

Moyamoya disease in a young woman with intra- and extracranial vessels involvement on vessel wall imaging.

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INTRODUCTION

Moyamoya disease (MMD) was first described in 1957 as “bilateral hypoplasia of internal carotid arteries (ICAs)”.¹ Aside from involving the intracranial arteries, MMD can also affect extracranial ICAs and external carotid arteries (ECAs).²⁻⁴ High resolution magnetic resonance (MR) vessel wall imaging (VWI) is increasingly being used to help with the diagnosis and characterization of the condition focusing mainly on intracranial vessels and extracranial ICAs.⁵⁻⁹ We present a case of a young woman with MMD, demonstrating vessel wall enhancement of non-stenotic maxillary branches of bilateral ECAs.

CASE REPORT

A 25-year-old ethnic Chinese woman with no previous medical illness complained of left hand numbness 2 months before admission, which resolved after 2 weeks. The cervical spine MRI was normal. The nerve conduction study showed reduced amplitude of the left dorsal ulnar cutaneous nerve sensory potential. This was followed by an episode of left facial asymmetry 6 weeks later, which also resolved within one week. Being seen in another hospital, she was diagnosed as Bell’s palsy and was prescribed a short course of prednisolone. On the day of presentation to our hospital, she woke up with left sided body weakness involving her face and limbs. She was otherwise asymptomatic, not taking any other medications and had no family history of stroke.

Physical examination revealed a mild left upper motor neuron facial weakness. Her left upper limb power was 2/5 and lower limb power was 4/5. Her physical examination was otherwise normal.

Her investigations including full blood count, electrolytes, renal and liver function, erythrocyte

sedimentation rate, HIV status, homocysteine, HbA1c, antineutrophil cytoplasmic antibody, antinuclear antibody, double stranded DNA, complements, thyroid function test, rapid plasma reagin, lupus anticoagulant and thrombophilia screen tests, cerebrospinal fluid examination and echocardiogram were all normal. Her low-density lipoprotein was raised at 3.09 mmol/L (<2.59).

Her brain computed tomography (CT) showed a right anterior cerebral artery (ACA) territory infarct (Figure 1). MR angiogram and cerebral angiogram revealed long segment narrowing of the right ICA from bifurcation to the cavernous segment, attenuation and beaded appearance of all segments of both middle cerebral arteries (MCAs), ACAs, anterior communicating arteries, posterior communicating arteries and right posterior cerebral artery. The left ICA tapers at the distal end. Multiple net like collaterals showing puff of smoke appearance were seen. No significant stenosis of the ECAs vessels was seen on cerebral angiogram (Figure 2). MR vessel wall study using SPACE (Sampling perfection with application optimized contrast using different flip angle Evolution) revealed long segment of concentric wall enhancement of ICAs involving the petrous, cavernous and extracranial C1 segment (more on the right side). There was enhancement of both ECA maxillary artery branches (Figure 2).

The diagnosis of moyamoya changes secondary to vessel wall inflammation was considered in view of the extensive vessel wall enhancement. A right meningeal biopsy showed no evidence of vasculitis. She was given a 5 days course of intravenous methylprednisolone followed by low dose oral prednisolone. There was no recurrence of symptoms since. Follow up MR angiogram and VWI demonstrated persistent vessels wall enhancement and stenosis (Figure 3).

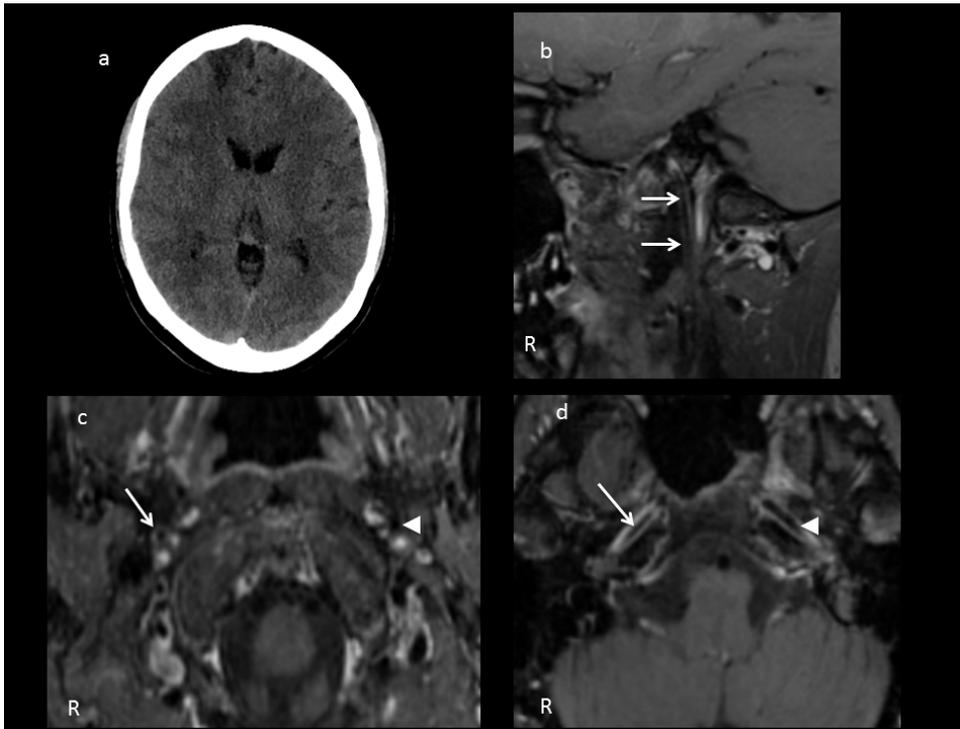


Fig 1. a: CT brain showing right ACA infarct. b-d: Vessel wall imaging using SPACE post gadolinium demonstrate concentric long segment right ICA vessel wall enhancement from C1 segment to distal ICA. For left ICA, vessel wall enhancement and narrowing only starts at foramen lacerum (arrowhead)

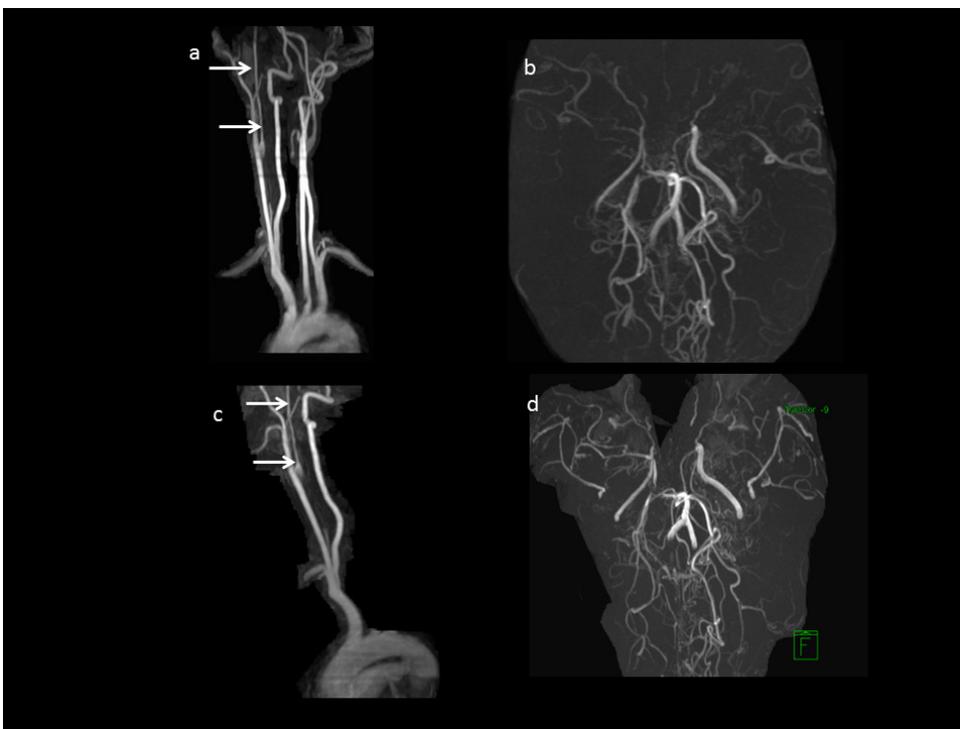


Fig 2. a-b: MRA Carotids and Circle of Willis (COW) demonstrating long segment right ICA stenosis and bilateral supraclinoid ICA stenosis with attenuation of both Carotid arteries and MCA. c-d: MRA carotids and COW 3 months after initial presentations and treatment demonstrating unchanged stenosis

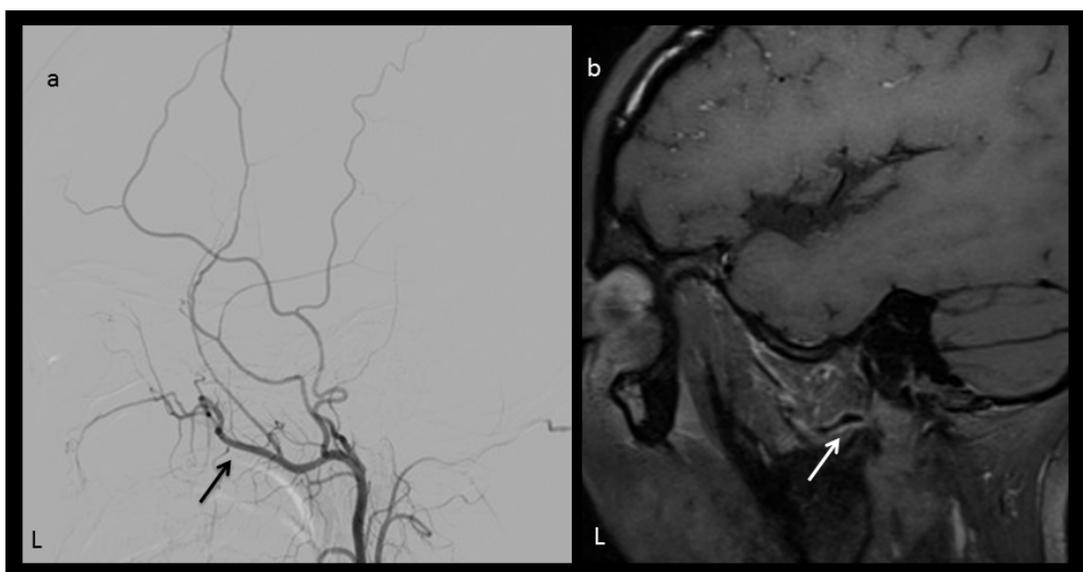


Fig 3. a: Left External carotid artery angiography demonstrating normal Maxillary artery branches (arrow)
 b: Vessel wall imaging using SPACE post gadolinium demonstrate concentric short segment left Maxillary arterial wall enhancement (white arrow)

DISCUSSION

MMD is classically characterized by progressive intracranial stenosis, especially bilateral distal internal carotid arteries (ICA), the proximal portion of the MCAs and ACAs.² The staging system for MMD first proposed by Suzuki and Takaku¹⁰ back in 1969 is still being used today (Table 1); commencing at the terminal portion of the ICA.¹¹ We believe that our patient has MMD based on the characteristic imaging features, and no other systemic pathology found. Our patient however, also has some atypical features, with extensive stenosis of extracranial ICAs and contrast enhancement of non-stenosed ECA branches on VWI.

The understanding on the changes in the external carotid system has improved with histopathological access to superficial temporal artery (STA) and middle meningeal artery (MMA) during bypass surgery. Yang *et al.*⁴ reported histopathological abnormalities in the STAs and MMAs of all his moyamoya cohort. In

angiographic studies, Hoshimaru *et al.*³ found that 20% of his moyamoya patients showed stenoses in the branches of the external carotid artery, while Komiyama *et al.*¹² found none in his 39 patients. Despite both being angiographic studies in Japanese moyamoya population, the discrepancy could be due to the angiography being performed at different stages of the disease or ECA stenosis happening infrequently in limited short segments that was missed. Extracranial ICA involvement in moyamoya are seen mostly in advanced disease. Using carotid ultrasound, Yasaka *et al.*¹³ and Yasuda *et al.*¹⁴ reported a reduction of the diameter at the proximal portion of the ICA, described as a champagne bottle neck sign appearance in 74% and 55.6% of their patients respectively, with all champagne bottle neck sign-positive arteries being exclusively present in those with Suzuki stage \geq III.¹⁴

In high resolution MR VWI, MMD typically shows concentric wall thickening with homogenous signal.^{5,15} Contrast enhancement of

Table 1: The Suzuki stages of Moyamoya disease

- Stage 1: Narrowing of the carotid bifurcation;
- Stage 2: Appearance of moyamoya vessels; dilatation of anterior and middle cerebral arteries;
- Stage 3: Intensification of moyamoya vessels; partial disappearance of anterior and middle cerebral arteries;
- Stage 4: Minimization of moyamoya vessels; advanced steno-occlusive changes of the ICA,
- Stage 5: Reduction of moyamoya vessels; absence of anterior and middle cerebral arteries;
- Stage 6: Disappearance of moyamoya vessels; only the collateral circulation from external carotid artery.

intracranial vessels is controversial, with some studies reporting no enhancement and others showing up to 90.6% of vessels demonstrating varying degrees of enhancement.⁵⁻⁹ This vast discrepancy on contrast enhancement may be due to the differing ethnic populations with heterogeneity of genetic background, the possibility of transient inflammatory changes, the presence of pseudoenhancements or slow flow artifacts and inclusion of patients with steno-occlusive disease other than MMD.^{6,7} The VWI of our patient demonstrated enhancement of bilateral ICAs, and both maxillary arteries in the setting of a normal ECA angiogram. As far as we know, there have been no previous reports on ECA enhancement in moyamoya patients on VWI. Previous studies have shown that vessel wall enhancement is closely related to progression of intracranial stenosis and may be seen in non-stenosed asymptomatic vessels.^{6,8} The ECA vessel wall enhancement here is potentially a marker for further stenosis changes as the disease advances. The discrepancy between the normal ECA angiogram and abnormal vessel wall enhancement also highlights the inherent limitation of luminal studies with conventional angiogram and emphasizes the advantage of MR VWI that allow depiction of vessel wall.¹⁶ Histopathologic correlation for MRI vessel wall enhancement is still scant but it has been said to correspond to vessel wall inflammation and/or increased *vasa vasorum density*.¹⁷ We postulate that histological changes of tunica intima thickening and fragmentation of elastica interna may have led to endothelial contrast leakage, causing enhancement on VWI.

An important differential diagnosis to consider in a young lady with cerebral infarcts and vessel stenosis is vasculitis. The biopsy of our patient however, did not show vasculitis changes. The patient also had no positive seromarkers to indicate a known vasculitis syndrome. We noticed similarities between our case and a recent case report on a woman with Takayasu arteritis, including the involvement of ECA and extracranial ICA on imaging.¹⁸ This indicate that there may be overlapping clinical features between moyamoya and Takayasu arteritis. Only one case of a patient with Takayasu arteritis and MMD have been reported previously.¹⁹

In conclusion, we present a case illustrating the varying involvement of intra- and extracranial vessels in a patient with MMD highlighting the advantage of higher spatial resolution provided by MR VWI that allow depiction of vessel wall compared to conventional luminal studies.

Keywords: moyamoya, external carotid artery, magnetic resonance vessel wall imaging

DISCLOSURE

Conflict of interest: None

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