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Pulmonary hypertension due to silicosis and right upper pulmonary artery occlusion with bronchial anthracofibrosis

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ABSTRACT

Bronchial anthracofibrosis is a rare disease defined as bronchial stenosis with black pigmentation and usually not associated with artery occlusion. The patient was an 81-year-old man with silicosis. He presented with dyspnea on exertion, and pulmonary hypertension due to right upper pulmonary artery occlusion without thromboembolism was diagnosed on the basis of the results of right heart catheterization and pulmonary angiography. Bronchoscopy demonstrated bronchial anthracofibrosis in the right upper lobe. These findings suggested that the cause of PH was silicosis and pulmonary artery occlusion with bronchial anthracofibrosis. He has been treated with home oxygen therapy and tadalafil, and his symptom and 6MWD remain stable.

1. Introduction

Bronchial anthracofibrosis is defined as bronchial stenosis with black pigmentation in the overlying mucous membrane; its causes include tuberculosis and dust exposure [1–3]. It is usually not associated with artery occlusion. On the other hand, pulmonary hypertension (PH) can occur with pneumoconiosis, which is associated with interstitial lung disease and hypoxic vasoconstriction [4]. Here, we report a rare case of PH due to silicosis and right upper pulmonary artery occlusion with bronchial anthracofibrosis. The imaging findings suggested that the bronchial anthracofibrosis was associated with the right upper pulmonary artery occlusion.

2. Case report

The patient was an 81-year-old man who had worked in a coal mine from the age of 20 years–30 years. Since receiving a diagnosis of silicosis and benign asbestos pleurisy based on thoracoscopic findings in 2011, he had been seen regularly at our hospital. During that time, no change was observed in the chest computed tomographic (CT) images. Between April and June 2015, he was repeatedly hospitalized for pneumonia. After the discharge in June 2015, he presented with dyspnea on exertion. Pulmonary function testing showed decreases in forced expiratory volume % in 1 second (FEV1%, 63.9%) and in diffusing capacity of the lungs for carbon monoxide/alveolar volume (DL_{CO}/VA, 2.91 mL/min/mmHg/L, 71.3%). Ultrasonic echocardiography (UCG) demonstrated an increase in the tricuspid regurgitation pressure gradient (TRPG, 48 mmHg). PH associated with silicosis was suspected, and he was hospitalized for further investigation and treatment.

The patient had a medical history of herpes zoster and was a former smoker (15 pack years). On admission, his temperature was 36.0 °C; blood pressure, 105/56 mmHg; pulse rate, 90 bpm; and saturation of percutaneous oxygen (SpO_2) in room air, 96%. His 6-min walk distance (6MWD) was 246 m, and the SpO2 was reduced to 88%. The physical finding was rhonchi in both lung fields.

The laboratory findings demonstrated increases in N-terminal pro-brain natriuretic peptide (NT-proBNP, 444.3 pg/mL), D-dimer (1.5 μg/mL), and C-reactive protein (CRP, 4.88 mg/dL). Autoantibody test findings for connective tissue disease and biomarkers of infection with HIV and mycobacterium were all negative. The bacterial and mycobacterial culture findings were also negative.

A chest X-ray image showed a nodular shadow in the right upper lung field, multiple granular shadows in both lungs, and right pleural effusion (Fig. 1). The chest CT images revealed mediastinal and hilar lymphadenopathies with egg-shell calcification and bronchial stenosis with a nodular shadow and lymphadenopathy in the right upper lobe. These imaging findings were no different from the previous ones. There was no finding suspicious for other diseases excluding silicosis. The

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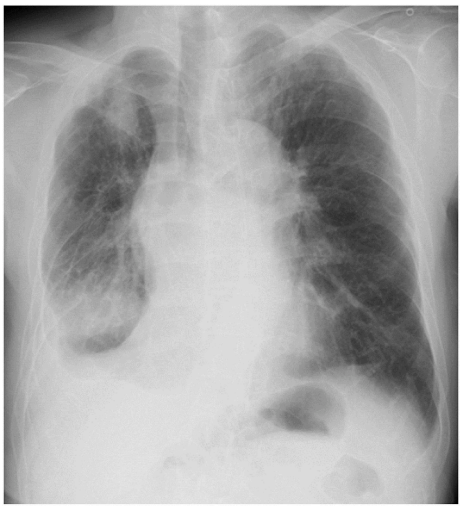

Fig. 1. Chest image showing a nodular shadow in the right upper lung field, multiple granular shadows in both lungs, and right pleural effusion.

contrast-enhanced CT showed right upper pulmonary arterial stenosis and main pulmonary artery dilation (Fig. 2). The ratio of the main pulmonary artery diameter to the ascending aorta diameter (PA:A ratio) was larger than 1. Thrombi were not found in the arteries.

A lung ventilation scintigram showed no abnormality (Fig. 3A). However, a lung perfusion scintigram showed a perfusion defect in the right upper lobe (Fig. 3B). Right heart catheterization (RHC) test results showed the following: mean pulmonary arterial pressure (mPAP) 32 mmHg, pulmonary vascular resistance (PVR) 4.69 Wood units, pulmonary artery wedge pressure (PAWP) 12 mmHg, and cardiac index (CI) 2.62 L/min/m^2. A pulmonary angiogram revealed right upper pulmonary artery occlusion without thrombi (Fig. 3C). In a coronal slice image of the chest CT, the right upper pulmonary artery was compressed extrinsically (Fig. 3D). Therefore, PH due to silicosis and right upper pulmonary artery occlusion without thromboembolism was diagnosed.

The bronchoscopic images demonstrated bronchial stenosis with carbon powder deposits, which were findings of bronchial anthracofibrosis, mainly in the right upper lobe (Fig. 4). This location was consistent with right upper pulmonary artery occlusion, which suggested that the bronchial anthracofibrosis was associated with the right upper pulmonary artery occlusion.

On the basis of a diagnosis of PH due to lung disease, the patient was treated with home oxygen therapy on exertion and tadalafil 20 mg/ daily. Half a year later, there was no change in mPAP (32–33 mmHg) in the RHC test results. However, his symptom and 6MWD were improved (246–285 m). The patient remains stable.

3. Discussion

In this report described a rare case of PH due to silicosis and right upper pulmonary artery occlusion with bronchial anthracofibrosis.

Chung et al. reported the bronchoscopic findings of bronchial stenosis with black pigmentation in the overlying mucous membrane and defined the findings as bronchial anthracofibrosis [1]. The bronchial stenosis was associated with peribronchial cuffing of the soft tissue or surrounding lymph nodes on chest CT. More than 60% of the patients (17/28) had active tuberculosis. Wynn et al. [2] and Naccache et al. [3] reported a case series of anthracofibrosis associated with exposure to

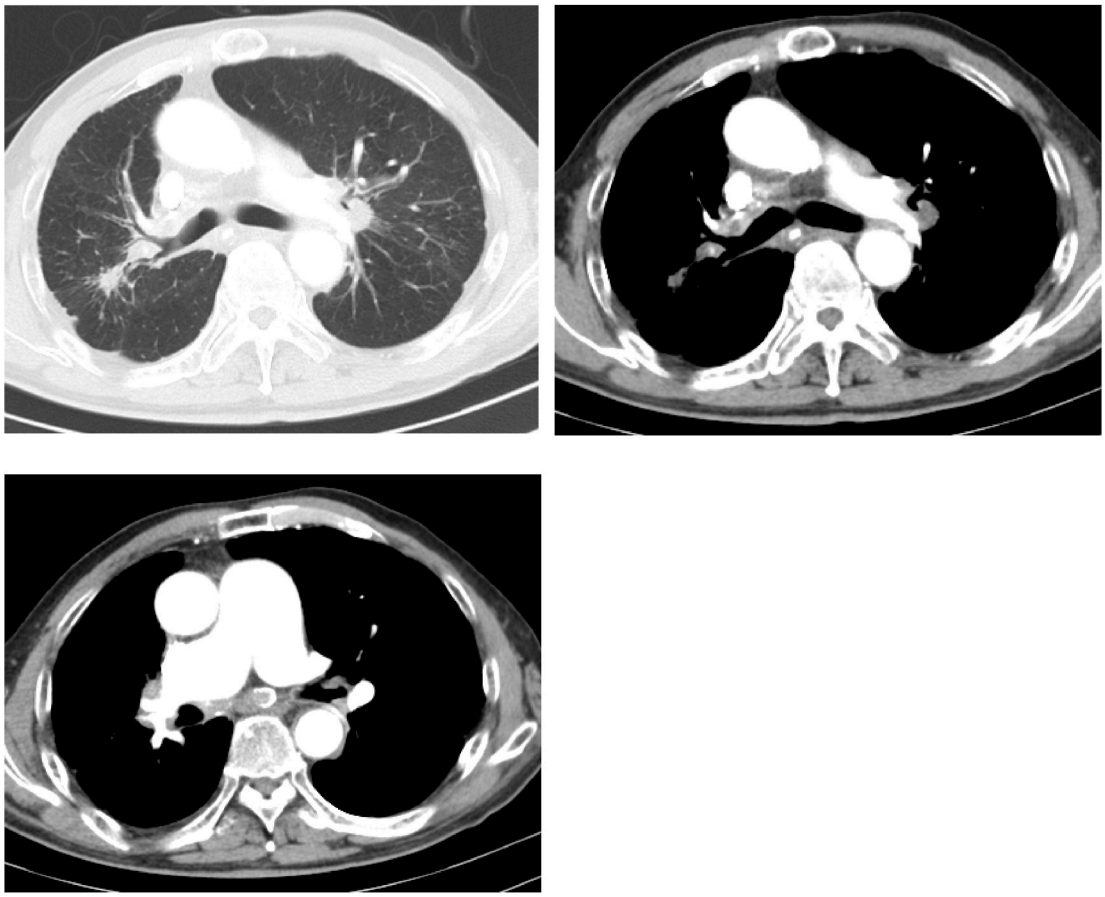

Fig. 2. Chest CT images showing mediastinal and hilar lymphadenopathies with egg-shell calcification, pulmonary arterial and bronchial stenosis with a nodular shadow, and lymphadenopathy in the right upper lobe, and main pulmonary artery dilation.

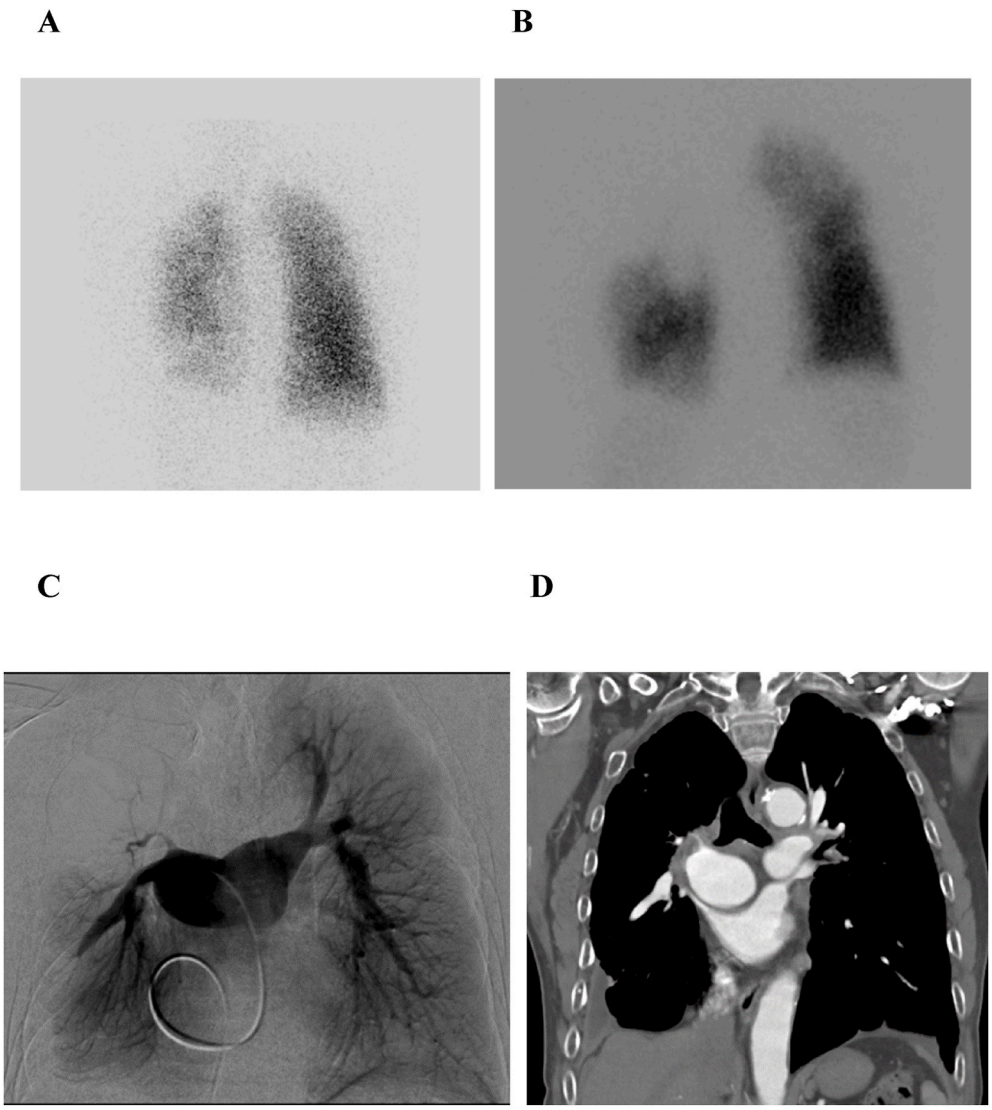

Fig. 3. (A) Lung ventilation scintigram showing no abnormality. (B) Lung perfusion scintigram showing a perfusion defect in the right upper lobe. (C) Pulmonary angiogram revealing right upper pulmonary artery occlusion. (D) Coronal slice image of the chest CT. The right upper pulmonary artery was compressed extrinsically without thrombi.

biomass fuel and mineral dust including silica.

The mechanism of bronchial anthracofibrosis remains unclear. The pathologic findings suggest that the lymph nodes compress or invade the adjacent bronchi, leading to fibrosis [1,3]. Although the relationship between anthracofibrosis and arterial occlusion is unknown, a case of bronchial anthracofibrosis with right lower pulmonary artery stenosis attributed to mixed mineral dust exposure including silica has already been reported [3]. On the other hand, pulmonary artery stenosis due to non-malignant lymphadenopathy rarely causes PH. However, there were some reports in patients with silicosis [5] and enlargement of anthracotic hilar lymph nodes [6]. Zelko et al. reported a mouse model of PH due to pulmonary fibrosis induced by crystalline silica [7]. Silica promoted the pulmonary vascular damage through mechanisms that might be involved in endothelial dysfunction, inflammation and vascular remodeling. In these reports, silica was associated with bronchial anthracofibrosis and pulmonary artery stenosis with silicosis. In our case, the patient had already received a diagnosis of silicosis. The lung lesions and mediastinal and hilar lymphadenopathies in the chest CT were no different from the previous ones. There was no finding suspicious for the other diseases. Therefore, the bronchial anthracofibrosis in the right upper lobe might have been caused by extrinsic compression from the lymphadenopathy owing to silicosis. The cause of pulmonary artery occlusion in the same location was also suspected to be the extrinsic compression from the lymphadenopathy. Finally, a diagnosis was made of PH due to silicosis and right upper pulmonary artery occlusion with bronchial anthracofibrosis associated with silicosis.

PH secondary to pneumoconiosis tends to correlate with the severity of the lung abnormality, but its prevalence is unknown [4]. The relationship between PH and bronchial anthracofibrosis is also unknown. Ko et al. reported that 11 of 21 patients (52%) with bronchial anthracofibrosis had UCG findings of PH, the criterion for which was mPAP >25 mmHg [8]. No significant difference was found in the diagnosis of UCG and CT when the CT criterion was a main pulmonary artery diameter >33 mm or a PA:A ratio >1. Although an RHC test result was unavailable, bronchial anthracofibrosis may be a high-risk factor for PH.

No specific and curative treatment for silicosis [9] or bronchial anthracofibrosis [10] exists. Zhang et al. reported the effect of percutaneous pulmonary artery stenting and balloon angioplasty for a PH patient with silicosis-induced pulmonary artery stenosis [5]. If the main cause of PH is pulmonary stenosis with silicosis, these treatments for PH may improve the clinical condition. In our case, the causes of PH were

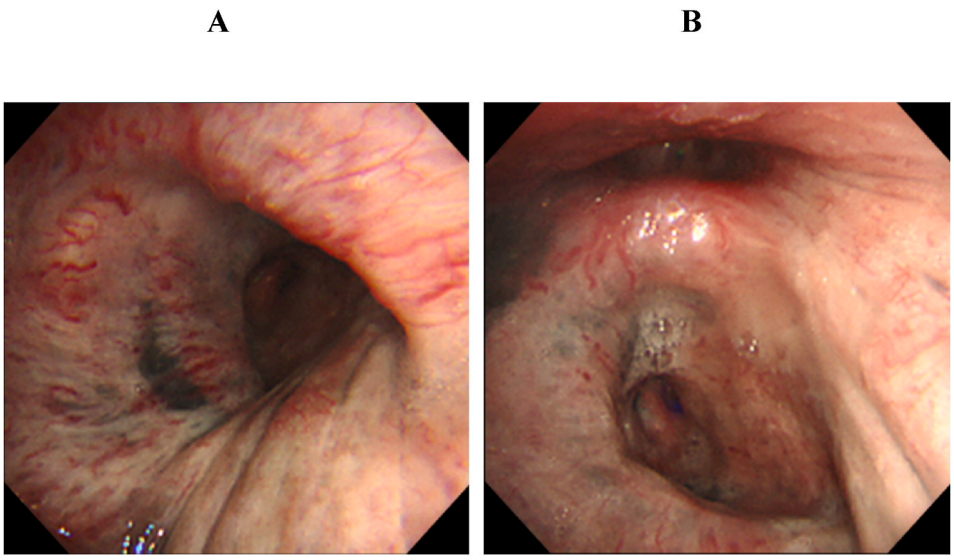

Fig. 4. Bronchoscopic images demonstrating bronchial stenosis with carbon powder deposits (anthracofibrosis), mainly in the right upper lobe. (A) Right main bronchus. (B) Right second carina.

not only pulmonary artery occlusion but also silicosis and bronchial anthracofibrosis. Therefore, this patient was been treated with home oxygen therapy [11] and phosphodiesterase-5 (PDE5) inhibitor [12] for PH due to lung disease, and his symptom and 6MWD remain stable. To our knowledge, this is the first reported case of treatments with home oxygen therapy and phosphodiesterase-5 inhibitor for PH due to silicosis and pulmonary artery occlusion with bronchial anthracofibrosis. Although these treatments carry a risk of worsening gas exchange, they may be considered as a treatment option.

4. Conclusion

Silicosis can be complicated by pulmonary artery occlusion and bronchial anthracofibrosis. Furthermore, patients with bronchial anthracofibrosis should be observed carefully because bronchial anthracofibrosis can be a high-risk factor for PH. Treatments with home oxygen therapy and phosphodiesterase-5 inhibitor may be considered as a treatment option to improve the symptom and 6MWD.

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Disclosure of COI

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