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Case Report

Giant cell arteritis with severe intracranial involvement diagnosed and treated early

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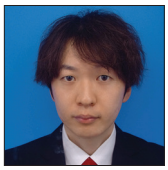

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ABSTRACT

Background: Ischemic cerebrovascular accidents (CVA) occur in 3.3–7.2% of patients with giant cell arteritis (GCA), and intracranial vessels are rarely affected. We, herein, report a case of intracranial GCA with rapidly progressive multiple intracranial vascular lesions.

Case Description: A 76-year-old woman visited a local doctor due to a headache; then, it improved spontaneously. Three months later, she suddenly had cerebral infarctions of bilateral pons and cerebellum. Magnetic resonance angiography (MRA) revealed the left internal carotid artery (ICA) occlusion, the right vertebral artery (VA) occlusion, and the left VA stenosis. She was diagnosed with atherothrombotic stroke and dual antiplatelet therapy was administered. However, 2 weeks later, the left VA stenosis was aggravated. Therefore, we reviewed the data of MRA performed 3 months ago and noted no lesions in the ICA and VA. T1 black-blood post-gadolinium imaging sequence magnetic resonance imaging (MRI) revealed vessel wall enhancement in the bilateral VA, left ICA, and bilateral superficial temporal artery. We performed a temporal artery biopsy and diagnosed her with GCA. The progression of the intracranial vascular lesions was decelerated by oral glucocorticoid administration.

Conclusion: Intracranial vascular lesions in GCA can be formed later than initial symptoms, such as headache, and aggravated despite improvement in headache. In patients with GCA, evaluating intracranial vessels as a control is useful for distinguishing them from arteriosclerotic lesions at the onset of CVA. Intracranial GCA is characterized by rapidly progressive vascular lesions in the bilateral ICA and VA. In addition, T1 black-blood post-gadolinium imaging sequence MRI may lead to early diagnosis and treatment.

Keywords: Giant cell arteritis, Ischemic cerebrovascular accidents, Multiple intracranial vascular lesions, Rapidly progressive lesions

INTRODUCTION

Giant cell arteritis (GCA) is a primary large-vessel vasculitis found commonly in older people aged 50 or more years and mainly affects the extracranial branches of the carotid artery.[4,6-8,18] Initial symptoms of GCA mainly include headache, tenderness of the temporal arteries, jaw claudication, and fatigue.[13,18] Concomitant ischemic cerebrovascular accidents (CVA) have been reported in 3.3–7.2% of patients with GCA.[3,5] Cerebral infarctions in patients with GCA are mostly caused by poor blood circulation due to severe stenosis or occlusion of extracranial vessels.[12,16] GCA with lesions affecting intracranial vessels (intracranial GCA) is rare, but patients with intracranial GCA have poor prognoses because the disease is refractory to treatment and

has a high mortality rate.[12,13] Moreover, in most cases, cerebrovascular lesions are not evaluated at the onset of GCA symptoms, such as headaches.[1,7,9,10,12-15,19] We, herein, report a case of intracranial GCA diagnosed and treated relatively early with good outcomes, where imaging evaluation of cerebrovascular lesions could be performed continuously throughout the various stages of intracranial vascular lesions, including the onset of GCA, onset of CVA, the advanced stage, and the stable stage.

CASE DESCRIPTION

A 76-year-old woman visited a local doctor immediately after she experienced headaches in the left and right temporal regions of the head. Magnetic resonance imaging (MRI) results showed no remarkable abnormal findings, and the doctor took a wait-and-see approach. The headache resolved spontaneously and did not relapse afterward. The patient had a history of hypertension and was undergoing treatment with oral antihypertensives.

Three months later, the patient visited our hospital because she suddenly experienced slurred speech and lightheadedness. Physical findings included disorientation, dysarthria, ataxic symptoms in the right upper and lower limbs, and inability to walk. MRI revealed infarction lesions in the left and right sides of the pons and cerebellum [Figure 1a], while the left internal carotid artery (ICA) and the right vertebral artery (VA) were not well visualized in magnetic resonance angiography (MRA) [Figure 1b]. Digital subtraction angiography (DSA) revealed occlusion of the V4 portion of the right VA [Figure 1c], stenosis of the V4 portion of the left VA [Figure 1d], and occlusion of the left supraclinoid ICA [Figure 1e]. No clear stenotic lesion was observed in the superficial temporal artery (STA) [Figure 1f]. Electrocardiography showed sinus rhythm, and echocardiography showed no finding suggestive of cardiogenic cerebral embolism. The C-reactive protein (CRP) level was 1.06 mg/dL in blood testing, and the erythrocyte sedimentation rate (ESR) was not measured. The patient was diagnosed with atherothrombotic brain infarction and treated with the oral administration of aspirin and clopidogrel.

However, dysarthria was aggravated 2 weeks later. MRI revealed a new cerebral infarction in the left pons [Figure 2a]. DSA showed aggravation of the stenotic lesion in the left VA and laminar flow in the right posterior cerebral artery

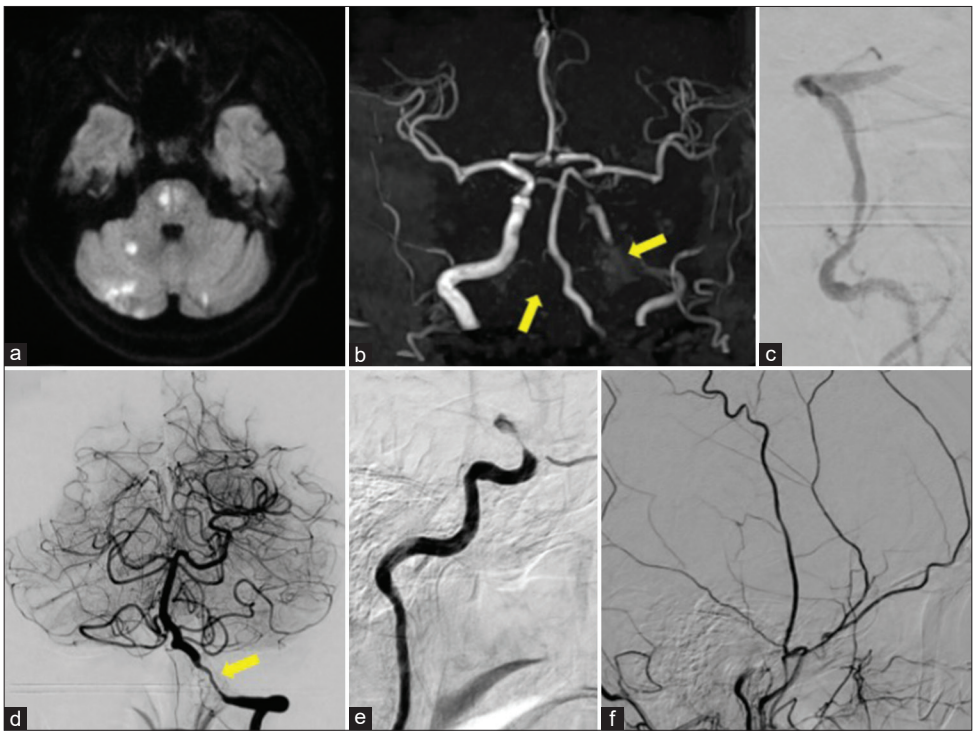

Figure 1: (a) Diffusion-weighted image revealed cerebral infarctions in the left and right sides of the pons and cerebellum, and (b) the left internal carotid artery (ICA) and the right vertebral artery (VA) were not well visualized in magnetic resonance angiography (arrow). Digital subtraction angiography revealed (c) occlusion of the V4 portion of the right VA and (d) stenosis of the V4 portion of the left VA (arrow), as well as (e) occlusion of the left supraclinoid ICA. (f) Meanwhile, no clear stenosis of the superficial temporal artery was observed.

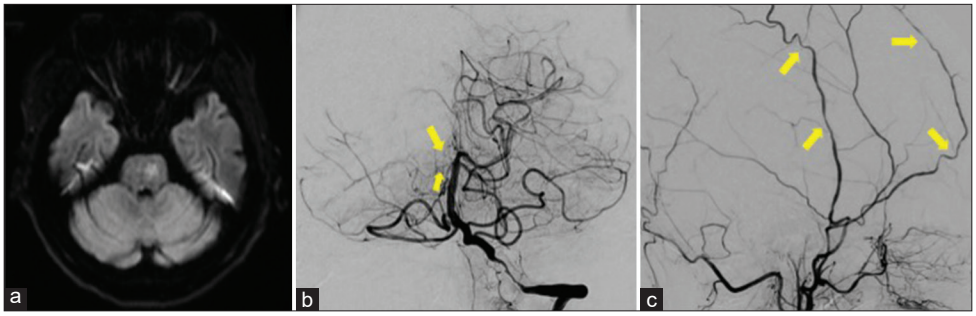

Figure 2: (a) Diffusion-weighted magnetic resonance imaging revealed a new cerebral infarction in the left pons. (b) In the front view of digital subtraction angiography (DSA), the right posterior cerebral artery and the right superior cerebellar artery showed laminar flow (arrow) due to blood flow through the right posterior communicating artery, and blood flow from the left vertebral artery was predicted to be reduced. (c) The lateral view of DSA revealed a narrowing of the superficial temporal artery at multiple sites (arrow).

and superior cerebellar artery due to blood flow through the right posterior communicating artery [Figure 2b]. Moreover, multiple stenotic lesions were observed in the STA [Figure 2c]. We obtained images acquired at the onset of headaches from the previous doctor. As the MRA data showed no vascular lesions, such as stenosis or occlusion, in cerebral vessels [Figure 3], we understood that the vascular lesions were aggravated rapidly within 3 months. In addition, T1 black-blood post-gadolinium imaging sequence MRI revealed vessel wall enhancement (VWE) in the left and right VA, left ICA, and left and right STA [Figures 4a-d]. The CRP level was 0.66 mg/dL and the ESR was 17 mm/1 h in blood testing. The level of myeloperoxidase-anti-neutrophil cytoplasmic antibody, serine proteinase3-anti-neutrophil cytoplasmic antibody, anti-cyclic citrullinated peptide antibody, anti-double stranded DNA immunoglobulin G, and anti-Sm antibody was normal. The patient was suspected of having vasculitis, and a temporal artery biopsy was performed before she was finally diagnosed with GCA [Figures 5a and b].

Oral administration of prednisolone 60 mg/day was initiated. Afterward, cerebral infarction did not relapse, the prednisolone dose was gradually reduced, and antiplatelet drugs were changed to aspirin alone. The patient recovered sufficiently to walk independently and was discharged 2 months later. As of 9 months after discharge, the oral prednisolone dose was gradually reduced to 10 mg/day, and MRA showed no aggravated intracranial vascular lesions [Figure 6], and very mild dysarthria was the only residual neurological symptom.

DISCUSSION

Typical pathological findings in GCA are giant cell granulomas formed in the vicinity of destroyed internal elastic lamina and inflammation affecting all the adventitia, media, and intima of arteries.[18] These pathological changes can cause stenosis, occlusion, and although rarely, dissection of cerebral arteries, leading to cerebral infarction.[10] Meanwhile, intracranial cerebral arteries have a very thin wall and very low levels of elastic fibers in the media and adventitia.[16,17] Therefore, intracranial vascular lesions are rarely found in patients with GCA.[12]

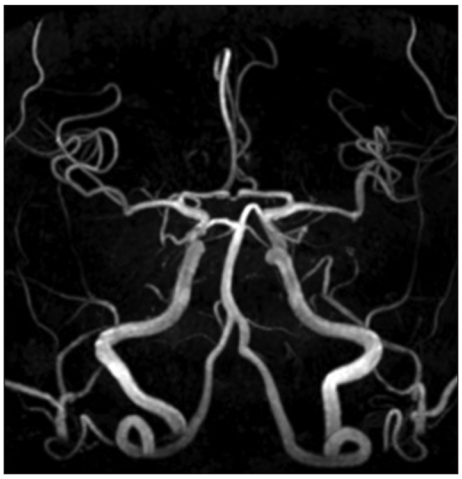

Figure 3: No intracranial vascular lesions were observed in the magnetic resonance angiography acquired at the onset of the signs of giant cell arteritis.

It is unknown when the progression of intracranial vascular lesions starts in patients with GCA. According to Alsolaimani *et al*., the signs of GCA preceded CVA in as many as 64% of cases and occurred concurrently with CVA in 17%, while 19% had no signs of GCA. In addition, there was a median interval of 60 days from the onset of the signs of GCA to the onset of CVA.[1] To the best of our knowledge, cerebrovascular evaluation at the onset of the signs of GCA has never been performed in cases with the

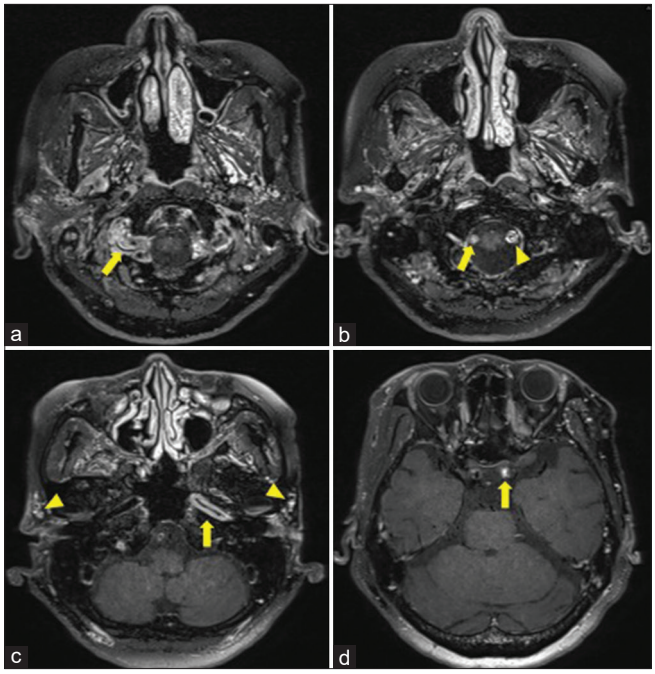

Figure 4: T1 black-blood post-gadolinium imaging sequence magnetic resonance imaging. (a) Vessel wall enhancement (VWE) of the V4 portion of the right vertebral artery (VA) was observed (arrow). (b) More distally, the right VA was occluded (arrow), and the VWE of the V4 portion of the left VA was observed (triangle). (c) VWE was observed in the petrous portion of the left internal carotid artery (ICA) (arrow), and VWE was also observed in the left and right superficial temporal artery (triangle). (d) Occlusion was observed in the supraclinoid portion of the left ICA (arrow).

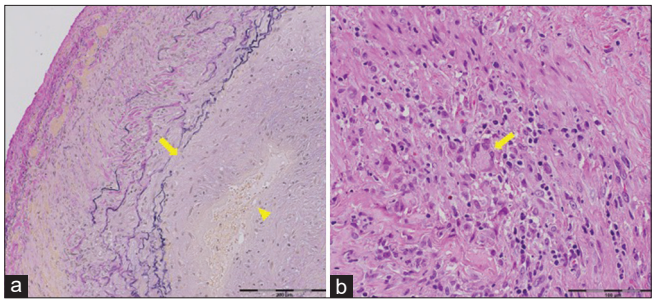

Figure 5: Photographs of pathological sections of the biopsied temporal artery. (a) Elastica van Gieson staining revealed the rupture of the elastic lamina near the intima (arrow) and the narrowing of the vessel lumen due to the marked thickening of the intima (triangle). (b) Hematoxylin and eosin staining revealed multinucleated giant cells in the media (arrow) and infiltration of histiocytes and lymphocytes.

GCA signs preceding CVA.[1,7,9,10,12-15,19] In this case, MRA was performed at the onset of the signs of GCA and revealed no intracranial vascular lesions. Furthermore, despite improving the headache, multiple severe intracranial vascular lesions were observed 3 months later. This suggests that in patients

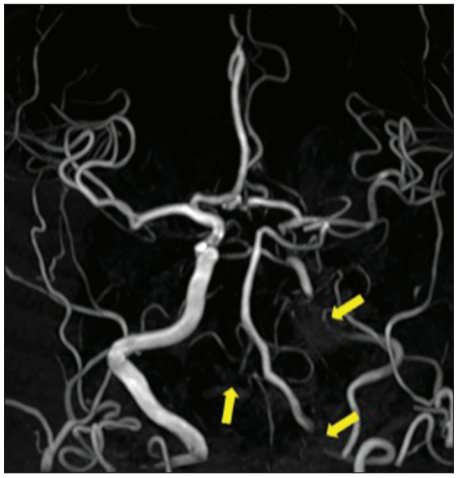

Figure 6: In the magnetic resonance angiography acquired 9 months after discharge, no progression of intracranial vascular lesions was observed, but the visualization of the left internal carotid artery and the right vertebral artery (VA) did not improve, and the left VA also remained poorly visualized (arrow).

with GCA, intracranial vascular lesions may start developing later than the signs of GCA and progress regardless of the improvement in the signs of GCA. In cases without the signs of GCA or those with the GCA signs occurring concurrently with CVA,[1,7,13] intracranial vascular lesions might have been aggravated while no signs of GCA were observed, and earlier slight signs might have been overlooked. Physicians should be fully aware of the possible delayed formation of intracranial vascular lesions in patients with GCA.

Intracranial vascular lesions found in middle-aged and older patients with cerebral infarction are difficult to determine whether they are arteriosclerotic lesions or vasculitic lesions. In fact, in multiple case reports on intracranial GCA, including the present case, patients were diagnosed with atherothrombotic brain infarction at the onset of cerebral infarction and received standard treatment.[7,14] In patients with GCA, evaluating intracranial vessels as a control is useful for distinguishing them from arteriosclerotic lesions at the onset of CVA. In patients with CVA, it is also necessary to perform medical examinations with GCA in mind. Imaging findings suggest that vasculitis includes bilateral long lesions.[2,7] In this case, lesions were found in the left and right VA, and the one in the left VA was an unusually long lesion. Furthermore, T1 black-blood post-gadolinium imaging sequence MRI can visualize inflammation of blood vessel walls and can be an additional diagnostic tool for GCA.[11] In fact, VWE was noted where the lesions were observed in MRA and DSA, suggesting vasculitis. Based on these findings, we could make the diagnosis of intracranial GCA and provide therapeutic intervention early.

Unlike extracranial GCA, intracranial GCA has very poor prognoses, even with glucocorticoid or immunosuppressant treatment.[12,13] Therefore, it is necessary to make strategies for the early diagnosis of intracranial GCA. In the consultation of patients with CVA, as a first step, physicians should conduct a detailed interview about the signs of GCA experienced over several months and attention should be paid to the increased inflammatory response of unknown causes in blood tests. In the next step, physicians confirm imaging findings suspicious for intracranial GCA. Specifically, besides the bilateral long lesions, the ICA and VA are frequently affected in patients with intracranial GCA.[6,7] Moreover, the conjunction of headache with stroke caused by bilateral vertebral and basilar arteries involvement in the elderly is highly suggestive of intracranial GCA.[7] Finally, T1 black-blood post-gadolinium imaging sequence MRI should be considered to confirm the presence of vessel wall inflammation. We believe that these practices as soon as possible will lead to the early diagnosis of intracranial GCA. Regarding the management of patients with previously diagnosed GCA, physicians should be fully aware of the possible delayed formation of intracranial vascular lesions, and it is important to conduct regular imaging tests. In this case, the patient was initially not suspected of having vasculitis as there were no typical signs of GCA or clear blood test findings indicating inflammation at the onset of cerebral infarction. However, we realized that the intracranial vascular lesions were not typical arteriosclerotic lesions because they were bilateral and rapidly aggravated. Then, we understood that multiple cerebrovascular lesions aggravated rapidly in 3 months based on MRA images taken at the onset of GCA by the previous doctor, which showed that the intracranial vascular lesions were absent 3 months ago. Finally, the patient was suspected of having vasculitis and was diagnosed and treated relatively early with favorable outcomes.

CONCLUSION

In patients with GCA, the progression of intracranial vascular lesions may begin later than the signs of GCA and continue despite the improvement in the symptoms of GCA. Physicians should be fully aware of the possible delayed formation of intracranial vascular lesions in patients with GCA, and continuous evaluation of intracranial vessels since the onset of GCA may be helpful for early diagnosis at the onset of CVA. In addition, T1 black-blood post-gadolinium imaging sequence MRI may lead to early diagnosis and treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The author(s) confirms that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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