



Isolated Unilateral Pulmonary Artery Hypoplasia with Accompanying Pulmonary Parenchymal Findings on CT: A Case Report

폐병변으로 나타난 고립성 편측성 폐동맥발육부전의 CT소견: 증례 보고

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Unilateral pulmonary artery hypoplasia or agenesis without congenital cardiovascular anomalies is rare in adults. We report a case of a 36-year-old man with isolated left unilateral pulmonary artery hypoplasia with recurrent hemoptysis. On computed tomography (CT), the left pulmonary artery showed hypoplasia with multiple collateral vessels seen in the mediastinum and the left hemithorax. Also, parenchymal bands and peripheral linear opacities were seen in the affected lung, which were probably due to chronic infarction induced by unilateral pulmonary artery hypoplasia. There are only a few reports focusing on the radiologic findings in the pulmonary parenchyma induced by unilateral pulmonary artery hypoplasia, such as parenchymal bands and peripheral linear opacities. Therefore we report this case, which focused on the CT findings in the pulmonary parenchyma due to isolated unilateral pulmonary artery hypoplasia.

Index terms

Pulmonary Artery
Hypoplasia
Hemoptysis
Multidetector Computed Tomography

Received June 16, 2016

Revised July 4, 2016

Accepted September 8, 2016

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INTRODUCTION

Unilateral pulmonary artery hypoplasia occurs due to an embryologic developmental failure in the bud of the sixth aortic arch (1-3). Unilateral pulmonary artery hypoplasia is frequently associated with cardiovascular anomalies (3), but unilateral pulmonary artery hypoplasia without a cardiovascular anomaly is called 'isolated' unilateral pulmonary artery hypoplasia and this entity is much less common (2, 4). Common presentations include shortness of breath, recurrent pulmonary infections, and hemoptysis (5); however, some patients are asymptomatic. Due to the nonspecific nature of the symptoms and imaging findings,

it is easy to misdiagnose lung parenchymal change induced by unilateral pulmonary artery hypoplasia as other lung parenchymal diseases or pneumonia. There are only a few reports focusing on the radiologic findings in the pulmonary parenchyma, such as parenchymal bands caused by unilateral pulmonary artery agenesis (6). Therefore, this report describes the computed tomography (CT) findings in the pulmonary parenchyma due to isolated unilateral pulmonary artery hypoplasia.

CASE REPORT

A 36-year-old male was admitted to our hospital for the sec-

ond time due to recurrent hemoptysis. He was an ex-smoker with a smoking history of 10 pack-years. He visited our hospital due to an episode of hemoptysis. The initial chest CT showed multifocal parenchymal bands in the left lung with patchy ground glass opacities, which were considered to be caused by

aspirated blood. Initially it was thought that the parenchymal bands occurred due to previous infectious sequelae, and a definite diagnosis was not made at that time. He underwent bronchial artery embolization, which improved his symptoms.

During his second visit to our hospital, chest radiography (Fig.

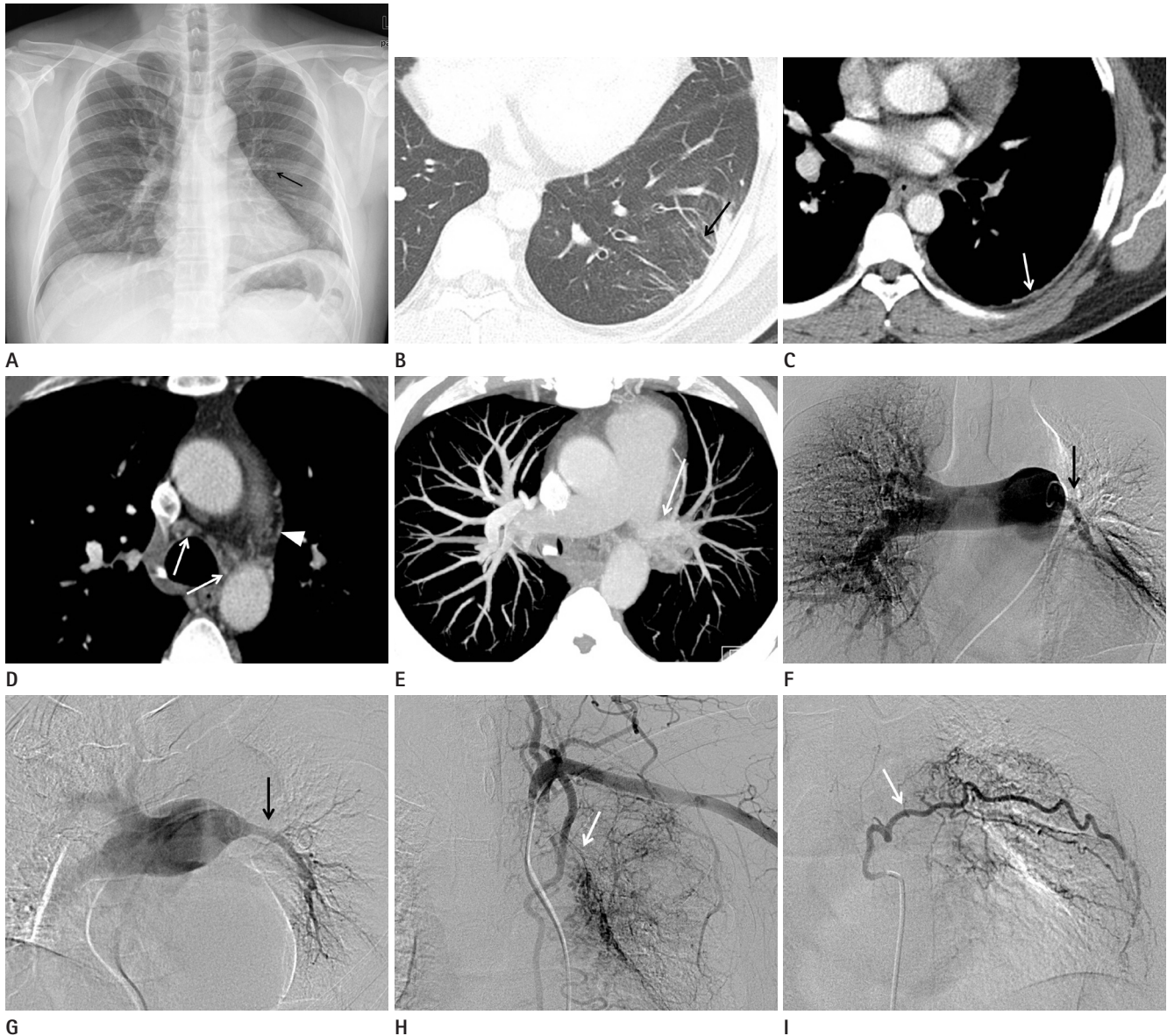


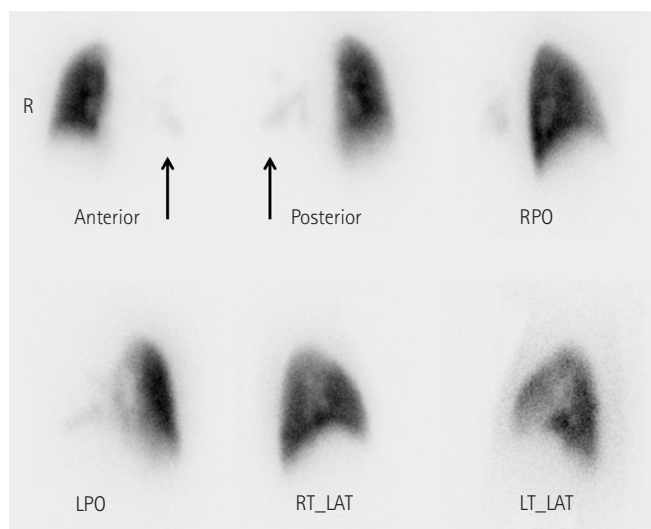
Fig. 1. Isolated unilateral pulmonary artery hypoplasia in a 36-year-old man who presented with hemoptysis.

A. PA chest radiograph shows a small left hilum (arrow) compared to the right hilum. The left hemidiaphragm is slightly elevated, but mediastinal structures are not displaced.

B-E. On chest CT with lung setting (**B**), parenchymal bands are seen in the posterolateral portion of the left lower lobe (arrow). Mediastinal setting of chest CT (**C**, **D**) shows serrated pleural thickening in the left posterolateral portion of the hemithorax (arrow). Tortuous prominent collateral vessels are noted in the mediastinum (arrows, arrowhead). Maximum intensity projection image (**E**) shows a hypoplastic left pulmonary artery (arrow).

F-I. Pulmonary angiography (**F**, **G**) shows a small left pulmonary artery (arrows), compared to the right pulmonary artery. Left subclavian angiography (**H**) shows fine systemic collateral vessels (arrow) that supply the left lung. Left intercostal angiography (**I**) shows collateral branches of the left intercostal artery (arrow) with abnormal lung parenchymal staining.

CT = computed tomography



J

Fig. 1. Isolated unilateral pulmonary artery hypoplasia in a 36-year-old man who presented with hemoptysis.

J. The lung perfusion scan shows a near total perfusion defect (arrows) in the left lung.

1A) showed decreased left-sided pulmonary vascularity, especially in the hilar area, and subtle elevation of the left hemidiaphragm, but no definite mediastinal shifting. A chest CT showed parenchymal bands and peripheral linear opacities in the left lung, which might be due to chronic lung infarction. Serrated pleural thickening was seen in the left hemithorax, corresponding to dilatation of the intercostal arteries. Left pulmonary artery diameter was small with decreased left-sided pulmonary vasculature. There were multifocal hypertrophied bronchial artery collateral vessels. Maximum intensity projection image showed a hypoplastic left pulmonary artery (Fig. 1B-E).

Pulmonary arteriography showed hypoplasia of the left pulmonary artery. Angiography showed that multifocal collateral arteries had arisen from the left subclavian artery and the left intercostal arteries (Fig. 1F-I). The patient received embolization of the collateral arteries via the left bronchial and left upper intercostal arteries, with PVA particles. Hemoptysis was completely resolved after embolization.

The patient was not aware of any childhood pulmonary infections or any specific family history of pulmonary disease. Trans-thoracic echocardiography (TTE) and a lung perfusion scan were performed for further evaluation of unilateral pulmonary artery hypoplasia. TTE revealed a normal left ventricular systolic function with normal valvular morphology. TTE did not show any septal defects or other cardiovascular anomalies or any evidence

of pulmonary arterial hypertension. The lung perfusion scan showed a near total perfusion defect in the left lung (Fig. 1J). Hemoptysis was controlled and the patient was discharged from the hospital.

DISCUSSION

Unilateral pulmonary artery hypoplasia or agenesis is a rare congenital anomaly with an incidence of 1 in 300000. It occurs due to an embryologic developmental failure in the bud of the sixth aortic arch (3). It is frequently associated with other congenital anomalies, such as tetralogy of Fallot, a ventricular septal defect, transposition of great vessels, and aortic arch anomalies (2). Unilateral pulmonary artery agenesis as an isolated phenomenon without congenital cardiovascular anomalies is even more rare (7).

Terminologies related to unilateral pulmonary artery hypoplasia such as agenesis, aplasia, complete absence, and/or stenosis have been used interchangeably. Angiographic evidence was used to diagnose unilateral pulmonary artery aplasia by confirming the absence of the ipsilateral pulmonary artery. However, failure to fill the ipsilateral pulmonary artery with contrast material does not always indicate the absence of the pulmonary artery. At thoracotomy or on postmortem examination, a patent and small pulmonary artery has been observed even when there was no filling of the pulmonary artery in the previous pulmonary arteriography. This might be caused by increased bronchial arterial flow (1). It is unclear whether unilateral pulmonary artery hypoplasia is congenital or acquired (1). However, certain imaging characteristics may be useful in differentiating between congenital absence and hypoplasia of the pulmonary artery. Patients with acquired unilateral hyperlucent lungs have bronchiectasis or other destructive processes in the lungs with markedly impaired ventilation and mediastinal swing during inspiration. Patients with congenital unilateral pulmonary artery hypoplasia show normal ventilation in the affected lung and none or minimal mediastinal swing. In addition, a patient with congenital unilateral pulmonary artery hypoplasia shows more abundant systemic collateral vessels throughout the affected lung than a patient with acquired unilateral hyperlucent lungs (1). Therefore, we believe that the imaging findings in our patient were likely caused by congenital abnormalities.

Common presentations of unilateral pulmonary artery hypoplasia include shortness of breath, recurrent pulmonary infections, and hemoptysis (5); however, some patients are asymptomatic. Hemoptysis has been reported in approximately 18–20% of patients with unilateral pulmonary artery hypoplasia (8). Because of the nonspecific symptoms and imaging findings, it is easy to misdiagnose parenchymal change induced by unilateral pulmonary artery hypoplasia as other lung parenchymal diseases or pneumonia. In our case, initially it was thought that the parenchymal bands occurred due to the previous infectious sequelae, which delayed the diagnosis of unilateral pulmonary artery hypoplasia. CT findings of chronic lung infarction may include parenchymal bands, wedge-shaped opacities or irregular peripheral linear opacities (8). In the study by Sakai et al. (6), in eight patients with unilateral pulmonary artery agenesis, CT findings such as serrated pleural thickening were observed in six patients (75%), subpleural parenchymal bands in five (63%), and mosaic attenuation in three (38%) on the affected lung. In our case, parenchymal bands, serrated pleural thickening, and peripheral linear opacities were seen on CT altogether. These findings were probably due to parenchymal changes induced by unilateral pulmonary artery hypoplasia.

There is no general consensus on the treatment of unilateral pulmonary artery hypoplasia (9). However, there are several case reports of unilateral pulmonary artery hypoplasia with hemoptysis that were successfully treated with variable therapeutic options, such as pneumonectomy, vasodilator therapy, and embolization of collateral arteries (10).

In conclusion, if a patient presents with parenchymal findings, such as parenchymal bands, as well as hypertrophied collateral vessels, the possibility of unilateral pulmonary artery hypoplasia should be considered.

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폐병변으로 나타난 고립성 편측성 폐동맥발육부전의 CT소견: 증례 보고

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선천성 심혈관 기형을 동반하지 않은 편측성 폐동맥발육부전 및 무발생은 성인에게서 드문 질환이며, 이러한 경우를 고립성 폐동맥발육부전이라고 한다. 반복되는 객혈을 주소로 한 컴퓨터단층촬영에서 폐실질에 이상소견을 보인 36세 남성의 고립성 폐동맥발육부전 증례를 보고한다. 흉부 컴퓨터단층촬영에서 왼쪽 폐동맥의 발육부전과 여러 측부혈관들이 관찰되었고, 더불어 고립성 폐동맥 발육부전으로 인한 만성 폐경색과 관련된 것으로 보이는 폐실질밴드와 폐 주변부 선상음영도 관찰되었다. 고립성 폐동맥발육부전의 경우, 폐실질밴드와 폐 주변부 선상음영과 같은 폐실질의 영상 소견에 주안점을 둔 보고는 매우 드물다. 따라서 이번 증례에서 고립성 폐동맥발육부전에 의한 폐실질의 컴퓨터단층촬영 소견을 보고한다.

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