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Moyamoya Phenomenon Secondary to Intracranial Atherosclerotic Disease: Diagnosis by 3T Magnetic Resonance Imaging

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Abstract

Moyamoya phenomenon occurs in response to an occlusive vasculopathy affecting the distal internal carotid artery and its proximal branches. The nature of the occlusive vasculopathy is unknown in most patients. We present a patient in whom 3T magnetic resonance imaging was used to examine the arterial wall at the site of occlusion in a patient with unilateral moyamoya phenomenon. Signal characteristics were consistent with atherosclerotic disease. 3T magnetic resonance imaging may be useful for distinguishing the underlying etiology of moyamoya phenomenon in some patients.

Keywords

Moyamoya; Atherosclerosis; Stroke; MRI

Background and Purpose

Moyamoya collaterals form in response to progressive narrowing or occlusion of the distal internal carotid artery (ICA) or its proximal branches. The underlying pathology may be variable and includes atherosclerotic disease [1,2]. A reliable method to distinguish atherosclerosis from other causes of occlusive vasculopathy would have considerable clinical value [3].

Magnetic Resonance (MR) methods for characterization of atherosclerotic plaques have been in development for several years [4,5]. These tools offer the potential to distinguish atherosclerotic disease from other causes of arterial narrowing or occlusion in patients with moyamoya phenomenon. We report a patient with moyamoya phenomenon in whom 3T-MR imaging was used to identify elements of atherosclerotic plaque at the site of the occlusion.

Clinical Presentation

A 46-year-old gentleman with past medical history significant for dyslipidemia, hypertension, and coronary artery disease presented with complaints of a stepwise decline in cognitive function and numbness in his right hand and face. The patient was moderately disabled at time

of presentation to us in clinic. He was dependent on family for daily affairs and was living with family members. The majority of the dependency related to cognitive dysfunction with poor short term memory (1/3 objects at 5 minutes) and mild language difficulties. His presenting modified Rankin Score (mRS) was 3. His symptoms had progressed over a 2-year period despite aggressive medical therapy including an antiplatelet regimen and, at the time of presentation to our institution, anticoagulation using coumadin. Neurological examination revealed a significant reduction in cognitive speed, abstract reasoning, and short-term memory. He also had marked decrease in light touch and pinprick sensation in the right upper extremity. His exam was otherwise unremarkable.

Standard MRI of the brain performed 1-month prior revealed multiple small areas of infarction in the centrum semiovale and corona radiata (Figure 1). Cerebral angiography performed 12 months prior to his presentation at our institution (Figure 2a) demonstrated high-grade stenosis at the origins of the left A1 and M1 segments and hypertrophy of lenticulostriate vessels consistent with moyamoya collaterals. The right ICA and middle cerebral artery (MCA) were normal. The right A1 segment was small with no flow across the anterior communicating artery to the left hemisphere. We repeated his angiogram at the time of his evaluation at our institution for consideration of possible angioplasty or stenting. This revealed a progressive stenooclusive process with interval occlusion of the left M1 segment (Figure 2b) and persistent left moyamoya collaterals.

Methods

We enrolled the patient in an Institutional Review Board approved pilot study designed to investigate the feasibility of arterial wall imaging in moyamoya disease with 3T-MR. The patient provided informed consent. All images were acquired at a 3T Siemens Allegra head-only system (Siemens Medical Solutions, Malvern, PA) with a head coil. The patient was scanned first with a 3-dimensional (3D) time-of-flight (TOF) sequence to cover the area of interest (TR/TE = 43 ms/4.7 ms, flip angle = 18°, field of view (FOV) = 175 mm × 215 mm, data matrix = 208 × 512, interpolated slice thickness = 1 mm, total three slab with 30 slices each slab, and total acquisition time = 7 min 48 sec).

T1-weighted, T2-weighted, and proton-density-weighted images were collected in 12 slices that were perpendicular to the axis of the distal internal carotid artery. For T1-weighted acquisition, a segmented turbo-spin-echo sequence (TSE) was used with double saturation pulses to saturate inflow blood signals from arteries and veins (TR/TE = 600 ms / 9.8 ms, segmented number = 7, slice thickness = 3 mm, no slice gap, fat saturation, FOV = 144 mm × 160 mm, interpolated data matrix = 564 × 640, image averaged number = 2, and acquisition time = 2 min 10 sec). The proton-density and T2-weighted images were collected in one scan with the same TSE imaging sequence aforementioned, but with TR/TE of 3000 ms/9.9ms and 3000ms/59 ms. The location of diseased left distal ICA is shown in Figure 3a. Magnified images (Figure 3b, proton-density-weighting and Figure 3c, T2-weighting) demonstrated a narrowed left artery with thickened intima, with the distal right ICA for comparison. The hypointense area close to the lumen was consistent with lipid deposit (Arrow, Figure 3c).

Owing to the complete occlusion of the M1 segment, the patient underwent surgical revascularization including both a left superficial temporal artery (STA) to MCA bypass utilizing the posterior branch of the STA as well as a left encephaloduroarteriosynangiosis procedure (EDAS) utilizing the anterior branch of the STA. He had an uneventful hospital course and was discharged to home on postoperative day three. At one year post-surgery, the patient had demonstrated a marked improvement in cognitive function including normal short term memory (3/3 objects at 5 min) and no language difficulties. He was living independently and was looking to return to work. This qualifies as an mRS=1. One year follow-up angiography

(not shown) revealed excellent revascularization of the left MCA territory via direct STA-MCA collaterals, as well as newly developed indirect pial collateralization from the EDAS.

Discussion

This report illustrates the potential of MR imaging to identify patients with moyamoya phenomenon secondary to atherosclerotic steno-occlusion. Moyamoya phenomenon is likely a secondary response to many different causes of arterial stenosis and occlusion [1]. Previous reports have recognized that atherosclerosis may be one of a number of causative factors for this phenomenon [6].

Application of special imaging techniques, faster coils, and high-resolution scanners have significantly improved MR resolution, contrast, and signal-to-noise ratio such that visualization of plaque components is now feasible [5]. Recent work suggests that intraluminal areas with decreased signal intensity on T2-weighted images (Figure 3c) may indicate lipid infiltration [4,10,11]. These results are consistent with severe intracranial atherosclerosis and, in conjunction with the patient's clinical history, strongly suggest that the underlying cause of this patient's moyamoya syndrome was severe atherosclerotic vasculopathy. It must be noted, however, that pathologic proof of atherosclerosis in this patient is lacking, and that it is possible that other pathological processes may cause signal loss as well.

In addition, the application of this technique to smaller vessels, such as the proximal middle and anterior cerebral arteries, will require further investigation. The visualization of these branches was poor in this subject, possibly owing to their small size as well as the angle of the imaging slab relative to the vessel axis. The slab was positioned orthogonal to the distal internal carotid artery to maximize the flow signal within the residual lumen.

Narrowing of the major intracranial arteries occurs secondary to a number of different vasculopathies, including inflammatory conditions, sickle cell anemia, Long-standing basilar meningitis, prolonged/repeated x-ray exposure, Down's syndrome, and atherosclerosis [12]. Angiography only allows the diagnosis of luminal narrowing which alone may not be able to definitively isolate the cause of narrowing. Moreover, the *in-vivo* diagnosis of these vasculopathies is often difficult or impossible. For example, the diagnosis of primary angiitis of the central nervous system requires a brain biopsy [13]. Identification of atherosclerosis would be of great clinical importance, particularly for the ability to exclude other causes of arterial narrowing and guiding therapeutic decision-making. This is particularly relevant given the development of dedicated intracranial stents for the treatment of atherosclerotic stenosis [14] and the recent observation of a high-risk re-stenosis group, consisting of young women with supraclinoid stenosis [15]. These patients may have an underlying pathology other than atherosclerosis. MR imaging may be able to separate these two groups. It is also possible that 3T-MR imaging of blood vessel walls might also allow the *in-vivo* identification of other pathologic processes as well.

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Figure 1.

Figure 2.

Figure 3.

Figure 4.