More Precise Imaging Analysis and Diagnosis of Moyamoya Disease and Moyamoya Syndrome Using High-Resolution Magnetic Resonance Imaging

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■ BACKGROUND: **The diagnosis of moyamoya disease (MMD) is often uncertain. Moyamoya syndrome (MMS) is often misdiagnosed as MMD. High-resolution magnetic resonance imaging (HR-MRI) enables vessel wall assessment to obtain more precise diagnoses. The aim of this study was to determine the true etiologies of arterial steno-occlusion in patients with an angiographic diagnosis of MMD or MMS using HR-MRI.**

■ METHODS: **HR-MRI was performed in 21 adult patients with angiographically proven MMD or MMS. A definite diagnosis was based on the HR-MRI findings. The diagnoses made via the 2 different imaging technologies were compared, and significant findings were analyzed.**

■ RESULTS: **A total of 21 patients were enrolled, including 7 patients with angiographically proven MMD and 14 patients with angiographically proven MMS. Among the 7 patients with MMD, HR-MRI confirmed the diagnosis of MMD in 6; the remaining patient was considered to have atherosclerosis in the bilateral distal internal carotid arteries (ICAs) and the left middle cerebral artery. Among the 14 patients with MMS, HR-MRI confirmed MMD in 6 patients (including 2 patients with unilateral MMD), atherosclerosis in 5 patients (including 3 patients with bilateral atherosclerosis and 2 with unilateral atherosclerosis), arterial dissection of the left ICA in 1 patient, and MMD in the left cerebral hemisphere with atherosclerosis in the right hemisphere in 2 patients.**

■ CONCLUSIONS: **Differentiating MMD from MMS is difficult in certain situations, and HR-MRI may help provide a more in-depth understanding of MMD and MMS, thereby achieving a more reliable diagnosis.**

INTRODUCTION

Moyamoya disease (MMD) is a rare cerebrovascular occlusive disease characterized by progressive narrowing of the distal internal carotid arteries (ICAs) and the secondary development of small collateral vessels.[1,2] Although MMD was identified roughly 50 years ago, its etiology remains unclear.[3] Given the difficulty of correlating the disease with pathological findings, the diagnosis of MMD relies exclusively on imaging results. To diagnose MMD, atherosclerosis and other occlusive diseases that involve mainly the distal ICA or proximal middle cerebral artery (MCA) must be excluded.[4] Similar to MMD, these cerebrovascular occlusive diseases also can cause cerebral artery steno-occlusion combined with moyamoya vessels (MMV), a condition known as moyamoya syndrome (MMS).[5,6]

Conventional arterial imaging techniques, such as digital subtraction angiography (DSA) and magnetic resonance angiography, focus on the vessel lumen. Differentiating MMD from MMS is difficult in certain situations, especially in adult patients with atherosclerotic risk factors.[7-10] Thus, the clinical diagnosis of MMD or MMS based on DSA is unreliable in some cases. Some patients

Key words
- High-resolution magnetic resonance imaging
- Intracranial atherosclerosis
- Moyamoya disease
- Moyamoya syndrome

Abbreviations
ACA: Anterior cerebral artery
BA: Basilar artery
DSA: Digital subtraction angiography
HR-MRI: High-resolution magnetic resonance imaging
ICA: internal carotid artery
ICAD: Intracranial atherosclerotic disease
MCA: Middle cerebral artery
MMD: Moyamoya disease
MMS: Moyamoya syndrome
MMV: Moyamoya vessels
TE: Echo time
TR: Repetition time

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with MMS may be misdiagnosed when their imaging findings are similar to those associated with MMD, and some patients with MMD may be diagnosed with MMS owing to confounding factors.

High-resolution magnetic resonance imaging (HR-MRI) with high in-plane resolution allows for vessel wall assessment to obtain more precise diagnoses.[9,10] Although the MCA is a small and deep intracranial artery, recent studies have suggested that the MCA wall and lumen can be identified and characterized on HR-MRI with good interobserver reproducibility.[11,12] Studies using HR-MRI have demonstrated that the arterial wall of the stenotic area in MMD is associated with inward remodeling, smaller vessel outer diameter, concentric occlusive lesions, and a homogeneous signal intensity, whereas the arterial wall in intracranial atherosclerotic disease (ICAD) is more likely to present with outward remodeling, normal or greater than normal vessel outer diameter, eccentric occlusive lesions, and a heterogeneous signal intensity[10-17] (**Figure 1**). Because ICAD is common in adult patients, MMS caused by atherosclerosis can be confusing. Therefore, in the present study we focused mainly on patients with MMS with atherosclerotic risk factors. We sought to assess the true etiology of cerebral artery steno-occlusion in these patients, and to identify patients with MMS who were misdiagnosed as having MMD. To the best of our knowledge, this is the first report to analyze MMS by comparing HR-MRI and DSA findings.

METHODS

Patients

We prospectively enrolled patients who presented at our hospital's Department of Neurosurgery between September 2015 and May 2016 with suspected MMD. The study population comprised 7 patients with MMD and 14 patients with MMS diagnosed by DSA, according to the 2012 updated diagnostic criteria for MMD.[4] The patients with MMS had bilateral narrowing of the ICAs or MCA with moyamoya-type baseline collateral vessels, along with 1 or more atherosclerotic risk factors (eg, hypertension,

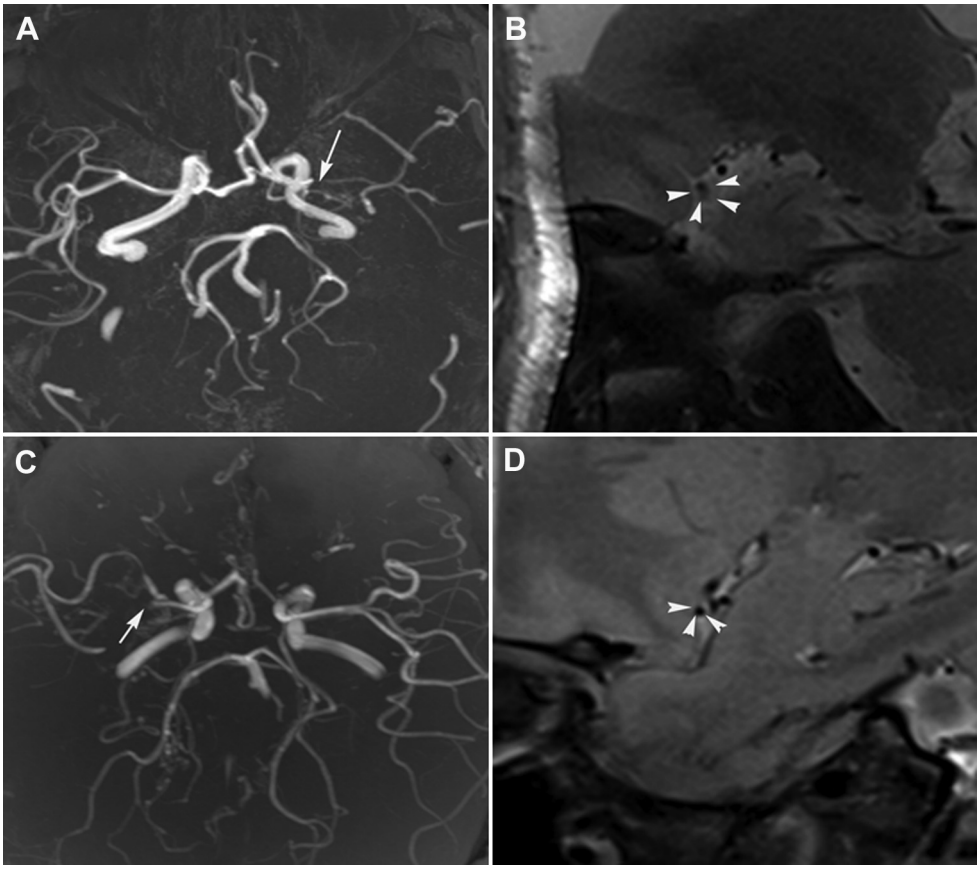

Figure 1. (**A** and **C**) Characteristic features of high-resolution magnetic resonance imaging (HR-MRI) in a patient with intracranial atherosclerotic disease (ICAD) (**A**) and a patient with moyamoya disease (MMD) (**C**). Both patients had severe stenosis of the right or left M1 segment (*arrows*). (**B**) HR-MRI image of a patient with ICAD showing the stenotic segment of the left M1 presenting with positive remodeling, normal outer vessel diameters, eccentric occlusive plaque, and heterogeneous signal intensity (*arrowheads*). (**D**) Inward remodeling, smaller outer vessel diameters, and concentric occlusive lesions in the stenotic segment of the right M1 in a patient with MMD.