Case Report

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Recurrent ischemic stroke from reversible extracranial internal carotid artery and middle cerebral artery vasospasm: A case report

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Abstract:

Idiopathic internal carotid artery (ICA) vasospasm is a rare cause of ischemic stroke. Its pathophysiology remains unclear and diagnostic and treatment protocols are yet to be defined. A 45-year-old male, presenting with recurrent transient dizziness, blurred vision, and speech disturbances, was diagnosed with recurrent ischemic stroke caused by bilateral ICA and middle cerebral artery (MCA) vasospasm, and the vascular ultrasound and imaging techniques have grabbed the reversible changes in a short time. This case underscores the importance of considering idiopathic ICA vasospasm as a potential cause of recurrent ischemic stroke, even in the absence of common diagnostic markers. The case also indicates the possible, albeit rare, involvement of the MCA in this condition. Therefore, it is crucial to maintain a high index of suspicion for idiopathic ICA vasospasm in similar clinical presentations and to explore more inclusive diagnostic criteria.

Keywords:

Idiopathic carotid artery vasospasm, ischemic stroke, recurrent cerebral infarcts

Introduction

Idiopathic internal carotid artery (ICA) vasospasm remains a rare and under-recognized cause of ischemic stroke, which is clinically characterized by recurrent cerebral infarctions coupled with the reversible occlusion or stenosis of the ICA.[1,2] Despite the reports of about 30 cases to date, a common characteristic of each has been the exclusive involvement of the extracranial ICA. In this report, we present a novel case of recurrent ischemic stroke caused by the both middle cerebral artery (MCA) and the bilateral ICA vasospasm. This case may illuminate new clinical manifestations of idiopathic ICA vasospasm, informing future diagnostic and therapeutic strategies, particularly for young patients.

Case Report

Our patient, a 45-year-old male was admitted to our department after experiencing recurrent transient dizziness and blurred vision for more than 20 years. In addition, these symptoms had progressively intensified, accompanied by speech disturbances and sluggish 9 months before admission.

Initially, the patient neglected medical consultation despite the recurrent blurred vision and dizziness due to the mild and transient nature of his symptoms. However, 10 years prior to the current admission, his symptoms escalated to included increased dizziness with transient blindness in both eyes. After finally seeking medical attention, an initial magnetic resonance imaging (MRI) was suggested. Magnetic

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resonance angiography (MRA) revealed severe stenosis of the right MCA without the presence of any cerebral infarctions [Figure 1]. However, a Digital Signature Algorithm (DSA) was performed 3 days later which reported mild stenosis in the distal right ICA, while the right MCA appeared normal [Figure 2]. This was the extent of the workup, and the patient had no further follow-ups until the aforementioned transient symptoms started to once again worsen.

Nine months before the current admission, the patient was referred for exacerbated dizziness, loss of vision in the left eye, speech disturbances, sluggishness, and memory impairment all following a bout of fatigue. The MRI revealed acute and subacute cerebral infarctions in the bilateral basal ganglia, corona radiata, centrum semiovale, and the frontal lobe. Concurrently, MRA images revealed bilateral occlusion of the ICA, MCA, and anterior cerebral artery [Figure 3]. Given that a transesophageal echocardiography identified a patent foramen ovale (PFO), the initial etiological diagnosis was cardioembolism. The patient underwent percutaneous PFO closure and was administered antiplatelet agents, and his speech impairment subsequently improved. However, a persistent tubular field of vision in the left eye remained. One month later, MRA was repeated and no abnormalities were detected.

Five months before the current admission, though seemingly asymptomatic, the patient was admitted to our department for further diagnostic evaluation and treatment. Neurological examination revealed incomplete motor aphasia, mild cognitive impairment, and visual disturbances. Further investigations including cerebrospinal fluid examination and whole-exome sequencing revealed no abnormalities with the exception of a missense mutation in the MTHFR. A computed tomography (CT) examination highlighted bilateral elongation of the styloid process. Notably, transcranial Doppler evaluation of cerebral hemodynamics, conducted in various head positions, disclosed a decrease in blood flow velocity of the left MCA with head rotation (relatively low pulsation changes in the spectral morphology, with an average decrease of 0.1 in P1 and approximately 19% in average peak velocity). Subsequently, the left styloid process was surgically amputated.

Despite the surgery, the patient continued to experience dizziness and blurred vision even 4 months after the surgery and was once again admitted to our department. Further imaging examinations revealed occlusion of the right ICA and severe stenosis of the left ICA [Figure 4], although no new infarctions were identified. On the following day, the DSA only showed occlusion of the right ICA. Based on the patient's medical history, nimodipine (30 mg, three times a day, 7 days, oral) and papaverine (60 mg, twice a day, 7 days, intravenous) were administered. The CT angiography demonstrated normalization of the corresponding arteries within 8 days of this intervention [Figure 4].

Nimodipine (45 mg, three times a day, oral) was continued after the patient was discharged. However, the patient still had transient ocular symptoms or dizziness

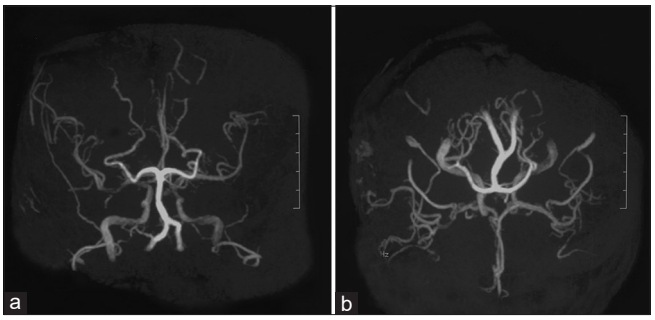

Figure 1: The magnetic resonance angiography in the superior-anterior (a) and superior-posterior (b) view showed the severe stenosis of the right middle cerebral artery 10 years ago

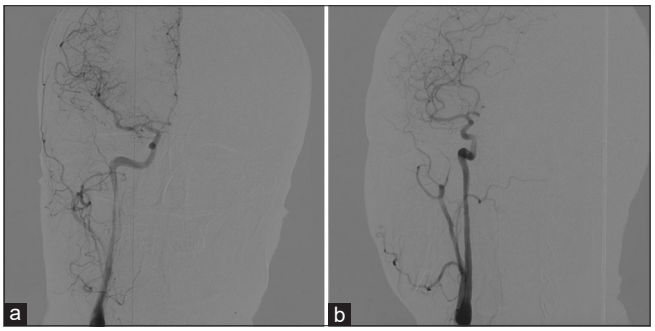

Figure 2: The Digital Signature Algorithm showed the mild stenosis of the distal right internal carotid artery in anterior-posterior (a) and lateral (b) views 10 years ago

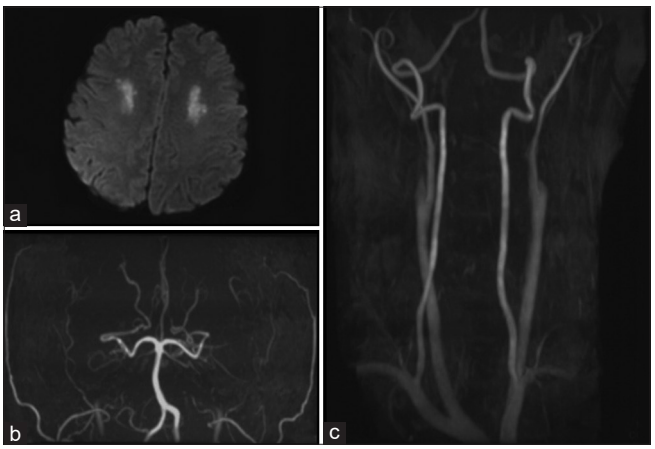

Figure 3: The images at 9 months ago. (a) The acute-subacute bilateral infarctions, (b and c) The occlusions of bilateral internal carotid artery, middle cerebral artery and anterior cerebral artery

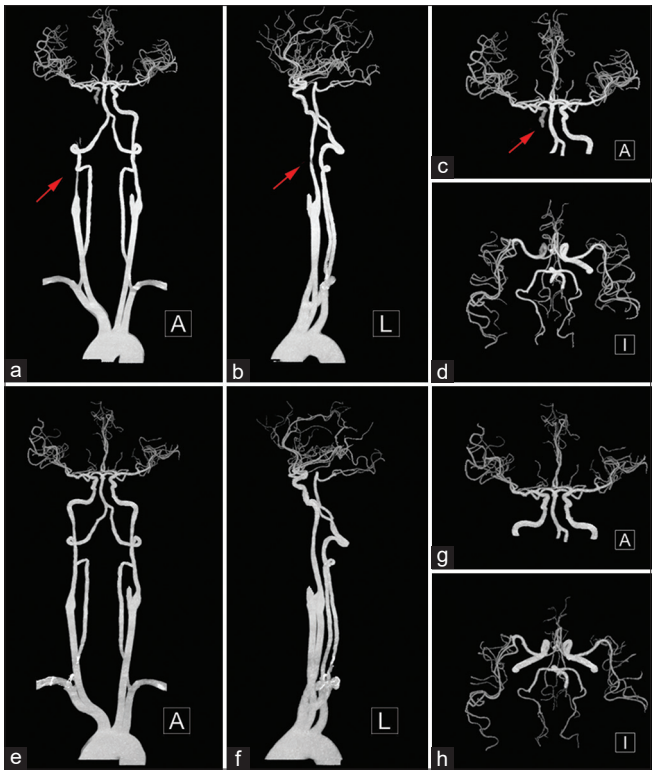

Figure 4: The computed tomography angiography at 4 months after surgery. (a-d) On admission, the occlusion of the right internal carotid artery (ICA) and the stenosis of the left ICA. (e-h) On follow-up at 8 days, bilateral ICAs were normal. The red arrow indicated the stenosis segment

during the 6-month follow-up and did not undergo further treatment.

Discussion

This case presents a young male patient diagnosed with an acute cerebral infarction secondary to reversible stenosis or ICA occlusion. Laboratory test results for inflammation, immunity, and heredity were unremarkable. The Eagle syndrome was taken into account because of elongation of the bilateral styloid processes and the changes of blood flow in the left MCA. Nonetheless, the assumption failed given that the attacks were independent of the head position and the persistent vascular changes occurred after the initial styloidectomy.[3-5] In light of the bilateral symmetric infarctions and reversible cerebrovascular stenosis/occlusion, a possible diagnosis of reversible cerebral vasoconstriction syndrome (RCVS) was considered. However, the absence of classic RCVS features, such as thunderclap headache, "string of bead" appearance on imaging, and involvement of vertebrobasilar arteries made this diagnosis less probable.[6]

Other than one instance of right MCA occlusion, the evidence from this patient's case primarily supports the diagnosis of idiopathic ICA vasospasm. The reversible vasospasms were mainly captured by MRA and DSA throughout the course of disease. DSA is the gold standard. However, comparing with DSA, the MRA revealed high sensitivity (70%) and specificity (97%) for intracranial stenosis, especially for 70%–100% stenosis.[7,8] In this case, severe stenosis of the right MCA was observed on MRA, which may be *in situ* stenosis or attributed to a decreased blood flow from the right ICA spasm. However, 3 days later, DSA still showed the stenosis of the ICA while MCA appeared normal. We considered that there was MCA vasospasm occurred. This rare cerebrovascular disease is predominantly characterized by the involvement of the ICA. Although the underlying mechanisms are still unclear, it is suspected that the lack of parasympathetic innervation in the extracranial ICA makes it sensitive to sympathetic vasomotor stimulation, resulting in vasospasms.[4,5,9] In this case, we postulated that overexertion or cold activated the patient's sympathetic nerve and promoted the release of vasoconstrictive neurotransmitters, which resulted in the changes in the carotid artery.[9,10] In addition, Tokunaga *et al*. put forward an idea that the ICA stenosis at the C1-2 vertebral levels may be relevant to the superior cervical ganglion, which may explain the residual ocular symptoms after carotid artery stenting.[2] While the involvement of the MCA in idiopathic ICA vasospasm remains largely unexplored, the presentation of this patient suggests that MCA involvement is plausible, although extremely rare.

To date, the absence of diagnostic guidelines for idiopathic ICA vasospasm and the lack of understanding of its underlying mechanisms present significant challenges. Potential reasons for the relative absence of MCA involvement in previous cases include the potential for less severe clinical manifestations that may not prompt seeking medical attention, thereby missing MCA stenosis altogether, the rarity of involvement, and the potential for involvement only in early stages or in younger patients. Further studies are warranted to investigate this condition's underlying mechanism and these theories and expand our understanding of the pathogenesis of idiopathic ICA vasospasms. Therefore, more cases are required to enrich and refine the clinical manifestations of this disease. Significantly, there is no standardized treatment at this point. Ganglion blocks, carotid artery stenting as well as conventional medications, such as calcium antagonist, steroid, papaverine, nitroglycerin, have not seen a long-term effective outcome.[1,2,4,11,12] The normal caliber of the ICAs had been restored in this patient using papaverine and nimodipine in the acute phase, but there were still attacks during the long-term follow-up. Further treatment measures need to be explored.

In summary, idiopathic ICA vasospasm is generally recognized to involve the ICA. However, this case

demonstrates that MCA involvement, although rare, is a possibility. As such, additional cases and further research are required to enrich our understanding of the clinical manifestations and potentially broaden the scope of this condition.

Ethics Committee approval and Declaration of Helsinki

Ethics Committee of Xuanwu Hospital, Capital Medical University (No.: [2019]093; Dated on Nov 26th, 2019) has approved the project, and the work that was undertaken conforms to the provisions of the Declaration of Helsinki.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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