Case Report

Isolated congenital interruption of the right interlobar pulmonary artery with unilateral interstitial lung abnormality

Mitsukuni Sakabe a,*, Kazunori Tobino a, b, Kazuki Uchida a, Yosuke Murakami a, Yuri Hiramatsu a, Hiroaki Ota a, Takafumi Kawabata a

a Department of Respiratory Medicine, Iizuka Hospital, 3-83 Yoshiomachi, Iizuka, Fukuoka, 820-0018, Japan
b Department of Respiratory Medicine, Juntendo University, School of Medicine, 2-1-1 Bunkyo-Ku, Hongo, 113-8421, Japan

ARTICLE INFO

Keywords:
Congenital interruption of right interlobar pulmonary artery CT
Interstitial lung abnormality

ABSTRACT

We report a case of an isolated congenital interruption of the right interlobar pulmonary artery with unilateral interstitial lung abnormality. 3D-CT with enhancement showed absent right interlobar pulmonary artery without any other abnormalities of the pulmonary artery and an enlarged inferior phrenic artery. High-resolution CT demonstrated ground-glass opacities, reticular changes, and small cysts in the right middle and lower lobes, which were compatible with interstitial lung abnormality. The patient was diagnosed with an isolated congenital interruption of the right interlobar pulmonary artery since chronic pulmonary thromboembolism, structural heart disease, systemic congenital disease, and systemic vasculitis were ruled out.

1. Introduction

Isolated congenital interruption of the right interlobar pulmonary artery is an extremely rare condition. To the best of our knowledge, only one case report has described the chest computed tomography (CT) findings of this condition, which showed unilateral interstitial changes in the middle and lower lobes of the right lung. In this report, usual interstitial pneumonia (UIP) pattern-like lesions such as bronchial dilatation, ground-glass opacities, reticular changes, and honeycombing in the right lower lung lobe were observed. These abnormalities were thought to be due to changes in blood flow owing to pulmonary artery defects. We report a case with CT imaging findings that differ from the previous report.

2. Case presentation

A 62-year-old female patient was referred to our hospital for suspected recurrent pulmonary embolism (PE). She complained of dry cough and chest pain for 2 days. Eighteen months previously, she was found to have abnormal electrocardiography (ECG) patterns (i.e., nonspecific T-wave abnormalities) without any symptoms and was diagnosed with PE of the right interlobar pulmonary artery on CT perfusion scan. Since then, she had been treated with warfarin; however, her condition and ECG patterns remain unchanged. She had no other medical history and had never smoked.

Her oxygen saturation was 94% at room air and her other vital signs were normal. A physical examination revealed normal breathing and cardiac sounds and no edema. Laboratory test values were as follows: white blood cells, 5660/mm³; hemoglobin, 11.6 g/

Abbreviations: ECG, Electrocardiography; PE, Pulmonary embolism; UIP, Usual interstitial pneumonia.

* Corresponding author.
E-mail address: msakabeh1@aih-net.com (M. Sakabe).

https://doi.org/10.1016/j.rmcr.2022.101734
Received 15 March 2022; Accepted 31 August 2022
Available online 6 September 2022

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dl; platelets, 115,000/mm$^3$; serum lactate dehydrogenase, 223 U/l (normal, 119–229 U/l); serum aspartate and alanine aminotransferase, 34 U/l and 19 U/l (normal, 0–35 U/l); serum albumin, 2.8 g/dl (normal, 4–5 g/dl); serum C-reactive protein, 0.57 mg/dl (normal, < 0.2 mg/dl); serum D-dimer, < 0.5 μg/ml (normal, < 0.5 μg/ml); and brain natriuretic peptide, 631.2 pg/ml. Serum protein

Fig. 1. Chest radiograph shows reticular opacities in the right middle and lower lung field without volume change.

Fig. 2. A and B, Contrast-enhanced chest CT (LightSpeed 16, General Electric Medical Systems, Milwaukee, WI) with coronal reconstruction and 3D CT shows abrupt interruption of the right interlobar artery (white arrow) without endoluminal changes, and a normal-sized right main pulmonary artery and right upper lobe artery. There is no evidence of multiple bilateral pulmonary arterial abnormalities usually seen in chronic pulmonary thromboembolism. C, Contrast-enhanced chest CT with coronal reconstruction shows prominent right inferior phrenic artery (white arrow).
C and protein S activity were normal, and antinuclear antibody, anti-phospholipid antibody, PR3-ANCA and MPO-ANCA assays were negative. ECG showed nonspecific T-wave abnormalities. Transthoracic echocardiography revealed normal chamber sizes, normal left ventricular function, and no evidence of other structural heart diseases. Chest radiograph showed reticular changes in the right middle and lower lung fields without volume change (Fig. 1). 3D-CT with enhancement showed absent right interlobar pulmonary artery without any other abnormalities of the pulmonary artery, and an enlarged inferior phrenic artery (Fig. 2). High-resolution CT demonstrated ground-glass opacities, reticular changes, and small cysts in the right middle and lower lobes (Fig. 3), which were compatible with interstitial lung abnormality. Lung perfusion scan showed total defect in the right middle and lower lobes (Fig. 4). The patient was diagnosed with an isolated congenital interruption of the right interlobar pulmonary artery, since chronic PE, structural heart disease, systemic congenital disease, and systemic vasculitis were ruled out. Her symptoms improved on the 2nd day of admission, and her condition stabilized without any treatment. Therefore, it was considered that there was no relationship between her symptoms and the isolated congenital interruption of the right interlobar pulmonary artery.

3. Discussion

To the best of our knowledge, there is only one case report on the CT findings of an isolated congenital interruption of the right interlobar pulmonary artery [1]. Chest radiographic findings showed a normal lung volume, fine reticular opacities in the right middle and lower lung field, and similar size of both hilum. Chest CT findings were described as UIP pattern-like changes such as bronchial dilatation, ground-glass opacities, reticular changes, and honeycomb cysts in the right middle and lower lobes. Multiple enlarged collateral systemic arteries without patent ductus arteriosus were also observed.
The pathophysiology for this condition may be related to the partial involution of the proximal sixth aortic arch [1]. The blind end of the pulmonary artery at the hilum may cause the anastomoses of pulmonary vessels to systemic arteries. The idiopathic pulmonary fibrosis-like changes seen are thought to represent lung injury (i.e., ischemia, infarction, bleeding and inflammatory change) caused by the pressure gradient between these vessels or a high oxygen saturation level [1]. Our patient also had an enlarged inferior phrenic artery distributed to the right lower lobe, but the pulmonary parenchymal change was thought to be different from the previous case. The difference in the CT appearance between our patient and the previously reported case was probably due to the difference in the grade of the lung injury.

The most important differential diagnosis was chronic PE in which the presence of multiple bilateral arterial abnormalities and occlusion of one main pulmonary artery mimic proximal interruption, which has been reported in only 3% of cases [2,3]. Our patient did not manifest these findings and warfarin therapy was not effective. Other differential diagnoses were pulmonary artery involvement in Takayasu’s arteritis or Behçet’s disease, for which our patient did not meet the diagnostic criteria [4,5].

4. Conclusion

We describe the rare case of unilateral interstitial lung abnormality due to isolated congenital interruption of the right interlobar pulmonary artery. We believe that our case will add to our understanding and recognition of the spectrum of this rare condition.

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