Limb-shaking TIA Related to Moyamoya Disease: Diagnosis with Magnetic Resonance Imaging and Magnetic Resonance Angiography

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Abstract-

Purpose: In a case with moyamoya disease, we found the magnetic resonance image and magnetic angiographic studies were helpful for the definite diagnosis of the disease.

Case report: A young adult presented limb-shaking transient ischemic attacks caused by moyamoya disease. The magnetic resonance angiography proved the steno-occlusive lesions in the major arteries of circle of Willis, and the magnetic resonance images demonstrated compensatively congested and dilated leptomeningeal vessels and lenticulostriate arteries with exhausted vasomotor elasticity.

Conclusion: In this case, without the aid of conventional angiography, the noninvasive magnetic resonance studies offered explicit imaging evidence to support the diagnosis and to illuminate the patient's clinical manifestation.

Key Words: transient ischemic attack, moyamoya disease, magnetic resonance image, magnetic resonance angiography

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INTRODUCTION

Moyamoya disease is characterized by progressive stenosis or occlusion of bilateral internal carotid artery bifurcation and abnormal vascular network at the base of the brain. On conventional cerebral angiography, filling of the basal vascular network and collaterals produces a cloudy image resembling a puff of smoke, which is called moyamoya vessels[1,2]. There are no clinical manifestations that are characteristic for moyamoya disease, although affected children most frequently present with recurrent ischemic events and older patients with intracerebral hemorrhage[3,4]. The pathological findings in moyamoya disease are distinct with segmental narrowing of major branches of the circle of Willis, and the microscopic features include thickening of intima and media, markedly wavy internal elastic lamina, proliferation of smooth muscle cells, and the absence of inflammatory changes[3,5-7].

Usually, the conventional angiography via invasive

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catheterization technique is prerequisite to establish the diagnosis of moyamoya disease. Recently, a few literatures discovered that magnetic-resonance image (MRI) and three-dimensional magnetic-resonance angiography (MRA) helped in diagnosing the occlusion of cerebral major vessels and identifying moyamoyoa vessels in cases with moyamoya disease. A recent revised guideline in 1995 also acknowledged these noninvasive methods substituting for invasive angiography to be helpful for the diagnosis of moyamoya disease, but in children only[3]. Here we present a young adult patient with moyamoya disease diagnosed with the aids of MRI and MRA.

CASE REPORT

A 31-year-old woman suffered from occasional numbness and clumsiness on the right hand for 1 year. Six months prior to admission, she started to have intermittent coarse shakings on the right hand and arm and briefs of right hand drop, which were occasionally accompanied with right facial myokymia and speech arrest. The involuntary shakings were mostly elicited when she was hyperventilated in anger or exercise, or used her right arm on continuous activities more than 1 minute.

She was healthy without history of central nervous system infection, cranial irradiation, vasculitis, or malignancy, and had no risk factors for atherosclerosis, hyperviscosity state, or cardiac source of emboli. There was no family history of early-onset vascular disease. She was a non-smoker, and used oral pills briefly and irregularly. Laboratory tests including hematological and coagulopathy screening, lipid profiles, C-reactive protein (CRP), antinuclear antibody, antiphospholipid antibody and cerebrospinal fluid studies were all normal.

Repeated electroencephalographies showed no epileptic discharge, but there were focal delta waves at the left frontal area during hyperventilation, which induced the aura of right hand shaking. Carotid ultrasonography disclosed decreased peak systolic flow velocity on the left proximal internal cerebral artery, and relatively high flow volume on bilateral vertebral arteries. Transcranial Doppler study also revealed reduced peak systolic velocity 36 cm/sec in left MCA and 64 cm/sec in right MCA, low pulsatile index in bilateral MCAs (0.39, 0.49 respectively), and high peak systolic velocity in basilar artery (132 cm/sec). Both vascular sonographic findings proved the insufficiency of anterior circulation with hemodynamic compensation from the posterior circulation.

MRI was performed with a 1.5T scanner (Genesis_Signa; GE Medical System, Milwaukee, U.S.A.), and MRA a 3D time-of-flight (TOF) sequence with spoiled gradient-recalled acquisition in the steady state. The coronal T2 imaging disclosed abnormal vascular signals in bilateral lenticulostrate perforators (Figure 1), and the fast fluid-attenuated inversion-recovery (FLAIR) revealed congested and dilated leptomeningeal vessels at bilateral cerebral hemispheres (Figure 2), indicating static flow in these vessels. MRA with Gadolinium contrast enhancement revealed significant stenotic-occlusive lesions in the proximal segments of bilateral MCAs, more severe in the left side, and bilateral anterior cerebral arteries (ACAs), with relatively flourishing flow in the branches of the posterior cerebral arteries (PCAs) (Figure 3). Poor run-off in the distal branches of left MCA was also noted. Further cerebral

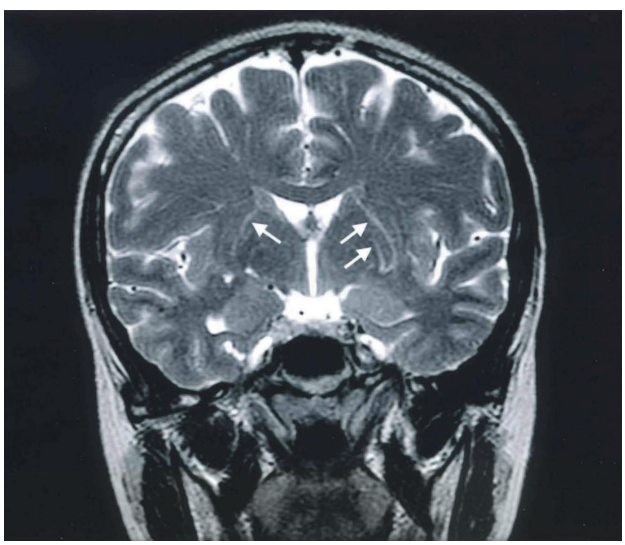

Figure 1. Coronal T2 fast spin echo (TR/TE: 4716/102) MR image reveals the course of perforating arteries in bilateral basal ganglia (arrows).

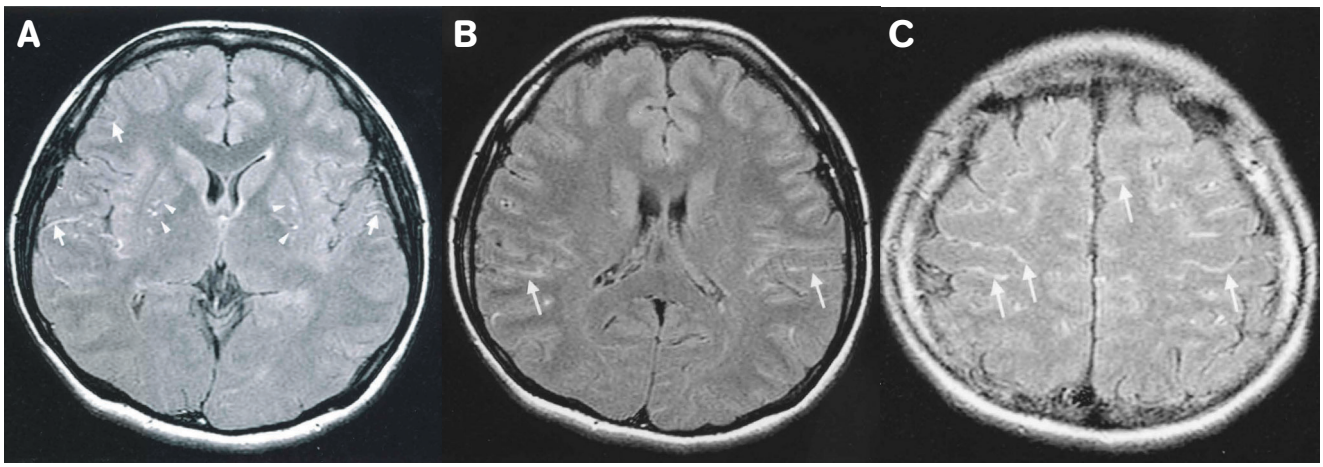

Figure 2. (A) Axial FLAIR (TR/TE/TI: 10,002/105/2,200) MR image reveals multiple areas of high signal intensity in leptomeninges (arrows) and in perforating arteries in basal ganglia (arrowheads). (B)(C) Axial FLAIR (10,000/105/2,200) MR images reveal multiple areas of high signal intensity in leptomeninges (arrows). Both b and c were interpreted as depicting the leptomeningeal ivy sign.

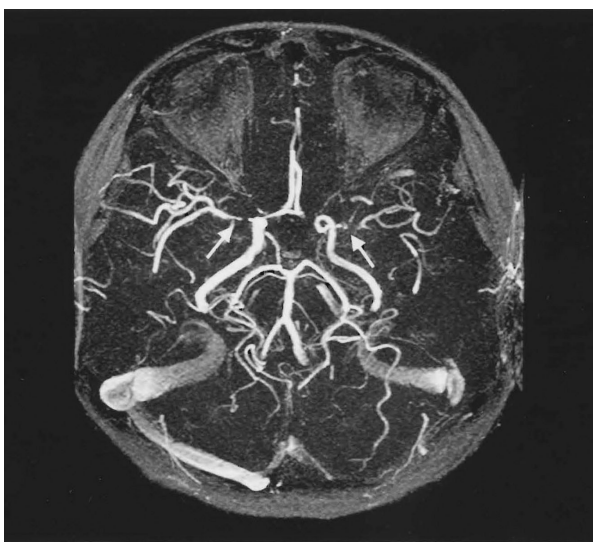

Figure 3. Maximum intensity projection image of MR angiogram reveals severe steno-occlusion of bilateral MCA stems (arrows).

angiography was not performed since her family strongly refused any invasive examination.

At first, she declined the offer of bypass surgery, and tried conservative treatment and avoided provoking factors such as strenuous exercise and emotional agitation. The shaking attacks on right hand got more frequent with daily attacks about 9 months later, and then she finally received bypass surgery on left superficial temporal artery-middle cerebral artery at another hospital in northern Taiwan, and the TIAs on right hand completely vanished after revascularization surgery. Unfortunately, she started to have limb-shaking TIAs on left hand about 4 months after surgery, and she was ready to take revascularization surgery on the other side soon.

DISCUSSION

In this case, the diagnosis of moyamoya disease was confirmed by the fact of severe steno-occlusion at the proximal segments of the MCAs and ACAs with no identified etiologies, such as atherosclerosis, autoimmune disorders, post-radiation state or meningitis. The limb-shaking, characterized by brief, repetitive jerking movements of the arm or leg, is an uncommon manifestation of transient cerebral ischemia attacks (TIA) and results from perfusion insufficiency and exhausted vasomotor reactivity on the affected hemisphere[8-11]. Orthostatic position change, prolonged upright posture, hyperventilation, or hypotension can trigger these involuntary movements. Mostly the limb-shaking TIAs were

reported to be related to extracranial or intracranial carotid atherosclerotic atherosclerotic stenosis[10,12,13], but few cases associated with moyoamoya disease were documented[7,14].

TIA is not a common clinical manifestation among adult patients with moyamoya disease. According to the report from research committee on Spontaneous Occlusion of the Circle of Willis, Ministry of Healthy and Welfare in Japan (MHWJ), hemorrhagic stroke is more prevalent in adult moyamoya cases, occurring in 66% with predominance in females, and cerebral infarction or TIA is less common[3,4,7]. Perfusion insufficiency was confirmed to be the main pathophysiologic etiology for these transient ischemic events or a progression of an ischemic deficit. The exhausted vasomotor reactivity in affected cerebral vessels was proved by hypercapnic test, and usually evidenced with maximal vasodilation of collateral vessels on images[14,15].

In order to improve the inadequate cerebral perfusion, collateral pathways develop gradually. These may include basal telangiectasia, transdural collaterals and leptomeningeal collaterals. There are reports exhibiting the unique MRI findings of these collaterals. For example, the "vault moyamoya" means the dilated leptomeningeal vessels at the cortical surfaces[6], and the "ivy sign" on MR FLAIR images illustrates the slow flow in engorged leptomeningeal vessels[16]. The significantly larger vessels along cortex evidenced on the MR images suggest that the extreme dilatation are compensated for cerebral hypoperfusion[17], and the vessels lose their vascular elasticity gradually. In order to compensate for the low perfusion, the dilated leptomeningeal vessels will draw more blood flow from posterior circulation or external carotid circulation[21]. In our case, MR images showed the congested and dilated deep perforators and leptomeningeal vessels clearly. Obviously, the dilated vessels meant the poor vascular reactivity, which explained why limb-shaking TIAs were provoked by hand activities when increasing demands of cerebral perfusion. The loss of vessel elasticity probably affected the prognosis of revascularization surgery, and acute cerebral hemorrhage or acute cerebral ischemia following procedure were reported[21,22].

MRA with contrast enhancement has been known of accuracy in assessment of steno-occlusive lesions of large vessels. The cerebral basal moyamoya vessels were not usually visualized on MRA, but the dilated leptomeningeal and transdural collateral vessels were explicated on MR images[18,19]. The MRA could be more helpful in pediatric cases, because the angiographic changes progressed more rapidly and prominently among childhood-onset moyamoya[20]. The sensitivity and specificity for diagnosis of moyamoya disease with MRI plus MRA were documented to be near 92% and 100% respectively as compared with conventional angiography in one lesion-by-the lesion analysis[18]. The value of MRI and MRA altogether in diagnosing moyamoya disease is demonstrated in our case, too. Although conventional angiography is still considered the diagnostic gold standard, this intrusive procedure actually carries a significant risk of ischemic complications in patients with moyamoya disease. A safer and easier substitution like MRI plus MRA appears to be a promising substitute.

In our adult patient with moyamoya disese, the MRA proved steno-occlusion in major arteries of circle of Willis. Although there were no moyamoya vessels shown on the MRA, the MR FLAIR images demonstrated the compensated dilated leptomeningeal vessels, which illuminated the gradual loss of vasoreactivity in the cerebral territory. Without the aid of angiography findings or perfusion study, this noninvasive MR image study proved to offer the clear imaging evidences and supported our clinical diagnosis.

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