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CASE REPORT

Atherosclerosis or Nonatherosclerotic Vasculopathy? Moyamoya Disease in a Mid-Aged Adult

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All causes of cerebrovascular disease should be considered in any patient presenting with ischemic stroke, despite the patient have concomitant risk factors for atherosclerosis. Here, we reported a case of Moyamoya disease (MMD) at a rather old age and with the presence of multiple risk factors for atherosclerosis. It is difficult to consider MMD at first glance, and it is easy to choose the wrong treatment in the first place. The key radiological feature for differentiation is the bilateral intracranial stenosis, rather than atherosclerotic changes, which are characterized as plaques with an uneven surface.

Key words: Atherosclerosis, moyamoya disease, stroke

INTRODUCTION

MMD is characterized by bilateral stenosis or occlusion of intracranial arteries and their branches. The most common presentations include ischemic stroke and transient ischemic attack. Here we describe the interesting clinical challenge posed by a 54-year-old woman, with multiple risk factors of atherosclerosis, presenting with acute stroke. Brain computed tomography (CT) revealed no hemorrhage. Magnetic resonance imaging(MRI) of the brain revealed acute cerebral infarction in the territory of the left middle cerebral artery and time-of-flight magnetic resonance angiography showed luminal narrowing at of the bilateral middle cerebral artery. We've consulted cardiologist for angioplasty or stenting but CT angiography demonstrated luminal stenosis without significant calcification, raising a suspicion of MMD. This case emphasizes that, in an adult with several predisposing risks for atherosclerosis, it is easy to consider such a patient as having atherosclerosis-associated ischemic stroke, without considering other causes, such as vasculopathy. Furthermore, angiography is the gold standard to confirm a diagnosis of MMD.

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CASE REPORT

The patient, a 54-year-old woman of Chinese ancestry, had received a diagnosis of Type 2 diabetes and hypertension 3 years earlier at other hospital, but she was noncompliant with medications. She presented to our hospital with a 3-day history of right-sided weakness and slurred speech. She denied having a headache, fever, or visual changes. The patient did not drink or smoke cigarettes; she reported no illicit drug use. She was afebrile, and her vital signs were normal at presentation. Physical examination showed the right central facial palsy and poor-extremity strength over the right upper and lower limbs. Her complete blood cell count, serum electrolytes, renal function, liver enzyme, and urine analysis were normal except the presence of dyslipidemia. Without the administration of contrast media, CT of the brain showed no evidence of hemorrhage, and following MRI of the brain revealed acute cerebral infarction in the territory of the left middle cerebral artery [Figure 1a]. Besides, luminal narrowing at the bilateral external and internal carotid arteries

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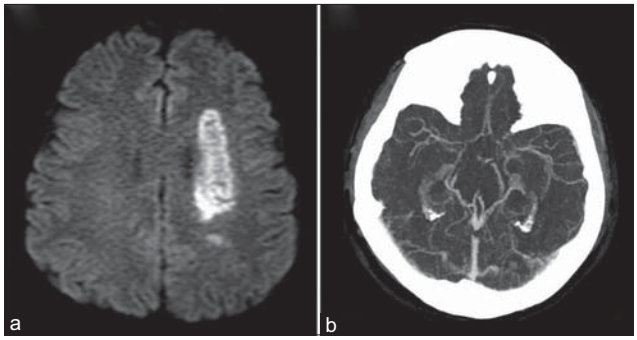

Figure 1: (a) Axial brain diffuse-weighted image magnetic resonance imaging. (b) Computed tomography angiography

was also noted. Under the impression of carotid stenosis, we consulted a cardiologist for scheduled angioplasty or stenting; however, preoperative CT angiography demonstrated luminal stenosis without significant calcification [Figure 1b], raising suspicion of MMD. The patient underwent conventional angiography [Figure 2], which confirmed the diagnosis of MMD. Through the process of shared decision-making, the patient opted to use medical treatment involving antiplatelet therapy. Both anti-diabetic and antihypertensive agents were administered according to the current guidelines. The focal neurological deficit improved after rehabilitation and no recurrent stroke developed during the 2-year follow-up period.

DISCUSSION

MMD, first described in 1957, is a chronic and progressive cerebrovascular condition that is characterized by bilateral stenosis or occlusion of intracranial arteries and their branches. Patients with the characteristic Moyamoya vasculopathy along with well-recognized medical conditions, such as sickle-cell disease, neurofibromatosis type 1, or Down's syndrome, are considered to have Moyamoya syndrome, whereas patients in whom the known risk factors are absent are classified as having MMD.[1] The definite etiology of MMD is unknown; current studies indicate that angiogenesis, genetic issue, and immune/inflammation play roles in the pathogenesis.[2] The epidemiological data display higher prevalence and incidence in the East Asian population.[2] MMD has a female preponderance; age at symptom onset has a bimodal distribution, with a major peak in children aged 5–9 years and another peak in adults in their 30s–mid-40s.[1]

The clinical findings range from ischemic to hemorrhagic stroke.[2] Ischemic stroke is more common in children, whereas hemorrhagic events are prevalent in adults.[1] Angiography, including conventional, magnetic resonance and CT angiography, is the gold standard for both diagnosis and the

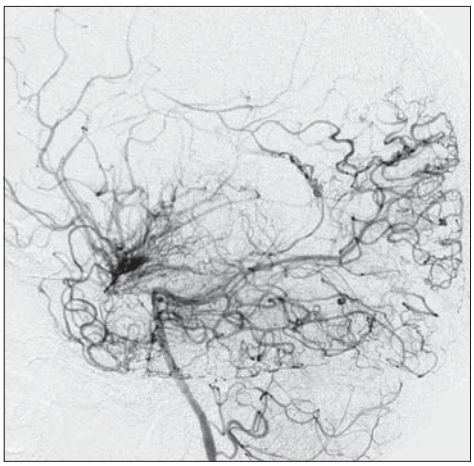

Figure 2: Conventional angiography showed prominent left lenticulostriate collaterals. Besides, the blood supply of the left middle cerebral artery is derived from the vertebral system and posterior communicating artery

assessment of MMD.[2] The Suzuki grading system, which is based on the severity and involvement of the moyamoya vessels in the internal carotid artery and external carotid artery (ECA) systems, classifies the angiographic findings into six categories.

Since the mechanism of MMD is under the hypothesis, there is no known cure for MMD. Practically, current treatments included surgical intervention and medication. Surgical revascularization, including direct (sparing the ECA for anastomosis), indirect (vascularized tissue contacting the brain) and combined bypass, is recommended to improve cerebral blood flow and ameliorating ischemic insult.[1,2] Medical therapy with antiplatelet agents, such as aspirin, has been used for the prevention of the recurrence of ischemic stroke in the long term.[2]

Endovascular techniques, including angioplasty and/or stenting, have been attempted; however, these are not recommended, due to the higher risks of recurrence and complications.[3] Carotid endarterectomy is suitable in patients with carotid stenosis due to atherosclerotic disease but not in patients with vasculopathies such as MMD. Since the choice of treatment is entirely different, distinguishing vasculopathy from atherosclerosis before initiating treatment is, therefore, important. In addition to CT scan in our case, high-resolution MRI technique is also helpful.[4]

While mid-aged-onset MMD is rare, clinicians should consider it in the differential diagnosis of patients presenting with ischemic stroke, including those with concomitant risk factors for atherosclerosis. Current treatments are designed for the secondary prevention of the recurrence of stroke. Endovascular intervention lacks evidence in the treatment of MMD.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her 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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