Treatment of an Unruptured Fusiform Aneurysm of the Internal Carotid Artery Associated With Wegener’s Granulomatosis by Endovascular Balloon Occlusion

—Case Report—

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Abstract

A 22-year-old woman developed an unruptured fusiform aneurysm of the internal carotid artery 7 months after being diagnosed with Wegener’s granulomatosis. Intracranial aneurysmal formation is an extremely rare complication of Wegener’s granulomatosis. This rare case of intracranial aneurysm was treated by endovascular balloon occlusion.

Key words: Wegener’s granulomatosis, intracranial fusiform aneurysm, carotid artery dilation, endovascular balloon occlusion

Introduction

Wegener’s granulomatosis is a type of systemic necrotizing, granulomatous vasculitis of the small and medium arteries, mainly in the respiratory tract, kidneys, and other organs. Wegener’s granulomatosis is strongly associated with the expression of antineutrophil cytoplasm antibodies (ANCA), specifically proteinase 3 (PR3)-ANCA. Neurological manifestations of Wegener’s granulomatosis are usually peripheral nerve disease, reported in 22–54% of patients.10 Central nervous system involvement is reported in 7–11% of patients.3,5,8,10 Previous reports have documented cerebral vasculitis manifesting as chronic hypertrophic pachymeningitis. Intracranial aneurysms in Wegener’s granulomatosis are extremely rare, and only one case of a clipping operation of a ruptured aneurysm has been reported.11 We treated a case of unruptured, Wegener’s granulomatosis-induced intracranial aneurysm by proximal endovascular occlusion, and describe the 4-year postoperative follow-up findings.

Case Report

A 22-year-old woman presented with a history of biopsy for intranasal granuloma. Immunochemical examination included PR3-ANCA under a diagnosis of Wegener’s granulomatosis in 2006. She was admitted for steroid pulse and intravenous cyclophosphamide therapy (7 treatments for a total dose of 5.15 g), but her paranasal sinusitis worsened. The disease responded to immunosuppressive therapy with rituximab. She received follow-up immunochemical examinations and paranasal magnetic resonance (MR) imaging studies every 2 months, with no apparent abnormalities. However, 7 months after the diagnosis of Wegener’s granulomatosis, paranasal MR imaging incidentally detected dilation of the left internal carotid artery (ICA) (Fig. 1A–C). Cerebral angiography detected an unruptured fusiform aneurysm (7-mm diameter) of the ICA and proximal part of the middle cerebral artery with a bleb on the bifurcation of the anterior choroidal ar-
Fig. 1 A–C: Preoperative T2-weighted magnetic resonance images, initial (A), after 3 months (B), and 7 months (C), showing an aneurysm of the left internal carotid artery. D–F: Postoperative T2-weighted magnetic resonance images, after 1 month (D), 1 year (E), and 4 years (F), showing the size of the aneurysm unchanged in the left internal carotid artery.

Fig. 2 Digital subtraction angiograms (DSA) of the left internal carotid artery, anteroposterior view (A) and lateral view (B), and three-dimensional DSA (C), showing an unruptured fusiform aneurysm with a small aneurysm on the bifurcation of the anterior choroidal artery.

Fig. 3 A, B: Oblique craniogram (A) and computed tomography scan of the skull (B) showing two detachable balloons (arrows) located at the intra-pyramidal portion of the internal carotid artery (ICA) placed for permanent proximal occlusion of the ICA. C, D: Digital subtraction angiograms performed after occlusion showing collateral flow in the middle cerebral artery (MCA) from the anterior communicating artery (C), and collateral flow of MCA and dilated fusiform lesion of the ICA from the posterior communicating artery (D). E: Non-subtracted left vertebral angiogram showing locations of the detachable balloons (arrows) and the fusiform lesion filled with contrast medium from the collateral circulation of the posterior communicating artery.

In our case, the ICA aneurysm appeared fusiform and complex, with a wide neck. Surgical flow alteration has been reported as an effective therapy for such cases, with excellent control rates and no new aneurysms in long-term follow up. Early ischemic attacks are uncommon complications of proximal occlusion. Delayed ischemic complications after direct surgical ligation of ICA occur at a rate of 7.5–20%, but at a rate of 3% with endovascular ICA occlusion. In our patient, endovascular ICA occlusion yielded an uneventful course without complications over several years. Monitoring of this patient will be continued to detect the appearance of new aneurysms or other complications.

This rare case of unruptured fusiform aneurysm associated with Wegener’s granulomatosis was treated by endovascular ICA occlusion and remained stable for 4 years. Evaluation of the true effectiveness of this treatment must wait for long-term follow up.

Discussion

Aneurysm formation in visceral arteries has been reported in Wegener’s granulomatosis, but intracranial aneurysms are extremely rare, with only one case of a tiny ruptured aneurysm at the junction of the left ICA and anterior choroidal artery treated by neck clipping. The present case of an unruptured fusiform aneurysm in Wegener’s granulomatosis was treated by endovascular balloon occlusion.

References

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