Visualization of Periventricular Collaterals in Moyamoya Disease with Flow-sensitive Black-blood Magnetic Resonance Angiography: Preliminary Experience

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Abstract

Fragile abnormal collaterals in moyamoya disease, known as “moyamoya vessels,” have rarely been defined. While flow-sensitive black-blood magnetic resonance angiography (FSBB-MRA) is a promising technique for visualizing perforating arteries, as of this writing no other reports exist regarding its application to moyamoya disease. Six adults with moyamoya disease underwent FSBB-MRA. It depicted abnormal collaterals as extended lenticulostriate, thalamic perforating, or choroidal arteries, which were all connected to the medullary or insular artery in the periventricular area and supplied the cortex. This preliminary case series illustrates the potential for FSBB-MRA to reveal abnormal moyamoya vessels, which could be reasonably defined as periventricular collaterals.

Key words: moyamoya disease, periventricular anastomosis, black-blood magnetic resonance angiography

Introduction

Intracranial hemorrhage is a devastating symptom of moyamoya disease.1) Fragile abnormal vascular collaterals, known as “moyamoya vessels,” are suspected as a source of bleeding.1,2) Although such collaterals are generally assumed to arise from dilated lenticulostriate arteries,3,4) the angiographical extension of dilated thalamic perforators or choroidal arteries is also known to involve bleeding.5) A pioneering study implied that all these types of collaterals arising from the lenticulostriate, thalamic perforating, and choroidal arteries clustered around the periventricular subependymal area to connect to the medullary arteries and were frequently associated with cerebral microbleeds.6) However, morphological details of the connection of such collaterals have not been sufficiently documented. Furthermore, because numerous overlapping vessels can obscure the view, angiography often fails to reveal these details.

Flow-sensitive black-blood magnetic resonance angiography (FSBB-MRA) is a recently introduced noninvasive black-blood imaging technique for visualizing perforating arteries.7,8) High-resolution 3-tesla FSBB-MRA can reveal tiny parenchymal arteries as well as cisternal and ventricular arteries in the coronal view of the brain. As of this writing, no previous report has addressed the use of FSBB-MRA in visualization of abnormal collateral vessels in moyamoya disease. Noninvasive detection of such collaterals might gain clinical significance in light of risk estimates of bleeding in moyamoya disease. In the present preliminary case series, this innovative imaging technique was applied to six patients with moyamoya disease to facilitate visualization and analysis of the morphological characteristics of periventricular collaterals.

Materials and Methods

I. Patients

Six adult patients (male 4, female 2) with moyamoya disease were included in the present study (Table 1). The age of these patients ranged from 34 years to 44 years. The mode of manifestation was intracranial hemorrhage in five patients and transient ischemic attack in one patient. All patients underwent magnetic resonance (MR) imaging including
FSBB-MRA, routine clinical 3-tesla MR imaging including susceptibility-weighted imaging (SWI), and conventional cerebral angiography during the same admission period. All patients provided written informed consent to the FSBB-MRA.

II. Imaging technique
A 3-tesla research MR scanner (Vantage; Toshiba Medical Systems Corporation, Otawara, Tochigi) with a 32-channel head coil was used to obtain FSBB images. These images were scanned as coronal sections with the following parameters: repetition time (TR)/echo time (TE), 35/13 ms; flip angle, 15°; acquisition matrix size, 384 × 384; and field of view (FOV), 192 × 192 mm in 1 axial 3D slab of 80 sections (0.8 mm thickness); and a parallel imaging factor of 2. The imaging field extended from the anterior horn to the atrium. A motion-probing gradient of b = 0.3 s/mm² was applied to dephase arterial blood flow in three directions. Total scan time was 8 m 31 s. In addition to source images with 0.8 mm thickness, minimum-intensity projection images were also generated as 2.5-mm thick and 10-mm thick slabs of overlapping volumes. Both minimum-intensity projection images and source images were assessed by a neuroradiologist and a neurosurgeon.

During the same admission period, all patients underwent MR imaging including FSBB-MRA, routine clinical 3-tesla MR imaging including susceptibility-weighted imaging (SWI) to detect the evidence of bleeding, and conventional cerebral angiography.

III. Analysis
Both a neurosurgeon (Takeshi Funaki) and a neuroradiologist (Yasutaka Fushimi) carefully compared the results of the arterial-phase angiography and FSBB-MRA for each patient. Periventricular anastomosis, the finding of interest in the present study, was defined as that between the perforating and medullary arteries or between the choroidal and medullary arteries, which was located around the periventricular area. Any topographical relationship between periventricular anastomoses and SWI-visible lesions was assessed on a workstation integrated into the image-archiving and communication system.

Results
Periventricular anastomoses were observed in FSBB-MRA images obtained from all six patients (Table 1). The morphologies of the periventricular anastomoses revealed in the FSBB-MRA images exactly coincided with those revealed in the arterial phase of angiography, confirming that FSBB-MRA truly depicted arteries. A total of 19 periventricular anastomoses were identified with FSBB-MRA. An SWI-visible lesion was identified at the exact sites of 11 anastomoses (57.9%).

Representative cases
Case 1: A 43-year-old female with moyamoya disease suffered from left temporal lobe hemorrhage. Left internal carotid artery angiography (Fig. 1a, b) revealed the medullary arteries derived from a lenticulostriate artery, suggesting an anastomosis between the lenticulostriate and medullary arteries around the periventricular area. FSBB-MRA more clearly revealed an anastomosis between these arteries at the lateral corner of the frontal horn of the ventricle with exact correspondence to angiography (Fig. 1c). Note that the medullary arteries have the largest caliber in the cortical area and smallest in their periventricular portion, indicating that the arteries originally arose from the cortical arteries. Blood flow was directed “ventriculofugally” toward

<table>
<thead>
<tr>
<th>Case</th>
<th>Age/Sex</th>
<th>Manifestation mode</th>
<th>Location of hemorrhage</th>
<th>Initial symptoms</th>
<th>Suzuki stage, R/L</th>
<th>Number of periventricular anastomoses (R/L)</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>43F</td>
<td>ICH</td>
<td>left temporal lobe</td>
<td>headache, speech disturbance</td>
<td>1/3</td>
<td>2 (1/1)</td>
</tr>
<tr>
<td>2</td>
<td>37M</td>
<td>ICH with IVH</td>
<td>right thalamus</td>
<td>headache, nausea</td>
<td>5/5</td>
<td>5 (3/2)</td>
</tr>
<tr>
<td>3</td>
<td>47M</td>
<td>ICH</td>
<td>left temporal lobe</td>
<td>hemiparesis, motor aphasia</td>
<td>4/4</td>
<td>3 (2/1)</td>
</tr>
<tr>
<td>4</td>
<td>37M</td>
<td>ICH</td>
<td>left insula and lateral part of thalamus</td>
<td>hemiparesis</td>
<td>0/3</td>
<td>3 (0/3)</td>
</tr>
<tr>
<td>5</td>
<td>34M</td>
<td>IVH</td>
<td>lateral ventricle</td>
<td>consciousness disturbance</td>
<td>5/5</td>
<td>5 (2/3)</td>
</tr>
<tr>
<td>6</td>
<td>44F</td>
<td>TIA</td>
<td>–</td>
<td>transient motor weakness</td>
<td>4/4</td>
<td>1 (1/0)</td>
</tr>
</tbody>
</table>

ICH: intracerebral hemorrhage, IVH: intraventricular hemorrhage, TIA: transient ischemic attack.
the cortical area in the medullary arteries; it thus travels opposite to the original direction of flow in the medullary artery. In this patient, FSBB-MRA of the contralateral side revealed a very similar finding.

**Case 2:** A 37-year-old male with moyamoya disease suffered from right thalamic hemorrhage. Meticulous reading of left carotid artery angiography might identify a tortuous perforating artery originating from the thalamotuberal artery and connecting to the medullary arteries, but numerous overlapping vessels obscure the view (Fig. 2a). FSBB-MRA more clearly revealed the thalamotuberal artery, which coursed around the periventricular area of the third ventricle and then abnormally extended laterally beyond the thalamus and connected to the medullary artery (Fig. 2b). Note that a microbleed was revealed at the inflexion point of the collaterals, the site supposed to be the first anastomotic site, located beneath the ependymal layer of the third ventricle. In this patient, FSBB-MRA of the contralateral side revealed a very similar finding, where the evidence of thalamic hemorrhage was observed.

**Case 3:** A 47-year-old male with moyamoya disease suffered from left temporal lobe hemorrhage. Right vertebral artery angiography showed the probable thalamogeniculate artery connecting to the insular artery and subsequently to the middle cerebral artery, revealing a somewhat arbitrary spatial relationship (Fig. 3a). FSBB-MRA more clearly demonstrated the anastomosis between the thalamogeniculate artery and the insular artery, a type of medullary artery originally derived from the middle cerebral artery (Fig. 3b). The anastomosis was located at the inferolateral margin of the thalamus near the inferior horn of the lateral ventricle. SWI revealed a microbleed at the exact point of the anastomosis (Fig. 3c). In this patient, FSBB-MRA of the contralateral side revealed a similar finding, where the evidence of temporal lobe hemorrhage was observed.

**Case 4:** A 37-year-old male with moyamoya disease suffered from hemorrhage extending through the lateral part of the thalamus and insula in the left...
hemisphere. Left internal carotid artery angiography showed an abnormal extension of the dilated choroidal artery (Fig. 4a, b). FSBB-MRA more clearly revealed anastomosis between the choroidal artery and the medullary artery beneath the lateral wall of the atrium of the lateral ventricle (Fig. 4c), with exact correspondence to the anterior-posterior view from the angiography. The evidence of a microbleed can be observed at the exact point of the anastomosis (Fig. 4c, d). In this patient, the anastomosis between the thalamogeniculate and insular arteries was also observed in the lateral part of the left thalamus, where the evidence of hematoma was observed.

Discussion

I. Periventricular anastomosis as a concept defining fragile collateral networks in moyamoya disease

In the present case series, all patients have a type of anastomosis between the perforating and medullary arteries or between the choroidal and medullary arteries. These types of anastomoses probably serve as a collateral to the cortex and compensate for the decrease in cerebral blood flow attributable to occlusion of the internal carotid artery. Such collaterals, although the subject of limited interest, have been denoted variously in previous reports as “anastomosis between the perforating branch and medullary artery,”9) “abnormal vessel network/medullary artery anastomosis,”4) or “lenticulostriate-medullary artery anastomosis.”3) Most of these collaterals, however, have barely been identified through meticulous angiographic observation and thus have rarely been

Fig. 3 Periventricular anastomosis originating in the thalamogeniculate artery (Case 3). a: Anterior-posterior view of vertebral artery angiography showing the thalamogeniculate artery (arrows), which connects to the insular artery (arrowhead) and subsequently to the middle cerebral artery (double arrowhead), revealing a somewhat arbitrary spatial relationship. b: Flow-sensitive black-blood magnetic resonance angiography more clearly demonstrating the anastomosis (asterisk) between the thalamogeniculate artery (arrows) and the insular artery (arrowhead), which is located at the temporal stem. c: Susceptibility-weighted imaging revealing a microbleed at the exact site of the anastomosis (double arrow).

Fig. 4 Periventricular anastomosis originating in the anterior choroidal artery (Case 4). a, b: Anterior-posterior (a) and lateral (b) views of left internal carotid artery angiography showing dilated anterior choroidal artery (arrows) connecting to the medullary artery (arrowhead). c: Flow-sensitive black-blood magnetic resonance angiography revealing anastomosis (asterisk) between the choroidal artery (arrows) and the medullary artery (arrowheads) located beneath the lateral wall of the atrium. d: Susceptibility-weighted imaging showing the microbleed (double arrow) coincident with periventricular anastomosis.
systemized. Furthermore, these reports focused only on perforating arteries such as lenticulostriate arteries as sources of collaterals. Morioka et al. stressed that abnormal extension or branching of the dilated choroidal artery also served as important collaterals possibly associated with bleeding. Although they did not clearly define the abnormal branches from the choroidal artery, we assumed from observation suggests that the medullary artery represents such branches.

All collaterals described in the present cases share one feature: all anastomosis sites were located in the periventricular area, that is, at the lateral corner of the anterior body of the lateral ventricle (Fig. 1), beneath the ependyma of the third ventricle (Fig. 2), superior to the inferior horn of the lateral ventricle (Fig. 3), and beneath the lateral wall of the atrium of the lateral ventricle (Fig. 4). It might thus be reasonable to classify all these types of anastomoses under one identifier as, say, periventricular anastomosis (Fig. 5).

These anastomoses cluster in the periventricular area possibly because of the presence of the subependymal artery, an anatomically hypothesized artery beneath the ependyma and originally described as the ventriculofugal (or centrifugal) artery. The subependymal artery might intervene between the perforating and medullary arteries or between the choroidal and medullary arteries in a specific pathological condition such as moyamoya disease. As shown in Fig. 5, periventricular anastomoses can be reasonably classified into three types: lenticulostriate, thalamic, and choroidal. These classifications differ only slightly from those proposed by Kazumata et al. The thalamic type could be subclassified into medial and lateral types according to the location of the anastomosis. The medial thalamic type could also include the connection between the perforating artery and the medial posterior choroidal artery in the roof of the third ventricle, which was not observed in the present series. It might also be acceptable to include in the choroidal classification the possible connection between the medial posterior choroidal artery and pericallosal artery through the corpus callosum, a condition not observed in the present series.

The distribution of periventricular anastomoses corresponds closely to common bleeding sites in moyamoya disease; that is, the basal ganglia, thalamus, temporal stem, and periventricular areas of the entire lateral and third ventricles. This evidence could support the hypothesis that periventricular anastomosis is a surrogate marker for bleeding, a consideration that should be tested in further studies.

II. Clinical importance of FSBB-MRA

FSBB-MRA is a high-resolution black-blood imaging method adequate for visualizing small perforating arteries in the general population and in patients with lacunar infarction. The imaging methods can more sensitively visualize the perforating arteries than time-of-flight MRA. In FSBB-MRA, the signal from rapidly flowing blood in the arteries is attenuated through the application of a very weak motion-probing gradient for signal dephasing, while the signal from slow-moving components, such as the flow in the veins, is much less affected. FSBB-MRA might have two benefits for detecting periventricular anastomoses. First, minimum-intensity projection coronal images with adequate slab thickness noninvasively facilitate visualization of periventricular anastomosis without any effect from numerous vessels overlapping and obscuring the view. Partial volume effect is avoid-
able by simultaneous reading of thin slice source images. Second, unlike conventional angiography, black-blood MRA can provide information on not only the tiny arteries but also anatomy of the parenchymal structure. Visualization of both anatomies seems essential to the evaluation of periventricular anastomoses. Although a recent study illustrated that 3-tesla time-of-flight MRA could also noninvasively depict moyamoya vessels,14) FSBB-MRA might provide better contrast than time-of-flight MRA for depicting both the collaterals and parenchyma.

In conclusion, this preliminary case series illustrates the potential of FSBB-MRA to reveal abnormal collaterals in moyamoya disease, or moyamoya vessels, characterized as those arising from the lenticulostriate, thalamic perforating, or choroidal arteries and connecting to the medial end of the medullary or insular artery in the periventricular area. The concept of periventricular anastomosis, an alternative definition of moyamoya vessels, might facilitate future optimal grading and classification of moyamoya vessels. The detection of periventricular anastomoses with FSBB-MRA could generate risk estimates of bleeding in moyamoya disease, and larger studies are required.

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Conflicts of Interest Disclosure

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

References


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