CASE REPORT

Subarachnoid Hemorrhage Caused by Ruptured Intracranial Fusiform Aneurysm Associated With Microscopic Polyangiitis
—Case Report—

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Abstract

A 44-year-old woman with microscopic polyangiitis (MPA) presented with a ruptured cerebral aneurysm. She was admitted to our hospital for further examination of progressive renal failure. She was found lying on the ward floor. Computed tomography showed subarachnoid hemorrhage, and digital subtraction angiography revealed a saccular-like aneurysm arising from the right distal posterior inferior cerebellar artery (PICA) at the non-branching site. We performed neck clipping of the right distal PICA aneurysm, which recurred 5 days after the operation. Second angiography demonstrated a right distal PICA aneurysm just above the site of the clip. Therefore, we performed trapping of the affected lesion with emergent bypass of the contralateral occipital artery to the ipsilateral PICA. Her postoperative course was uneventful until she coughed up blood and had gross hematuria 3 days after the second surgery. Histological examination of a renal biopsy specimen revealed crescentic glomerulonephritis. MPA was diagnosed on the basis of the cardinal symptoms, including progressive glomerular nephritis and the lung abnormality, as well as the presence of myeloperoxidase-antineutrophil cytoplasmic antibodies. After intensive treatment, she was discharged for rehabilitation without neurological deficit. MPA commonly affects small-sized vessels mainly in the kidneys and lungs and may lead to crescentic glomerulonephritis and pulmonary hemorrhage. MPA is rarely associated with aneurysms of medium-sized muscular vessels. Cerebral aneurysm is extremely rare in patients with MPA, but rupture of an intracranial fusiform aneurysm can be lethal, so screening of the intracranial vessels should be performed by magnetic resonance imaging in patients with MPA.

Key words: microscopic polyangiitis, cerebral aneurysm, fusiform aneurysm

Introduction

Primary vasculitis has been classified based on the size of the affected vessels (Chapel Hill Consensus Conference, 1994).6) Large-vessel vasculitis includes Takayasu’s arteritis and temporal arteritis; medium-sized vessel vasculitis includes polyarteritis nodosa (PN) and Kawasaki’s disease; and small-vessel vasculitis includes microscopic polyangiitis (MPA), Wegener’s granulomatosis, and allergic granulomatous angiitis.6) Vasculitis in small vessels is known as antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis because of the involvement of ANCA as a common pathogenetic factor.5) Histological examination of the involved organs has revealed necrotizing crescentic glomerulonephritis, and necrotizing vasculitis of the arterioles, capillaries, and venules with few immune deposits because ANCA appears to induce vasculitis by directly activating neutrophils. Therefore, ANCA-associated vasculitis is called pauci-immune vasculitis (pauci = few/little).

The major target antigens of ANCA-associated vasculitis are myeloperoxidase and proteinase 3.2,14) Myeloperoxidase-ANCA is related to MPA and allergic agranulomatous angiitis, and proteinase 3-ANCA is the marker antibody in Wegener’s granulomatosis.11) According to a broadly accepted recent classification, ANCA-associated vasculitis is characterized by the involvement of small vessels in various organs such as the lungs, kidneys, skin, and peripheral nervous system.1,4,7) MPA is the most common ANCA-associated small-vessel vasculitis, which mainly affects the kidneys and lungs.3,6) The kidneys are the most commonly affected organ, accounting for about 90% of patients; and kidney vasculitis can result in renal failure.13) Interstitial pneumonitis and pulmonary hemorrhage are also common.5) The diagnostic criteria for MPA have already been published by the Research Group of Intractable Vasculitis, Ministry of Health, Labour and Welfare of Japan.10) MPA is rarely associated with aneurysms of
medium-sized muscular arteries that are frequently encountered in PN.\textsuperscript{13} Only a few cases of MPA associated with aneurysms have been described in various organs except the brain.\textsuperscript{8,13} No previous cases of ANCA-associated vasculitis associated with cerebral aneurysm have been reported.

Here, we report the first documented case of MPA that developed subarachnoid hemorrhage (SAH) due to rupture of the intracranial aneurysm.

Case Report

A 44-year-old woman with a history of hypertension for 8 years and gross hematuria for the last 3 years had a high serum titer of myeloperoxidase-ANCA of 312 enzyme-linked immunosorbent assay (ELISA) units (normal < 20 ELISA units). She had undergone percutaneous renal biopsy 2 years earlier, which showed only minor glomerular abnormalities. Thereafter, her renal function deteriorated gradually and she was readmitted to our hospital to perform a second renal biopsy. She was found lying on the ward floor 2 days after the renal biopsy.

Computed tomography (CT) showed SAH mainly located in the cisterna magna and mild hydrocephalus (Fig. 1A). Her consciousness fully recovered after the scan, and she began to complain of headache. Digital subtraction angiography (DSA) revealed a saccular-like aneurysm arising from the right distal posterior inferior cerebellar artery (PICA) and a fusiform aneurysm of the left vertebral artery (Fig. 1B, C). Based on the location of the subarachnoid hematoma, rupture of the right distal PICA aneurysm was diagnosed, and emergent neck clipping was performed soon after the examination (Fig. 2A). Although premature rupture occurred in dissecting the aneurysm neck, bleeding was well controlled by applying a temporary clip, the aneurysm neck was secured, and neck clipping was accomplished in the usual manner followed by ventricular drainage because of tight posterior fossa.

Postoperatively, her consciousness level was rated as 14 on the Glasgow Coma Scale (E3V5M6), without paroxysm of the extremities. Five days after the operation, she experienced severe headache and rapid drainage of a great deal of ventricular hematoma from the ventricular catheter. CT revealed that the subarachnoid hematoma had increased slightly, and a new ventricular hematoma. DSA demonstrated recurrence of the right distal PICA aneurysm just above the site where the clip had been applied (Fig. 3). We reviewed the first operating video record and noticed a residual fusiform aneurysm (Fig. 2B). Therefore, we diagnosed fusiform aneurysm and decided to perform emergent trapping of the affected lesion.

During the operation, we found that the right occipital artery (OA) had already been occluded. Thus, the left OA was harvested, and left OA to right PICA anastomosis was performed before approaching the ruptured aneurysm. Although premature rupture occurred in dissecting the aneurysm, bleeding was easily controlled by placing a permanent clip just proximal to the lesion. Aneurysm trapping was completed by applying another clip on the parent artery just distal to the aneurysm (Fig. 2C), followed by opening of the anastomosis. Both the initial and second aneurysms were excised. Unfortunately, the distance between the two clips was too short to excise the afferent parent artery, so that no pathological diagnosis could not be obtained. We were only able to excise the two aneurysm walls.

The first aneurysm seemed to already have undergone fibrinoid degeneration, and histological examination showed the very fragile tissue was old hematoma. The second aneurysm wall consisted of a paper-like, whitish thin wall, and histological examination demonstrated a fibrin layer (Fig. 4). Therefore, the diagnosis was pseudoaneurysm. Considering the angiographical and intraoperative findings, our final diagnosis was fusiform aneurysm that had ruptured and formed pseudoaneurysm. Three days after the operation, the patient experienced severe respiratory distress and coughed up blood and had gross hematuria coincidently. Histological examination of the renal specimens, taken before the onset of the first SAH, revealed crescentic glomerulonephritis. Therefore, we diagnosed MPA on the basis of the cardinal symptoms of rapidly progressive glomerular nephritis, the lung abnormality, and a positive test for myeloperoxidase-ANCA. After initiation of steroid pulse therapy, cyclophosphamide pulse therapy, and hemodialysis, her renal and pulmonary func-
Fig. 2 Intraoperative photomicrographs at the first (A, B) and the second (C) surgeries. A: The “saccular” aneurysm (arrowheads) arising from the right posterior inferior cerebellar artery, parent artery (asterisk), was clipped with a Sugita aneurysm clip. B: Fusiform aneurysm (arrowheads) of the parent artery (asterisk) can be seen postoperatively. C: Both the fusiform aneurysm (asterisk) and a new pseudoaneurysm (arrowheads) were trapped to achieve hemostasis to prevent intraoperative premature rupture. The reddish-white pseudoaneurysm wall (arrowheads) is clearly visible. The arrow shows the aneurysm clip that was applied at the first surgery and the other two clips in the figure at the second surgery.

Fig. 3 A, B: Right vertebral artery angiograms just after the second subarachnoid hemorrhage, anteroposterior (A) and lateral views (B), demonstrating recurrence of the distal posterior inferior cerebellar artery (PICA) aneurysm at the same position as the initial aneurysm projecting superiorly. High magnification views of the aneurysms (insets) showing the fusiform aneurysm-like dilatation (arrows) and pseudoaneurysm (arrowheads). C: Left occipital artery angiogram after the second surgery showing patency of the distal PICA to the affected lesion. Arrowhead shows the site of the anastomosis. D: Right vertebral artery angiogram demonstrating complete disappearance of the aneurysm.

Fig. 4 Photomicrograph of the resected pseudoaneurysm showing the entire resected aneurysm wall. Hematoxylin and eosin stain, original magnification ×4. Inset: High magnification of the aneurysm wall showing only a fibrin layer without inflammatory cell infiltration or normal vessel structures. Hematoxylin and eosin stain, original magnification ×20.

Discussion

Our present patient was positive for myeloperoxidase-ANCA. Her renal function rapidly deteriorated after the second operation, and she suffered hemoptysis coincidentally that might have been caused by pulmonary hemorrhage. Histological examination revealed pauci-immune type crescentic glomerulonephritis based on the renal biopsy. These clinical and pathological findings are consistent with MPA, and she was diagnosed with MPA on the basis of the established diagnostic criteria.10

In general, patients with MPA commonly suffer from arteritis of the small-sized vessels. Arteritis of medium-sized vessels may also sometimes occur in MPA patients. About 23% of MPA patients had medium-sized renal artery involvement.15 However, aneurysm formation in medium-sized vessels is a rare feature of MPA3 but is frequently encountered in PN.12 Only a few cases of MPA have been associated with aneurysms of systemic, medium-sized muscular arteries. However, there have been no previous reports of MPA cases with a cerebral artery aneurysm. The present case of MPA with a cerebral aneurysm was detected because of rupture of the aneurysm.

MPA associated with aneurysms is very rare, so little is known about how aneurysm formation occurs in the affected vessels and what types of aneurysms may be formed. The exact pathogenesis and mechanisms are unclear. However, necrotizing changes of the affected vessels may occur followed by weakening of the vessel wall and aneurysm formation.8 The presence of coronary ectasia was detected in 14 of 16 patients with MPA, and
fusiform coronary aneurysms were identified in 4 patients using magnetic resonance angiography. Tissue inflammation may be involved to some extent. In our case, the pathological findings in the wall of the second aneurysm showed just a fibrin membrane, indicating a pseudoaneurysm. The aneurysm, which consisted of very fragile tissue, had already degenerated by the time of the second surgery, and had become old hematoma. We think that the first aneurysm may also have been a pseudoaneurysm. When she experienced recurrence of the aneurysm, we reviewed the initial intraoperative findings and noticed fusiform-like arterial ectasia at the affected parent artery. Furthermore, we retrospectively observed the same finding in both angiograms. A fusiform aneurysm of the right vertebral artery may have already existed at the time of the first surgery, and was present on the second DSA. We assume that she had ANCA-associated vasculitis at the distal PICA, which is a medium-sized muscular vessel. A fusiform aneurysm might have formed and ruptured at the most fragile point of the wall, resulting in the formation of a pseudoaneurysm. At the first operation, we did not remove the fusiform aneurysm, so that rupture of the aneurysm occurred again, leading to the second pseudoaneurysm.

The present case of MPA associated with cerebral fusiform aneurysm was detected because of aneurysm rupture. Cerebral aneurysm is extremely rare in patients with MPA, but rupture of an intracranial fusiform aneurysm can be lethal, so screening of the intracranial vessels should be performed by magnetic resonance imaging in patients with MPA.

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References


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