adult patients. Kim et al. (1) postulated that these unique CT features of lung cysts are the result of the destruction or underdevelopment of capillary networks at the alveolar level due to poor arterial supply or pressure damage from collaterals and somehow the inevitable results of this disease entity (1). Upper lobe dominant cysts could be explained by the poor oxygen supply of upper lobe as compared to the lower lobe, which is required for the normal development of alveoli. Chronic pressure damage from systemic collaterals in the subpleural area in adult patient which could possibly hamper the growth of alveolar tissue, and result in cysts formation (1).

The thickness of the current cysts wall is variable and a larger portion of subpleural cysts are thicker than 2 mm, with many cysts that share their wall with thickened interlobular septa (Fig. 1D). When we consider the definition of lung cysts from the Fleischner society, round parenchymal lucency with variable wall thickness usually less than 2 mm, current cyst walls are more likely to be composed, not just fibrous tissue of cyst, but also with collateral vessels (Fig. 1D, E) (10). So, these lung cysts in unilateral pulmonary vein atresia patients could possi-

bly get thicker and thicker as the disease progresses and collateral vessels grow.

We report the radiographic and CT findings of unilateral pulmonary atresia developed in a young adult male without symptoms. Along with findings of pulmonary vein atresia, we observed the cysts in affected lung, more on upper lobe and subpleural areas. These cystic lung lesions seem to be the late radiologic findings of unilateral pulmonary vein atresia and should be included in the differential diagnosis of cystic lung diseases on thin section CT.

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폐동맥 폐쇄를 가진 무증상 젊은 성인: 고해상도 CT에서의 낭성 폐 변화

이승철 · 이정근 · 박정희

편측 폐정맥 폐쇄는 매우 드문 기형으로, 보통 소아기에 반복되는 객혈이나 폐렴과 같은 증상을 동반한다. 지금까지 단지 1예의 무증상 성인 환자의 문헌 보고가 있다. 우리는 우측 폐정맥 폐쇄가 있지만, 증상이 없는 20세 남자 환자를 경험했다. 흉부 방사선 사진과 다검출기 CT에서 폐정맥 폐쇄, 폐동맥 저형성뿐 아니라, 낭성 폐 변화, 소엽 간 중격과 혈관 기관지 속의 비후, 간유리 음영이 고해상도 CT에서 보였다. 편측 폐정맥 폐쇄는 고해상도 CT에서 낭성 폐 변화를 보일 수 있는 또 다른 질환일 수 있다.

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