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【 CASE REPORT 】

Giant Cell Arteritis with Internal Carotid Artery Occlusion in the Absence of Typical Clinical Features

Mai Kikumoto[1], Shiro Aoki[1], Yuji Shiga[1], Naoto Kinoshita[1,2], Tomohisa Nezu[1], Hiroki Ueno[1], Koji Arihiro[3] and Hirofumi Maruyama[1]

Abstract:
A 65-year-old man presented with a slight headache and transient visual disturbance. Magnetic resonance imaging (MRI) revealed occlusion of the left internal carotid artery (ICA) and acute brain infarctions in both hemispheres, and a blood examination indicated inflammation. Gadolinium enhancement was observed in the walls of the temporal arteries and ICAs. After we diagnosed giant cell arteritis (GCA) by a temporal artery biopsy, aspirin and corticosteroids were administered. The typical symptoms of GCA, such as jaw claudication and temporal artery tenderness, were absent during the entire clinical course, and the findings of contrast-enhanced MRI contributed to the diagnosis.

Key words: giant cell arteritis, internal carotid artery occlusion, contrast-enhanced MRI, stroke

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Introduction

Giant cell arteritis (GCA) is characterized by the presence of granulation tissue on the arterial wall (1, 2). The typical symptoms are jaw claudication, temporal artery tenderness, and muscle pain, but some cases lack these clinical features. GCA is also known as a causative vasculitis of brain infarction, and 3-4% of patients with GCA experience a cerebrovascular ischemic event (3). Because GCA can have serious complications, including recurrent stroke and occlusion of the ophthalmic artery, treatment should be started immediately (2). When the typical clinical features of GCA are not observed, however, it is difficult to make the decision to proceed with a biopsy for the diagnosis.

We herein report a biopsy-proven case of GCA with left internal carotid artery (ICA) occlusion. The patient exhibited only slight neurologic deficits, but the findings of contrast-enhanced magnetic resonance imaging (MRI) enabled us to make the decision to perform a biopsy of the temporal artery. Contrast-enhanced MRI was helpful for detecting an undetermined cause of stroke.

Case Report

A 65-year-old man presented with a slight headache and an itchy forehead. He had experienced transient visual disturbance of the left eye that occurred every 2 to 3 days with a duration of about 30 seconds for 2 months after the initial headache. In addition, he suffered from recurrent transient paresthesia in the extremities that lasted for 20 to 60 seconds. He was a habitual smoker and had hyperlipidemia without any other risk factors of cerebrovascular events. He was not taking medication.

MRI performed at a local hospital showed slight stenosis in the left ICA distal to the dural ring (Fig. 1). An ophthalmic examination revealed no deficit in his visual ability.

Four months after the initial headache, he suddenly developed transient motor weakness in the right extremities, followed by aphasia. At admission to our hospital, his body weight was 57.2 kg, and his BMI was 22.3 kg/m^2, findings that were within the normal range for his age; however, he had lost 7 kg in the 3 months prior to admission. His body temperature was slightly elevated (37.3°C). The frontal headache had weakened but was still present. He had no jaw

[1]Department of Clinical Neuroscience and Therapeutics, Hiroshima University, Graduate School of Biomedical and Health Sciences, Japan, [2]Department of Neurology, Hiroshima Prefectural Hospital, Japan and [3]Department of Anatomical Pathology, Hiroshima University Hospital, Japan
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Correspondence to Dr. Shiro Aoki, shirou@hiroshima-u.ac.jp

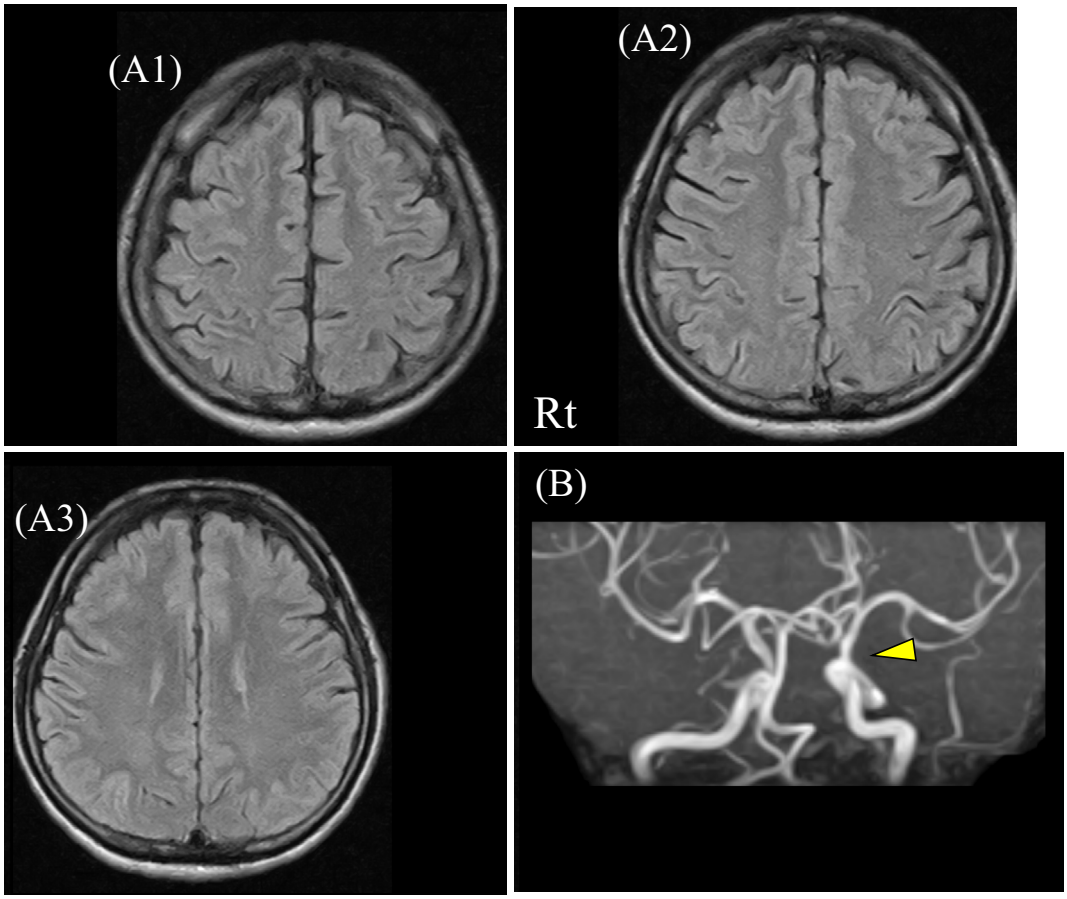

Figure 1. **MRI of the brain (axial FLAIR and MRA) performed two months after the initial headache. (A1-3) No hyperintensity is seen on FLAIR images. (B) MRA depicts slight stenosis in the left intradural ICA distal to the dural ring (arrowhead). MRI: magnetic resonance imaging, FLAIR: fluid-attenuated inversion recovery, MRA: magnetic resonance angiography, ICA: internal carotid artery**

claudication, muscle pain, or temporal artery tenderness.

On admission, he complained of slight aphasia and dysarthria. The visual disturbance partially remained in the left eye, but his symptoms were atypical of GCA with a sensation similar to photophobia. Brain MRI revealed bilateral watershed infarctions between the anterior, middle, and posterior cerebral artery territories that were distributed dominantly in the left hemisphere. The left ICA was occluded on magnetic resonance angiography (MRA). Stenosis was observed at the siphon of the right ICA (Fig. 2).

Laboratory tests revealed an elevated erythrocyte sedimentation rate (ESR; 105 mm for the first hour). Ferritin and C-reactive protein (CRP) levels were also elevated (365.1 ng/mL and 8.12 mg/dL, respectively), indicating inflammation. No specific autoantibodies for angiitis, such as anti-neutrophil cytoplasmic antibody, were detected.

Based on these findings, we made a provisional diagnosis of large-artery atherosclerosis and started dual antiplatelet therapy with aspirin and clopidogrel. As the ferritin, CRP, and ESR values continued to increase after starting treatment, and the episodes of headache and weight loss indicated vasculitis, we decided to conduct further examinations.

Contrast-enhanced MRI revealed vessel wall enhancement (VWE) in the bilateral temporal arteries, intradural ICAs, and vertebral arteries, predominantly on the left. The vessel walls at the occlusion of the intradural left ICA and the stenosis at the right siphon were also enhanced (Fig. 3). As these findings were indicative of GCA, a biopsy of the left superficial temporal artery was performed. Histopathology of the vessel wall revealed multinucleated giant cells in the tunica media (Fig. 4).

The diagnosis of GCA was confirmed by the result of the biopsy. The brain infarctions in the left hemisphere were considered to have been caused by hemodynamic changes due to the occlusion of the left ICA. The microvessels distal to the siphon in the right hemisphere were likely to have been occluded by vasculitis. We started oral prednisolone at 50 mg/day due to his mild symptoms; he did not need steroid pulse therapy to alleviate his neurologic deficits. Clopidogrel was terminated at the same time, and aspirin was continued.

After administering prednisolone, laboratory data showed a decrease in the ESR, CRP, and ferritin values. Serial follow-up MRI performed after starting prednisolone showed no recurrence of brain infarction or progressive stenosis in other arteries. The VWE of the temporal arteries gradually

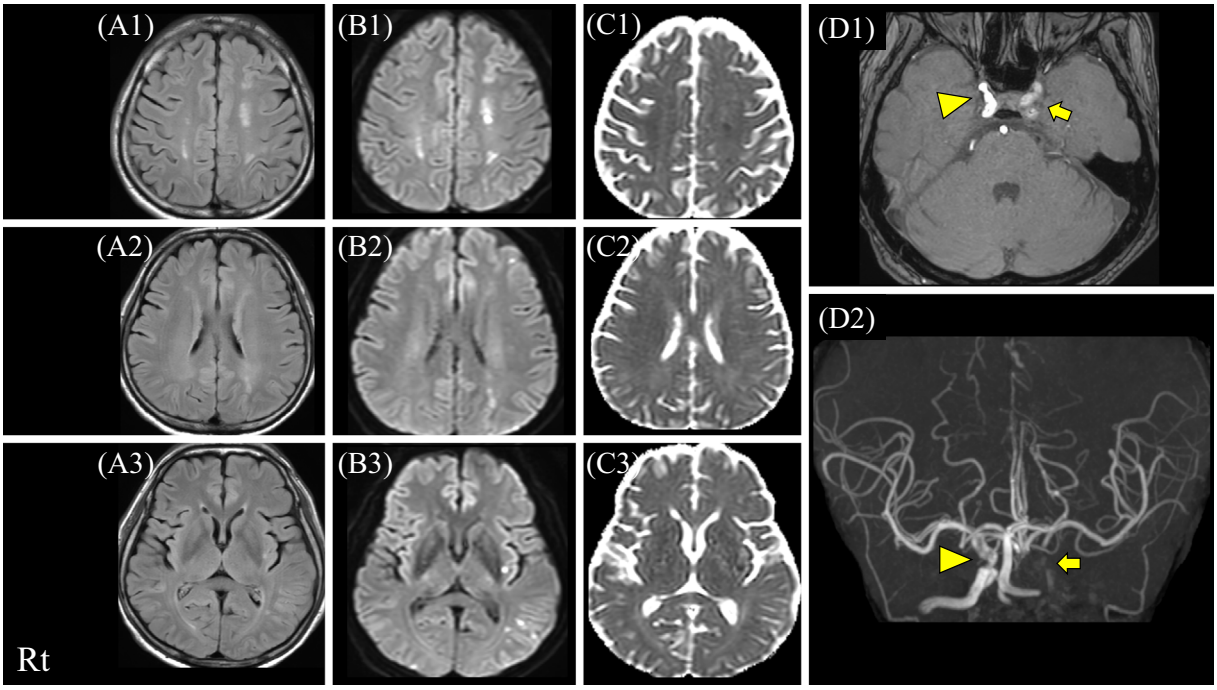

Figure 2. **MRI of the brain (axial DWI, ADC, and FLAIR), and MRA performed after admission. (A1-3, B1-3) FLAIR and DWI show hyperintense lesions in the bilateral watershed areas between the anterior, middle, and posterior cerebral artery territories, which are predominantly distributed in the left hemisphere. (C1-3) The ADC values of the lesion are slightly lower in the left hemisphere than in the right. (D1) MRA depicts occlusion in the left ICA (small arrow) and slight stenosis at the siphon of the right ICA (arrowhead). (D2) The left ICA is invisible on reconstruction of MRA images (small arrow). The stenosis of the right ICA is also observed at the siphon (arrowhead). DWI: diffusion-weighted imaging, ADC: apparent diffusion coefficient**

weakened. Oral prednisolone treatment was gradually tapered to 30 mg/day at 6 weeks after stroke onset without any relapse of neurological deficits. Furthermore, MRI performed 7 months after the stroke onset showed that the occlusion of the left ICA remained, without acute brain infarction, and the dose of prednisolone was tapered to 9 mg/day at 1 year after starting on steroids.

Discussion

We diagnosed GCA as a cause of bilateral brain infarction by a temporal artery biopsy in the present case, although typical clinical features of GCA were not observed. Contrast-enhanced MRI enabled us to make a decision to perform a biopsy by showing enhancement in the temporal arteries. We were able to start steroid therapy at the early stage by obtaining a diagnosis of GCA. VWE in the temporal artery on contrast-enhanced MRI is an important clue suggesting a diagnosis of GCA as a cause of stroke with undetermined etiology.

GCA is a type of large-vessel vasculitis that is usually accompanied by giant cells in the temporal artery (1, 2). The typical clinical features of GCA are jaw claudication, muscle pain, and temporal artery tenderness, which reflects inflammation. Although GCA is also known as a causative disease of cerebrovascular events, brain infarctions are uncommon among patients with GCA. A previous study reported that only 3-4% of patients with GCA present with a cerebrovascular ischemic event (3). Brain infarctions caused by GCA are more likely to occur in the vertebrobasilar than ICA territory (4). The low incidence of ischemic events in the ICA territory may correlate with the low frequency of severe arteritis in the ICA (5). A noteworthy feature of the present case is that the left intradural ICA became invisible on MRA within one and a half months and that the patient displayed a relatively slight neurological deficit despite an acute process.

High-resolution MRI at 3 Tesla using gadolinium-based intravenous contrast agents can be used to localize the involved cranial arteries by revealing mural inflammatory changes as increased mural contrast enhancement, and the diagnostic benefit of an evaluation by MRI has been also demonstrated (6-8). The diagnostic value of the enhancement in the temporal and occipital arteries in particular has been described in a previous report (6). The present patient showed atypical clinical signs of GCA, and VWE in the superficial temporal artery was helpful for deciding whether or not to perform a temporal artery biopsy. Of note, the wall of the intradural ICA was also enhanced as mentioned in a previous study (9). VWE is not typically associated with corresponding stenosis or occlusion (9), so a further examination, such as ultrasonography or MRA, is needed to evaluate the

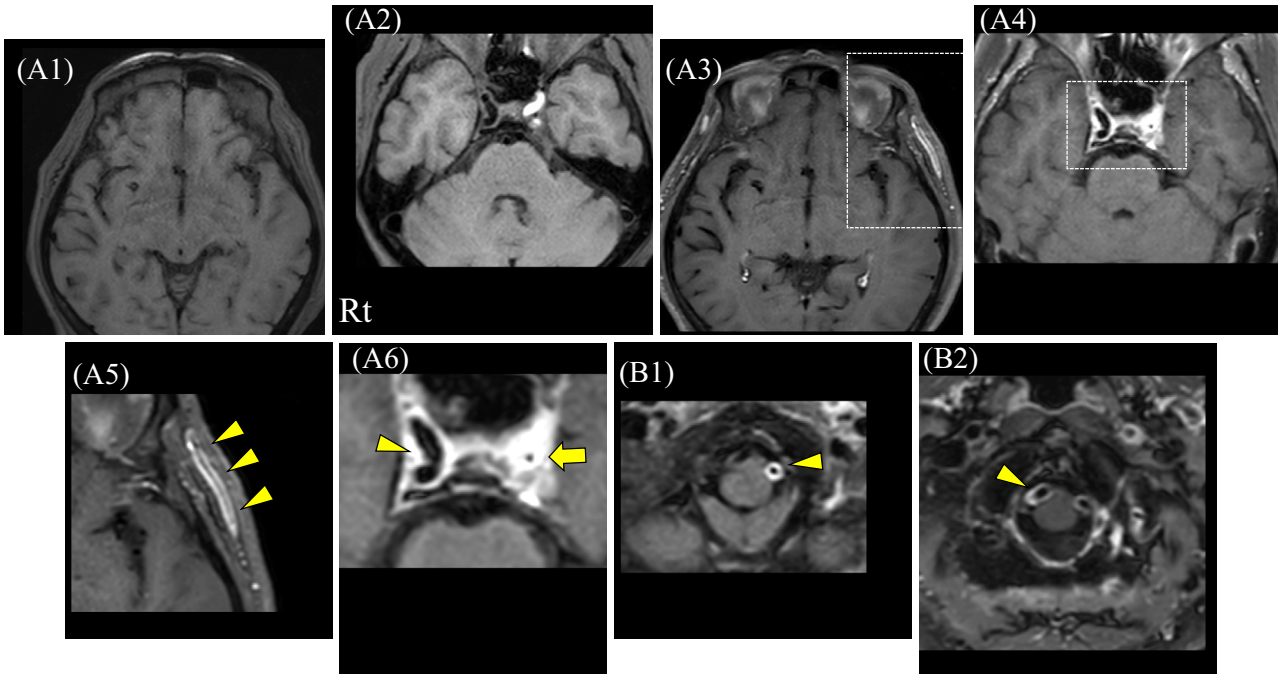

Figure 3. **Plain and contrast-enhanced MRI of the brain (axial CE-MSDE-3D TSE) performed after admission. (A1, 2) MDSE-3D TSE depicts the walls of the left temporal artery and ICAs with isointensity signals. (A3, 4) Vessel wall enhancement (VWE) is observed in the walls of the left temporal artery and ICAs on CE-MSDE-3D TSE. (A5, 6) Magnified images of the areas enclosed by dotted lines in A3, 4 show the enhanced artery walls of the left temporal artery (arrowheads) and ICAs (right: arrowhead, left: arrow). (B1, 2) The vertebral arteries are also bilaterally enhanced (arrowheads). MSDE-3D TSE: motion-sensitized driven equilibrium three-dimensional turbo spin echo sequence, CE-MSDE-3D TSE: contrast-enhanced three-dimensional turbo spin echo sequence**

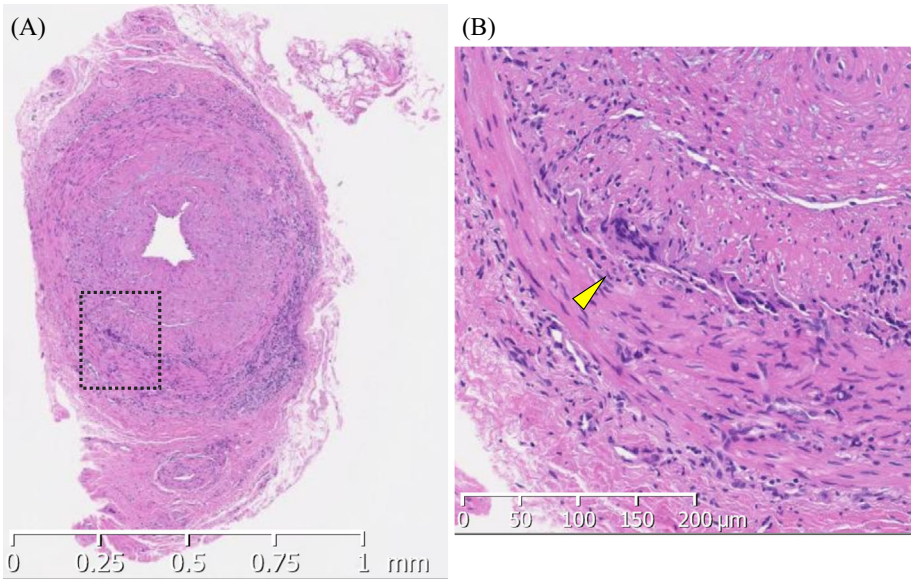

Figure 4. **Histopathology of the left superficial temporal artery. Left (A), ×2.5; right (B), ×10. (A) Diffuse filtration of inflammatory cells is observed mainly in the tunica media and adventitia. The intima and tunica media are thickened due to chronic inflammation, and fibrosis is seen. There is no obvious granuloma in the tissue. (B) Multinucleated giant cells are located in the tunica media (arrowhead) (Hematoxylin and Eosin staining).**

involved vessels.

GCA can result in severe sequelae, such as irreversible visual disturbance. Although steroid therapy should be started immediately, it is challenging to diagnose patients

without typical features as GCA at the early stage. VWE is observed in the temporal artery on contrast-enhanced MRI in many cases with GCA, and it can be useful for deciding whether or not to perform a biopsy. As a brain infarction with an undetermined cause can be related to vasculitis, including GCA, contrast-enhanced MRI should be considered in order to make a diagnosis. We can initiate appropriate treatment promptly by obtaining a GCA diagnosis at an early stage.

The authors state that they have no Conflict of Interest (COI).

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