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Case Report

Giant cell arteritis with simultaneous onset of multiple intracranial vascular occlusions: A case report

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ABSTRACT

Background: Giant cell arteritis (GCA) causes severe stenosis or occlusion of the arteries but rarely affects the intracranial arteries. We report a rare case of GCA along with autopsy results.

Case Description: A 69-year-old man developed gait disturbance due to vertebral artery (VA) occlusion. As is common in atherothrombotic stroke, dual antiplatelet therapy was administered. The patient's symptoms improved temporarily. However, his symptoms relapsed and his consciousness was acutely disturbed. Digital subtraction angiography revealed an appearance of stenosis of the internal carotid artery (ICA) C2 portion on the right side and decreased retrograde basilar artery (BA) blood flow through the right posterior communicating artery. Balloon angioplasty was performed, and BA blood flow increased. GCA was suspected, and a definitive diagnosis was made based on temporal artery biopsy findings. Steroid therapy was initiated but failed to control disease progression, and the patient died. The autopsy findings revealed GCA in the bilateral ICAs and VAs, and no signs of GCA were found in other intracranial arteries, despite occlusion on magnetic resonance angiography.

Conclusion: GCA of the intracranial blood vessels is rare and might be more likely to occur in the ICAs and VAs than in other intracranial blood vessels. GCA of the intracranial blood vessels has a poor prognosis, and as such, if rapid changes are observed in the ICAs or VAs, GCA should be considered a part of the differential diagnosis and immediate treatment should be administered.

Keywords: Angioplasty, Autopsy, Giant cell arteritis, Multiple intracranial vascular occlusions

INTRODUCTION

Giant cell arteritis (GCA) is a chronic granulomatous vasculitis of the large- and medium-sized arteries, and predominantly affects the extracranial arteries.[19] Cerebral infarction is a complication that occurs in 3–7% of GCA patients, which is a related vasculitis of extracranial arteries causing vertebral or internal carotid artery (ICA) stenosis.[8,11,17] Although some cases of GCA affecting the intracranial cerebral arteries have been described,[1,2,5,9,14-16,18] the case descriptions mainly included the results of imaging examinations, and none included cross-referencing with angiography of endovascular-treated intracranial lesions and autopsy results. We report a rare case of GCA with rapidly progressive multiple intracranial arterial occlusions that were refractory to endovascular therapy and had a fatal outcome, along with the subsequent autopsy results.

CASE DESCRIPTION

A 69-year-old man with a 1-month history of intermittent headaches and dizziness was transferred to our hospital due to a sudden onset of gait disturbance. The patient had untreated mild hypertension. Physical examination showed ataxia, but no other neurological deficits were noted. He also had a sore throat and his body temperature was 37.5°C. Magnetic resonance imaging (MRI) showed acute ischemic infarctions on both sides of the cerebellum [Figure 1a]. Magnetic resonance angiography (MRA) showed stenoses of the left ICA and bilateral vertebral artery (VA) occlusions [Figure 1b]. Digital subtraction angiography (DSA) revealed retrograde flow into the upper segment of the basilar artery (BA) through the right posterior communicating artery (PCoA) [Figure 1c]. Laboratory tests revealed the following results: low-density lipoprotein cholesterol, 92 mg/dL (optimal); C-reactive protein (CRP), 5.59 mg/L (elevated); and erythrocyte sedimentation rate, 110 mm/h (elevated). As is common in atherothrombotic stroke, dual antiplatelet therapy (daily aspirin dose of 100 mg and clopidogrel dose of 75 mg) was administered. A few days later, the CRP level decreased and fever was resolved so that we considered the elevated CRP on admission to be the result of viral infection.

After rehabilitation, the symptoms of cerebellar ataxia improved and he was discharged on day 21. At discharge, clopidogrel was changed to cilostazol (200 mg/day) as the latter has a lower risk of bleeding complications. However, dysarthria and gait disorders relapsed on day 32, and the patient was readmitted. The MRI showed diffuse cerebral infarctions in the right cerebellar peduncle and the left cerebellar hemisphere. DSA detected a decline in retrograde BA blood flow, which resulted in obstruction in the right anterior inferior cerebellar artery [Figures 1d-f]. Based on these results, the antiplatelet drug medication was restored to aspirin 100 mg/day and clopidogrel 75 mg/day.

On day 36, his consciousness was acutely disturbed**,** and MRI and DSA were urgently performed. DSA revealed an appearance of stenosis of the ICA C2 portion on the right side and decreased retrograde BA blood flow through the right PCoA, which was not observed at his first admission [Figures 2a,b]. These findings were suspected to be the cause of his consciousness disturbances [Figure 2c]. Therefore, balloon angioplasty was performed to dilate the right ICA stenosis. This treatment successfully increased the blood flow in the BA [Figure 2d], but the symptoms did not improve much. Vasculitis was suspected due to multiple unexplained stenoses that progressed in a short period of time and the increase in inflammation. Vessel wall thickening of the bilateral superficial temporal artery (STA) was confirmed in an echogram, and a STA biopsy was performed, demonstrating inflammatory reactions and giant cells in the vessels; therefore, the diagnosis of GCA was confirmed

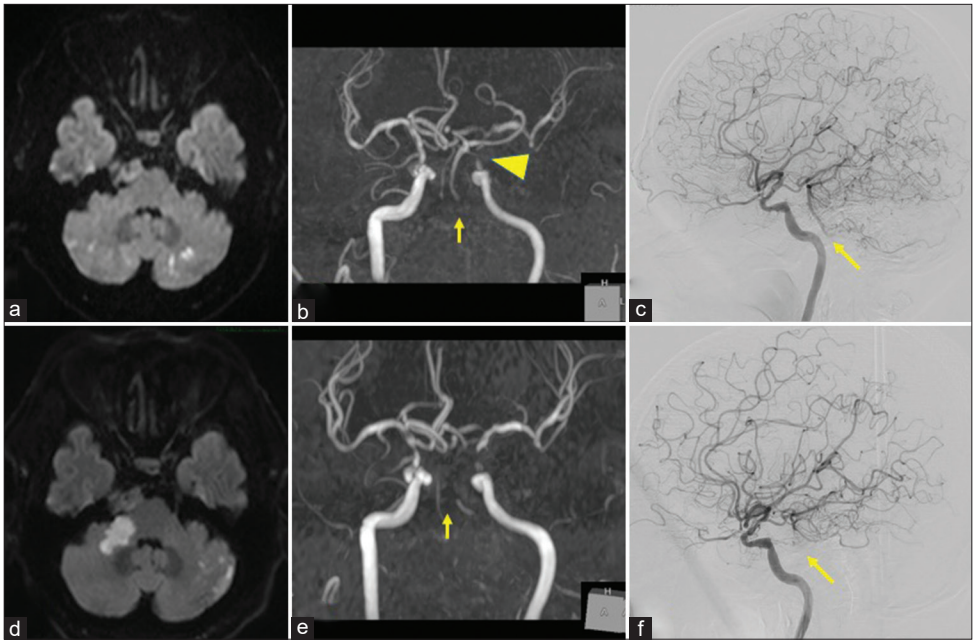

Figure 1: (a) Diffusion-weighted MRI showed bilateral cerebellar infarctions, and (b) MRA showed the left ICA stenosis (arrowhead) and the complete occlusions of the bilateral vertebral arteries (arrow) on admission. (c) Lateral view of the angiography showed retrograde filling of the BA and AICA from the right PCoA (arrow) on admission. (d) Diffusion-weighted MRI showed infarctions in the right cerebellar peduncle and the left cerebellar hemisphere, and (e) MRA showed poor visualization of the BA. (f) Lateral view of the angiography showed a decline in retrograde BA blood flow (arrow) compared to the image at admission. AICA: Anterior inferior cerebellar artery, BA: Basilar artery, ICA: Internal carotid artery, MRI: Magnetic resonance imaging, MRA: Magnetic resonance angiography, PCoA: Posterior communicating artery.

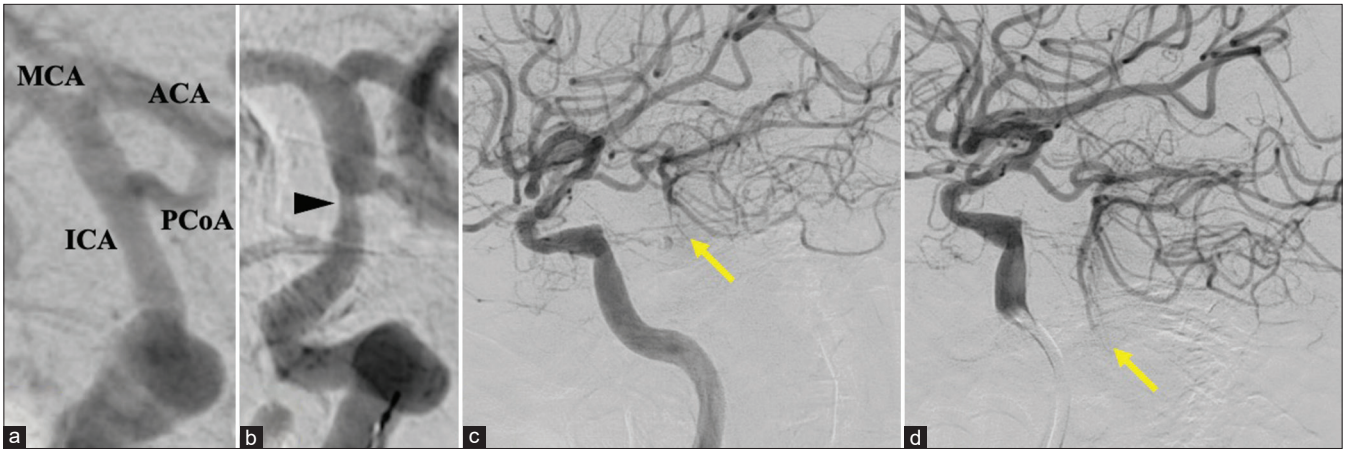

Figure 2: (a and b) Anteroposterior views of the cerebral angiography. (a) The right ICA at the C2 portion on admission. There was no sign of stenosis, (b) but severe stenosis appeared after his level of consciousness was decreased. (c and d) Lateral projections of the cerebral angiography of the right ICA. (c) Before angioplasty, (d) after angioplasty. Note that retrograde flow in the BA through the PCoA due to the bilateral VA occlusion was improved (arrow) because of the vasodilation of the right ICA. ACA: Anterior cerebral artery, BA: Basilar artery, ICA: Internal carotid artery, MCA: Middle cerebral artery, PCoA: Posterior communicating artery, VA: Vertebral artery.

pathologically. After the patient was treated with pulse dose steroid therapy (methylprednisolone 1 g intravenously for 3 days) followed by prednisolone (50 mg daily through a gastric tube), CRP levels temporarily improved. However, the progression of multiple vessel stenoses and occlusions could not be controlled, and the cerebral infarction worsened [Figure 3]. Extensive cerebral and brain stem infarction led to respiratory failure and the patient died on day 51 because he had not wanted any life-prolonging treatment, including intubation.

Autopsy result

The autopsy revealed cerebral infarctions in the lower right medulla and reticulum, which were thought to be the cause of respiratory depression. Microscopically, internal elastic lamina (IEL) disruptions with giant cells, granuloma, as well as transmural inflammatory infiltrates were observed in the bilateral ICAs and VAs [Figures 4a-c]. The internal elastic plate destruction of the right ICA was observed only at the site with inflammatory reaction and giant cells, so a balloon angioplasty was not indicated. The middle cerebral artery (MCA) and BA showed atherosclerosis, but no signs of GCA were found. Similarly, the bilateral anterior cerebral artery (ACA), posterior cerebral artery (PCA), superior cerebellar artery, and anterior inferior cerebellar artery also showed no signs of GCA and stenosis. In the extracranial arteries, GCA features were found in the thoracic aorta, left subclavian artery, and iliac artery. This suggested that the GCA developed systemically and spread to the ICAs and VAs, resulting in reduced intracranial blood flow and ultimately death due to extensive cerebral and brain stem infarction.

DISCUSSION

GCA predominantly affects the branches of the aorta, especially the branches of the ophthalmic and external carotid arteries, and rarely involves cerebral infarction.[19] The most cerebral infarctions associated with GCA are caused by hemodynamic insufficiency due to severe stenosis of extracranial blood vessels and by occlusion of extracranial blood vessels.[14,18] In the presented case, the autopsy revealed inflammatory cell infiltration of the IEL in bilateral VA that was obstructed from the beginning, left ICA stenosis, and right ICA where stenosis progressed gradually. Giant cells were also observed in some lesions. Although bilateral ACA, PCA, and right MCA were obstructed on the premortem MRA, the autopsy revealed no signs of vasculitis. In the epidural blood vessels, the subclavian artery was affected by arteritis, whereas the thoracic aorta had extensive atherosclerotic lesions and some arteritis lesions based on the pathological findings. Considering the autopsy, radiological, and clinical findings together, GCA resulted in the occlusion of the bilateral VAs at an early stage, narrowed the left ICA, and then spread inflammation to the right ICA, resulting in rapid stenosis. This rapid progression of GCA-induced inflammation to multiple intracranial vessels is rare, and studies which have assessed such cases with pathological results of intracranial blood vessels are also rare.[2,5,16,18] In our case, GCA affected the intracranial ICA and VA, but did not affect other vessels such as the MCA, ACA, and BA, which were also located in the intracranial portion. Some GCA cases demonstrated an affected PCA and MCA in radiographic features, but most of them did not show pathological results.[1,15,17] As far as we are aware, there are only a few reports showing pathological results of GCA

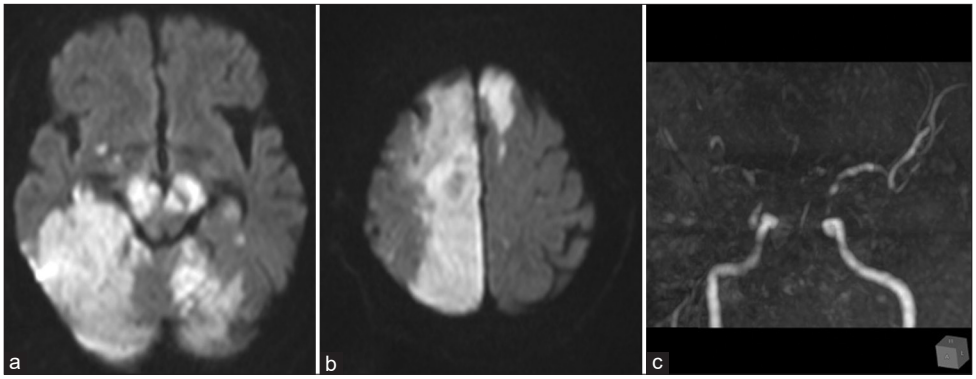

Figure 3: In spite of steroid therapy, (a and b) magnetic resonance imaging showed an exacerbation of the cerebral infarctions and (c) bilateral anterior cerebral arteries, the right internal carotid artery, right middle cerebral artery, and basilar artery were suspected to be occluded.

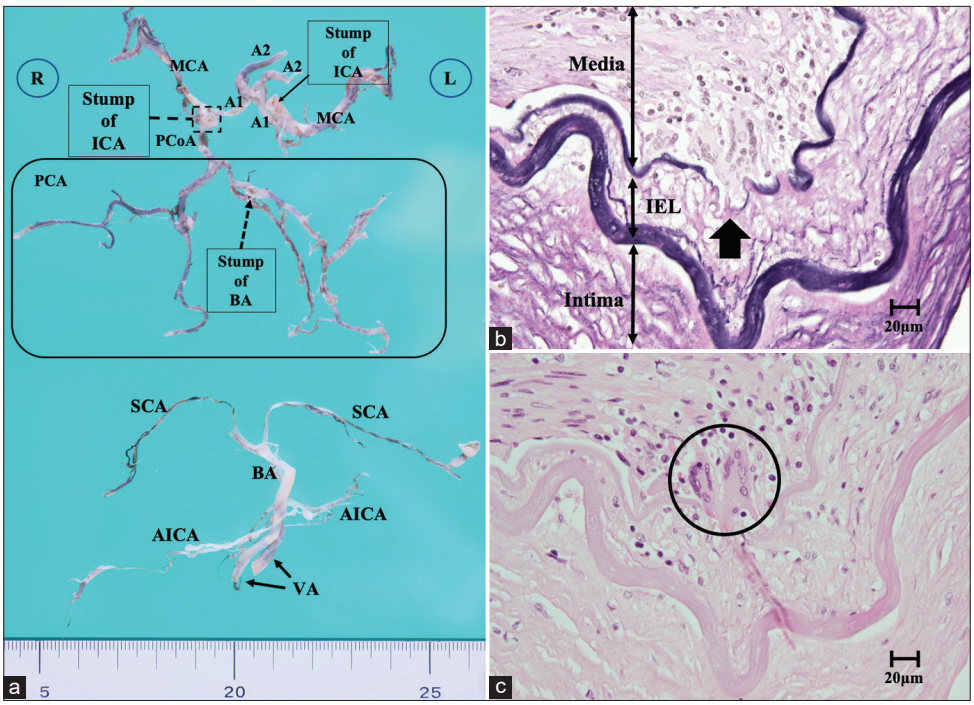

Figure 4: (a) Macroscopic photograph of the intracranial arteries. The left PCoA was hypoplasia, and the bilateral ICAs were cut at the C2 portion, and the BA was disconnected at the top. (b and c) Microscopic sections of the right ICA at the C2 portion. (b) Elastica van Gieson staining ×400, (c) hematoxylin and eosin staining ×400. All layers of the arterial wall are severely affected by the arteritis, and multinucleated giant cells invade the junction of the media and intima (circle). The IEL was also destroyed (arrow). A1: Anterior cerebral artery A1 segment, A2: Anterior cerebral artery A2 segment, AICA: Anterior inferior cerebellar artery, BA: Basilar artery, IEL: Internal elastic lamina, ICA: Internal carotid artery, MCA: Middle cerebral artery, PCoA: Posterior communicating artery, PCoA: Posterior communicating artery, SCA: Superior cerebellar artery, VA: Vertebral artery.

affecting intracranial arteries other than the ICA and VA.[4,12] As in our case, even in cases where MRA suggests that the ACA, MCA, or PCA stenosis is due to GCA, pathology results may not prove it. Pathological diagnosis is essential because it needs to be differentiated from atherosclerosis and other vasculitis such as primary central nervous system vasculitis.[3] Considering these cases, inflammation due to GCA rarely occurs in the MCA and BA that do not have an extracranial portion, on the other hand, it might be possible in the ICA and VA. An autoimmune reaction to the IEL has been suggested in GCA, therefore, intracranial blood vessels are supposed to be less likely to be inflamed due to the smaller amounts of elastic fibers.[14] Embryologically, the VA and BA are different vessels that anastomose during development, and it is considered that the ICA distal to the PCoA and the ICA proximal to it are embryologically different structures and cannot be treated identically.[7,10] This may be the reason for the regional distribution of inflammation by GCA.

According to the past reports involving inflammation of intracranial blood vessels, vasculitis progresses despite prednisolone administration, and the prognosis is often poor.[1,14] Therefore, it is important to differentiate GCA at an early stage and start treatment immediately. Although it is difficult to make a decision when there are complications such as infection, the inflammatory findings in the laboratory data and the fact that GCA often causes inflammation of the VA may be helpful.[11,17] In addition, stenosis in extracranial blood vessels such as the STA may be helpful in the diagnosis. In fact, the initial angiography in this case, with retrospective review, showed that blood flow in the STA was narrowed and delayed. Similar to the treatment option for GCA, except for steroids, both surgical and endovascular treatment options for intracranial stenoses and occlusions have been reported.[6,9,13] Successful endovascular treatment with balloon vasodilation and stenting has also been described in several patients with bilateral GCA-induced ICA stenosis that had progressed to cerebral infarction, despite medical treatment with immunosuppressants and antiplatelet drugs.[9] Furthermore, Dementovych *et al*. reported the successful treatment of complete VA occlusion secondary to GCA using endovascular intracranial angioplasty and stent placement.[6] In our case, recanalization of the VA may have been difficult because a good amount of time had passed since the complete occlusion of the VA and the lesion was long. Immediately following vasodilation with a balloon catheter in the ICA resulted in improved blood flow in the BA; however, the progression of stenosis in the intracranial artery, including the right ICA, could not be stopped, with subsequent worsening of the cerebral infarction. At the autopsy, marked luminal stenosis was observed in the treated right ICA. The effect of balloon dilation on the intracranial stenosis caused by GCA was only temporary; thus, a stent placement might have been needed. To the best of our knowledge, this is a rare case describing cross-referencing with angiography of endovascular-treated intracranial lesions and autopsy results of intracranial vessels severely impaired by GCA. Further studies of the pathology of intracranial GCA and treatment options, such as endovascular treatment, are needed.

CONCLUSION

We describe a case of GCA with extensive cerebral and brainstem infarction due to progressive stenoses of multiple intracranial arteries. GCA of the intracranial blood vessels is rare and might be more likely to occur in the ICAs and VAs than in other intracranial blood vessels. If multiple rapid changes in the ICA and VA are observed, GCA needs to be differentiated and immediate treatment is required.

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.

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