The use of vessel wall imaging to detect a de novo microaneurysm on the periventricular anastomoses in moyamoya disease: illustrative case

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BACKGROUND Moyamoya disease (MMD) is linked to the formation of intracranial aneurysms. The authors recently observed an effective use of magnetic resonance vessel wall imaging (MR-VWI) to detect de novo unruptured MMD-associated microaneurysms.

OBSERVATIONS The authors describe a 57-year-old female who was diagnosed with MMD 6 years ago after suffering a left putaminal hemorrhage. MR-VWI revealed point-like enhancement in the right posterior paraventricular region during the annual follow-up. On the T2-weighted image, this lesion was surrounded by high intensity. Angiography revealed a microaneurysm in the periventricular anastomosis. Right combined revascularization surgery was performed to prevent future hemorrhagic events. Another de novo circumferential enhanced lesion on MR-VWI appeared in the left posterior periventricular region 3 months after surgery. Angiography revealed that the enhanced lesion was a de novo microaneurysm on the periventricular anastomosis. The left combined revascularization surgery went well. The bilateral microaneurysms vanished on follow-up angiography.

LESSONS Unruptured MMD-associated microaneurysms on the periventricular anastomosis can be detected using MR-VWI. Revascularization surgery can eliminate microaneurysms by reducing hemodynamic stress on the periventricular anastomosis.

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KEYWORDS vessel wall imaging; moyamoya disease; microaneurysm; revascularization surgery

Hemorrhagic events in moyamoya disease (MMD) are caused by the disruption of inadequate collateral anastomoses or rupture of aneurysms associated with MMD.1,2 MMD-associated aneurysms are typically classified as microaneurysms on collateral anastomoses, which are synonymous with peripheral aneurysms, and saccular aneurysms on major arteries, such as the internal carotid arteries.3 MMD-related microaneurysms on collateral anastomoses are more likely to rupture than saccular aneurysms on major arteries.2

Magnetic resonance vessel wall imaging (MR-VWI) is used to assess the status of intracranial aneurysms. We previously reported that in unruptured intracranial saccular aneurysms, those with circumferential enhancement along the aneurysm wall (CEAW) on MR-VWI were associated with an unstable state that could lead to aneurysm rupture.4 We present a case in which MR-VWI was useful in detecting an MMD-associated microaneurysm before rupture.

Illustrative Case

Examination and history
A 51-year-old female had symptomatic left putaminal hemorrhage (Fig. 1A). She had a history of vasospastic angina and hyperlipidemia. She was diagnosed with MMD using magnetic resonance angiography (Fig. 1B) and treated conservatively. After rehabilitation, she recovered from aphasia and right hemiparesis and was discharged with no neurological deficits.
MMD.5,6 The prevalence of MMD-associated aneurysms varied according to the reports. Meyeong et al.7 found that 147 MMD-associated aneurysms in 118 of 2,728 adult MMD cases (4.3%) in a single institute. The clinical manifestation of MMD-associated aneurysms differed depending on where they were located. Unruptured MMD-associated microaneurysms along collateral anastomoses had a higher rate of growth or rupture than saccular aneurysm on major arteries.3 This was due to the vulnerability of microaneurysms, which are classified as pseudoaneurysms.8 Rhim et al.9 reported 77 adult MMD cases with hemorrhagic onset, of which 19 cases (24.7%) were caused by the rupture of microaneurysms. These findings could be linked to the fact that periventricular collaterals are a risk factor for de novo hemorrhage.10 Thus, surgical indication for cases with MMD-associated microaneurysms on the periventricular collateral is appropriate.7 Revascularization surgery may be an effective treatment for obliterating these microaneurysms by improving cerebral circulation and reducing chronic hemodynamic stress.2,11 When MMD-associated microaneurysms rupture, the treatment of choice is emergent surgical clipping or endovascular embolization.12–14

First Surgery

VWI with a single-slab three-dimensional (3D) T1-weighted fast spin echo sequence at the age of 57 revealed point-like enhancement at the right posterior paraventricular region (Fig. 2A). On a T2-weighted image, this enhancement was accompanied by surrounding high intensity (Fig. 2B). Digital subtraction angiography (DSA) revealed a microaneurysm on the periventricular anastomosis originating from the right posterior cerebral artery (Fig. 2C), which corresponded to the enhanced lesion on VWI. Despite the fact that she had no ischemic symptoms, we performed appropriate revascularization surgery to prevent future hemorrhagic events caused by the microaneurysm. An anastomosis of the right superior temporal artery (STA) to the middle cerebral artery (MCA) with encephaloduro-osynangiosis (EDMS) was performed. The postoperative period was uneventful. The enhanced lesion in the right hemisphere was not identified 3 months after surgery, and the aneurysm was completely gone on DSA (Fig. 2D).

Second Surgery

Three months after surgery, a ring-shaped enhancement with high T2 intensity surrounding it appeared in the left posterior paraventricular region (Fig. 3A, B). DSA revealed a de novo microaneurysm arising from the left posterior cerebral artery along the periventricular anastomosis, which did not exist prior to the first surgery (Fig. 3C and D). The ring-shaped enhancement on VWI was thought to show CEAW based on the fusion images with MRI and DSA (Fig. 3E). We performed left revascularization surgery due to the rupture risk of the de novo microaneurysm with CEAW. Left STA-MCA anastomosis with EDMS was performed. The postoperative period was uneventful. The enhanced lesion was not found 7 months after the second surgery, and the aneurysm completely disappeared on DSA (Fig. 3F and G). Single-photon emission computed tomography with N-isopropyl-p-iodoamphetamine showed improved cerebral blood flow in both hemispheres significantly (not shown).

Discussion

MMD With an Intracranial Aneurysm

Chronic hemodynamic stress may cause aneurysms formation in MMD.5,6 The prevalence of MMD-associated aneurysms varied according to the reports. Meyeong et al.7 found that 147 MMD-associated aneurysms in 118 of 2,728 adult MMD cases (4.3%) in a single institute. The clinical manifestation of MMD-associated aneurysms differed depending on where they were located. Unruptured MMD-associated microaneurysms along collateral anastomoses had a higher rate of growth or rupture than saccular aneurysm on major arteries.3 This was due to the vulnerability of microaneurysms, which are classified as pseudoaneurysms.8 Rhim et al.9 reported 77 adult MMD cases with hemorrhagic onset, of which 19 cases (24.7%) were caused by the rupture of microaneurysms. These findings could be linked to the fact that periventricular collaterals are a risk factor for de novo hemorrhage.10 Thus, surgical indication for cases with MMD-associated microaneurysms on the periventricular collateral is appropriate.7 Revascularization surgery may be an effective treatment for obliterating these microaneurysms by improving cerebral circulation and reducing chronic hemodynamic stress.2,11 When MMD-associated microaneurysms rupture, the treatment of choice is emergent surgical clipping or endovascular embolization.12–14

Observations

The unruptured MMD-associated microaneurysm was identified by MR-VWI in this case. The use of MR-VWI to assess the status of intracranial aneurysms has recently been reported in the literature. CEAW is seen on MR-VWI in ruptured saccular aneurysms.15 On the other hand, even in the unruptured saccular aneurysms, evolving aneurysms and aneurysms with daughter sac formation can exhibit CEAW.4,16 Unruptured aneurysms with CEAW are known to be at high risk of rupture, regardless of size. In the quantitative study, the degree of contrast ratio in unstable unruptured aneurysms was significantly higher than that in stable unruptured aneurysms.4 As a result, CEAW may serve as a biomarker for their instability.17 CEAW is also found in intracranial arterial dissection, where eccentric wall thickening is increased.18 The degree of contrast ratio varied depending on the conditions. In comparison to asymptomatic intracranial arterial dissection, acute symptomatic

FIG. 1. A: MR imaging of a left intracerebral hematoma at the time of initial presentation. B: MR angiography revealed steno-occlusive changes at the terminal portion of both internal carotid arteries, indicating MMD as the underlying pathology.

FIG. 2. A: VWI demonstrating the presence of a point-like enhancement in the right posterior paraventricular region (arrow). B: T2-weighted image of the surrounding high-intensity lesion (arrow). C: Right internal carotid angiography demonstrating a microaneurysm on the collateral circulation (arrowhead). D: The microaneurysm disappeared after revascularization surgery.

FIG. 3. A: T2-weighted image demonstrating the presence of a high-intensity lesion (arrow). B: Digital subtraction angiography demonstrating a de novo microaneurysm (arrowhead). C: Right internal carotid angiography demonstrating a microaneurysm (arrowhead). D: The microaneurysm disappeared after revascularization surgery.
microaneurysms along the developed collateral anastomoses in this intracranial arterial dissection, such as hemorrhage or headache, demonstrated a high level of CEAW. One of the underlying mechanisms for CEAW in saccular and dissecting aneurysms may be inflammation within the aneurysmal wall.

Strong hemodynamic stress was thought to be the cause of the microaneurysms along the developed collateral anastomoses in this case, which were successfully obliterated after revascularization surgery. On the contrary, right revascularization surgery has been linked to the formation of de novo microaneurysms in the left hemisphere. Ma et al. described the progression of impaired hemodynamics in the nonrevascularized hemisphere after unilateral revascularization. This could be because increased blood flow through the bypass affects compensation from other original collateral circulation. In this case, left revascularization surgery helped to eliminate the de novo microaneurysm and improve hemodynamic status in the left hemisphere.

In this case, we must debate the pattern of enhancement of MMD-associated microaneurysms. The right hemisphere microaneurysm showed point-like enhancement rather than circumferential enhancement. This pattern of improvement was comparable to the spot sign within the intracerebral hematoma, which was known as contrast extravasation and stagnation within the hematoma due to active hemorrhage. It is unclear whether this is due to partial aneurysmal wall enhancement or flow stagnation within the microaneurysm. The microaneurysm in the left hemisphere, on the other hand, showed circumferential enhancement, which could indicate the aneurysm wall itself, which we call CEAW. Though the resolution of the MRI was a limitation of this study, MR-VWI was useful in suggesting the presence of a de novo microaneurysm in a less invasive manner than cerebral angiography.

Lessons
Unruptured MMD-associated microaneurysms on the periventricular anastomosis can be detected using MR-VWI. Revascularization surgery can eliminate microaneurysms by reducing hemodynamic stress on the periventricular anastomosis.

References


**Disclosures**
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

**Author Contributions**
Conception and design: Endo, Ninomiya. Acquisition of data: Endo, Ninomiya. Analysis and interpretation of data: Endo, Ninomiya, Tashiro. Drafting the article: Ninomiya. Critically revising the article: Endo, Ninomiya, Tashiro. Reviewed submitted version of manuscript: Endo, Ninomiya, Tashiro. Kanoke. Approved the final version of the manuscript on behalf of all authors: Endo. Study supervision: Endo, Ninomiya, Tominaga.

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