【 CASE REPORT 】

Recurrence of Pulmonary Arteriovenous Malformation with Non-tuberculous Mycobacteria Infection Caused by Perfusion from the Pulmonary Artery and Bronchial Artery after Coil Embolization

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Abstract:
   Recurrence of an embolized pulmonary arteriovenous malformation (PAVM) is common after coil embolization. A 23-year-old woman who had undergone multiple instances of transcatheter coil embolization was admitted with hypoxia and hemoptysis. A PAVM in the left S6 was found to be recanalized by reperfusion through the pulmonary and bronchial arteries. The left S6 was partially resected; the specimen contained necrotic granulomas and non-tuberculous mycobacteria (NTM) around the PAVM. Clinicians should consider possible recurrence of PAVM after reperfusion of the pulmonary and bronchial arteries, as well as the risk of NTM infection during follow-up of patients who have undergone repeated coil embolization.

Key words: pulmonary arteriovenous malformation, coil embolization, reperfusion, bronchial artery, non-tuberculous mycobacteria

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Introduction

   A pulmonary arteriovenous malformation (PAVM) is a structurally abnormal blood vessel that results in capillary-free communication between the pulmonary and systemic circulations and produces an anatomic right-to-left shunt (1). Treatment for PAVM includes surgical resection and transcatheter coil embolization. Coil embolization is the preferred treatment because it is minimally invasive and can be repeated (2). Recurrence of PAVM is a common complication after coil embolization (3). However, recurrence of PAVM after reperfusion of the bronchial artery has not been reported.

   We herein report a case of recurrent PAVM that was caused by reperfusion of the bronchial and pulmonary arteries after coil embolization, accompanied by non-tuberculous mycobacteria (NTM) infection.

Case Report

   A 23-year-old woman presented to a local hospital for the treatment of a mandibular deformity and was found to exhibit hypoxemia. She had been diagnosed with PAVM associated with hereditary hemorrhagic telangiectasia 12 years earlier and had undergone multiple transcatheter coil embolization procedures to treat recurrences of PAVM. She was referred to our hospital for the evaluation of her hypoxemia and treatment of reperfusion of PAVM in the left S6 and left S10. Coil embolization was performed for the PAVM in the
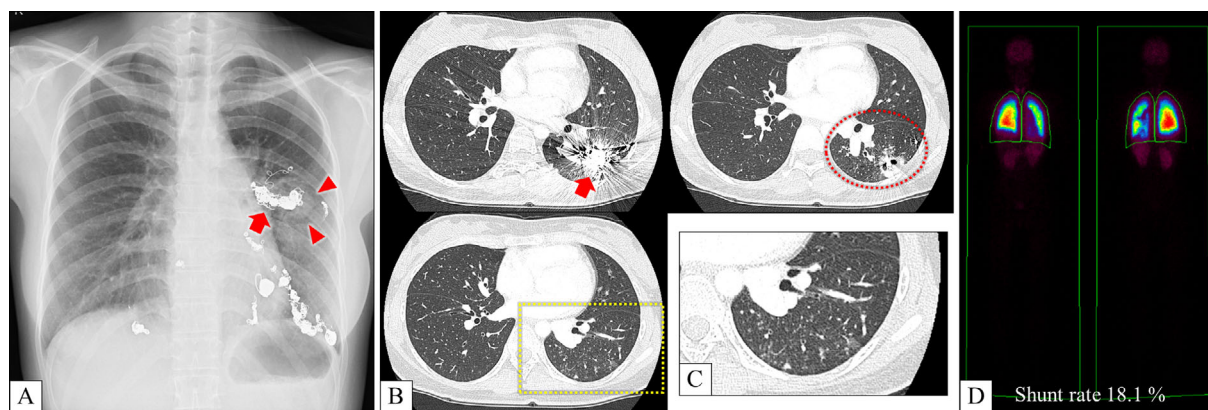
Figure 1. **(A) A chest radiograph showing the numerous coils used for embolization of PAVM in the lung fields bilaterally. In the left lung field, there were coils used for the embolization of the PAVM of the left S6 (red arrow) and sac increased by reperfusion (red triangles). (B) A computed tomography scan of the chest with the lung field setting showing a ground-glass shadow suggestive of bleeding around the PAVM in the left S6 (red dotted circle) and multiple granular shadows scattered throughout the lower left lobe (yellow dotted square, C). (D) Pulmonary blood flow scintigraphy showing the accumulation in organs other than the lungs with a shunt rate of 18.1%. PAVM: pulmonary arteriovenous malformation**

left S10. A month later, she was re-admitted to our hospital with hemoptysis.

The hemoptysis improved spontaneously after admission to hospital. The patient reported that 2 episodes of epistaxis, each lasting approximately 15 minutes, had occurred in the previous weeks. She had a blood pressure of 89/52 mmHg, a pulse rate of 76 beats/min, a peripheral capillary oxygen saturation of 93% in room air, and a body temperature of 36.4℃. Her peripheral capillary oxygen saturation decreased to 88% on exertion. Angiomas were present on her tongue and hands. A chest radiograph showed numerous coils that had been used for embolization of PAVM in the lung fields bilaterally, as well as a PAVM sac in the left S6 (Fig. 1A). Enhanced computed tomography (CT) of the chest and three-dimensional reconstructed images confirmed reperfusion of the PAVM sac in the left S6 where coil embolization had been performed in the past (Fig. 1B). Chest CT with lung field setting revealed a ground-glass shadow, suggestive of bleeding around the PAVM in the left S6, and multiple granular shadows scattered throughout left S6 (Fig. 1C). Blood tests were unremarkable. An arterial blood gas analysis on room air confirmed hypoxia (pH 7.45, PaO_2 68 Torr, $PaCO_2$ 31 Torr). Lung perfusion scintigraphy revealed the accumulation of the tracer in organs other than the lungs and a right-to-left shunt rate of 18.1% (Fig. 1D). Sputum smear examinations for bacteria, mycobacteria, and fungi were negative. Pulmonary arteriography showed reperfusion of the left A6, the PAVM in the left S6, and the draining vein (Fig. 2A). Angiography of the aorta to the bronchial artery also showed blood flow to the PAVM in the left S6 (Fig. 2B). No leak of the contrast agent from the PAVM could be confirmed; thus, an active bleeding source was considered unlikely. Further coil embolization was considered too difficult because of the recurrence involving the

pulmonary and bronchial arteries and coil embolization of the left A6 root. Therefore, the left S6 was partially resected. Lung perfusion scintigraphy revealed that the right-to-left shunt decreased to 12.2% after the surgery. A histopathologic examination of the resected specimen revealed an intermediate form of PAVM between the pulmonary artery and pulmonary vein (Fig. 3C). In addition, several bronchial artery-like arteries were observed near the PAVM, suggesting pathological reperfusion from the bronchial artery to the PAVM. Necrotic granulomas and mycobacteria suggestive of NTM infection were found around the PAVM (Fig. 3A, B, D). Culture of a sputum sample obtained before the operation revealed *Mycobacterium intracellulare* after the operation. Culture of the resected specimen was not performed, and polymerase chain reaction examinations of the resected specimen for *M. tuberculosis* and *M. avium* complex were negative. NTM infection was deemed to be limited to the left S6 because no granular shadows were present in other lung lobes on chest CT. Therefore, drug treatment was not administered for NTM infection; follow-up was performed with imaging examinations. There was no recurrence of hemoptysis or NTM during 12 months of follow-up after surgery.

Discussion

This case report highlights several important teaching points. First, PAVM can recur as a result of reperfusion through the pulmonary and bronchial arteries, and surgical treatment may be suitable in such cases. Second, NTM infection can develop around the PAVM because of a reduction in the blood flow after repeated coil embolization procedures.

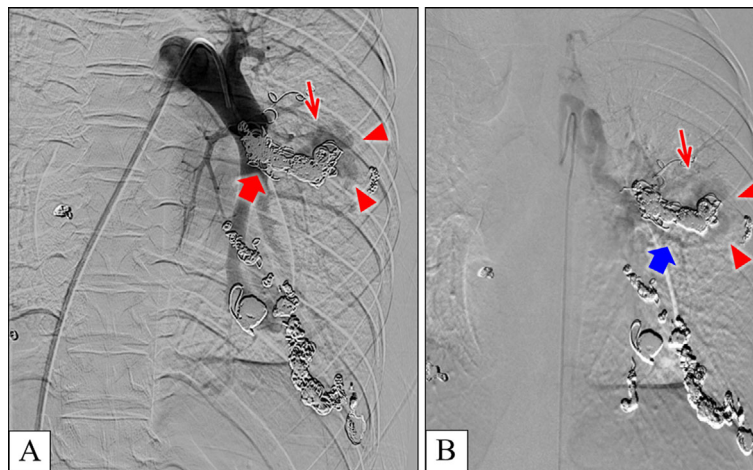Recurrence of PAVM after coil embolization can occur by

Figure 2. (A) A pulmonary arteriogram showing a pulmonary arteriovenous malformation (PAVM) sac in the left S6 (red triangle), a feeding artery from the left A6 (red arrow), and a draining vein (thin red arrow). (B) A bronchial arteriogram showing a feeding artery from the bronchial artery (blue arrow) perfusing the PAVM sac in the left S6 (red triangles) and draining vein (thin red arrow). PAVM: pulmonary arteriovenous malformation
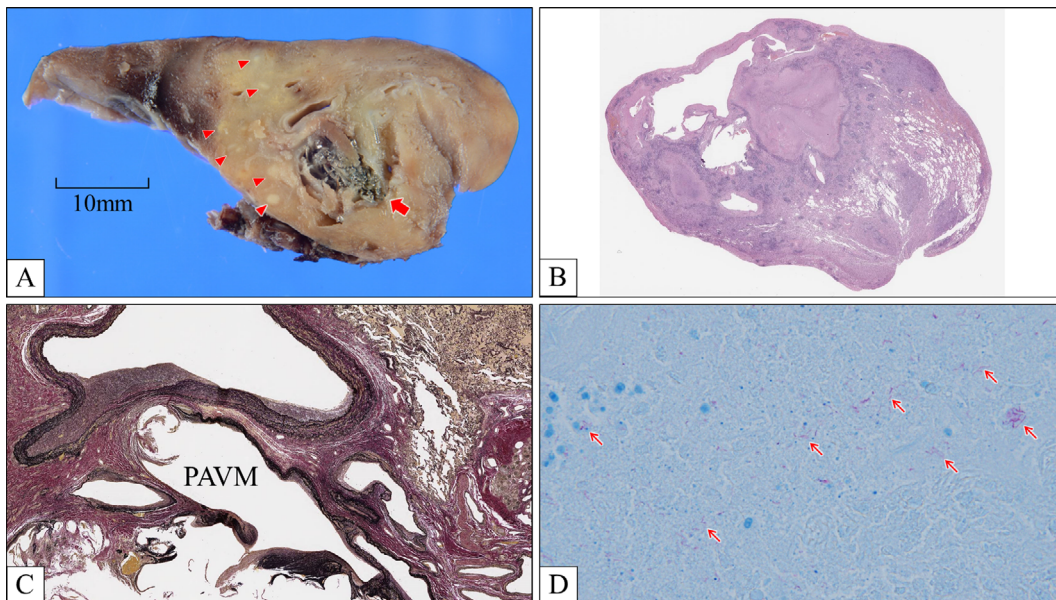

Figure 3. (A) A photograph of the surgically resected specimen showing abnormal vessels recanalizing the PAVM and the coil in the pulmonary artery (red arrow) and small nodules and a white necrotic region (red triangles). (B) Staining of the resected specimen with Hematoxylin and Eosin staining revealed caseous necrosis and mycobacteria around the PAVM. (C) Elastica van Gieson staining of the intermediate structure between the pulmonary artery and pulmonary vein was consistent with a PAVM. (D) Ziehl-Neelsen staining revealed rod-shaped bacteria suggestive of mycobacterial infection (thin red arrows). PAVM: pulmonary arteriovenous malformation

reperfusion via the pulmonary artery as well as the bronchial artery. Follow-up chest CT every few years is recommended to detect reperfusion after coil embolization of a PAVM (4). Although the possibility of reperfusion through the pulmonary artery and bronchial artery has been suggested (3), its frequency is unknown. Furthermore, there is no literature on reperfusion of the bronchial artery extending to a PAVM, which is considered a rare recurrence pattern.

Indeed, although multiple coil embolization procedures had been performed in our patient, reperfusion by the bronchial artery was not considered; a further, evaluation with contrast CT was not performed. In our patient, coil embolization reached the root of the left A6, because multiple coil embolization had previously been performed for the same PAVM of the left S6 during repeated reperfusion of the pulmonary artery. Therefore, the vessels branching from the

bronchial artery increased to compensate for the reduction in blood flow to the left S6, leading to reperfusion of the PAVM in the left S6. Furthermore, a high pressure blood flow from the bronchial artery caused backflow to the pulmonary artery, which led to reperfusion from the left A6. Given that coil embolization had been performed at the root of the left A6 and that the pulmonary and bronchial arteries were reperfused, further coil embolization was considered difficult. Therefore, we opted for surgical treatment, which allowed the recurrence from the pulmonary and bronchial arteries to be confirmed by a pathologic examination and visual inspection intraoperatively. Patients who experience repeated recurrences after multiple coil embolization procedures should be followed up carefully using contrast CT, with consideration of the blood flow in the pulmonary artery as well as in the bronchial artery. Surgical treatment may be necessary in these patients.

A PAVM in which the blood flow is decreased as a result of repeated coil embolization procedures may be accompanied by NTM infection. In a study of patients with chronic pulmonary thromboembolic pulmonary hypertension, NTM infection was reported to be significantly more likely to occur in the area where the blood flow in the lung was decreased as a result of the thrombus (5). In addition, Okuda et al. reported the case of a patient with chronic pulmonary thromboembolic pulmonary hypertension who developed a cavitary lesion caused by *M. intracellulare* in an area of the lung to which the blood flow from the pulmonary artery was decreased (6). Following successful pulmonary endarterectomy, the patient's hemodynamics significantly improved, and the lung cavitation disappeared. A reduction in the blood flow from the pulmonary artery may induce focal immunodeficiency. Kuroda et al. reported that, in cases of chronic pulmonary embolism, NTM infection in locally impaired perfusion of the pulmonary artery due to chronic emboli could be caused by a reduced accessibility of both blood-borne and humoral cell-mediated defensive factors and reduced lung tissue oxygenation due to chronic thrombus (5). A similar localized area of immunodeficiency may occur after repeated coil embolization procedures for PAVM. In our patient, the decrease in the blood flow to the left S6 following coil embolization to the root of the pulmonary artery of the left A6 may have caused the NTM infection. Indeed, the CT findings in this patient did not suggest NTM infection anywhere in the lung other than the left S6. Furthermore, the NTM infection did not recur in this patient despite a lack of pharmacologic intervention. Inflammation as a result of the NTM infection might have caused bleeding from the PAVM in the left S6. In previous reports, compli-

cations after coil embolization in patients with PAVM have included transient pleurisy and paradoxical embolism of devices, thrombi, or air bubbles (1, 2). However, lung infection after coil embolization, as in our patient, has not been reported. A risk of NTM infection because of reduced blood flow in the lungs should be considered in patients who have undergone repeated coil embolization procedures for PAVM. Therefore, careful observation is needed during follow-up to detect blood vessels that may cause reperfusion of PAVM, as well as NTM infection in the lung tissue around the PAVM. Furthermore, if abnormal findings are detected in the lung field around a pulmonary artery that has undergone multiple coil embolization procedures, a sputum examination and bronchoscopy are recommended to diagnose potential NTM infection.

In summary, we reported a case of PAVM that recurred as a result of perfusion from a bronchial artery after coil embolization and was accompanied by NTM infection. During follow-up for PAVM after repeated coil embolization procedures, it is necessary to check for reperfusion from the pulmonary and bronchial arteries and to monitor the lung field around the PAVM for signs of NTM infection.

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