Moyamoya Disease: Comparison of Assessment with 3.0-T MR Angiography and MR Imaging versus Conventional Angiography

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**Purpose**

1. Definition: Moyamoya disease (MMD) is a chronic cerebrovascular disorder that is characterized by progressive occlusion of distal part of internal carotid artery (ICA) and proximal part of middle and anterior cerebral arteries which are main branches within the circle of Willis[1].

2. Radiologic criteria for the diagnosis in MMD:

   (1) Stenosis or occlusion at the terminal portion of the ICA and the proximal portion of ACA and MCA.

   (2) Abnormal vascular network seen in the arterial phase in the vicinity of the arterial occlusion.

   (3) Bilateral involvement [2].

3. There is no previous investigation comparing 3.0-T magnetic resonance angiography (MRA) and magnetic resonance (MR) imaging with conventional angiography in moyamoya disease. We comparatively assessed the findings of those two methods of examinations in moyamoya disease.

**Methods and Materials**

**Patients**

From December 2006 to June 2009, 13 patients (4 males and 9 females; mean age / age range (years), 36 / 21- 54) with MMD were examined by both MRA (3.0-T) and conventional angiography at our institution (Saga University Hospital). Diagnosis was made on the basis of the findings by using conventional angiography. No patients underwent intracranial revascularization surgery. The mean interval time between the two types of examinations was 24 days. Patients were initially suspected of having MMD due to transient ischemic attack in 7 patients, cerebral infarction in 2 patients, intracranial
hemorrhage in 3 patients, and asymptomatic state in 1 patient. Of the 13 patients, 2 had a family history of MMD.

**Imaging Examinations Protocol**

MR imaging was performed using a 3.0-T MR unit (MAGNETOM Trio, A Tim System, Siemens AG, Erlangen, Germany). MRA was performed using the following parameters: repetition time (TR)/echo time (TE) (ms) = 22/3.1; flip angle (degree) = 18; field of view (FOV) (mm) = 200*166-180; matrix = 384*320-346; pixel spacing (mm) = 0.5*0.5; slice thickness/interslice gap (mm) = 0.6-1.0/0; number of slices = 102-150. Maximum-intensity projection reconstruction (MIP) images of TOF-MRA were generated. Fluid attenuated inversion recovery (FLAIR) imaging was performed using a fast inversion recovery sequence with parameters as follows: TR/TE/inversion time (TI) (ms) =9000/63-86/2500; flip angle (degree) = 120-150; FOV=220*178-213; matrix = 384*312-372; pixel spacing (mm) = 0.6*0.6; slice thickness/interslice gap (mm) = 6.0/1.2; number of slices =20.

Head angiographic examinations were performed using a clinical angiography system (Axiom Artis TA; Siemens AG, Erlangen, Germany). Angiographies of the bilateral common carotid arteries, internal carotid arteries, external carotid arteries, and either side of the vertebral artery were performed at least once for each patient.

**Imaging Evaluations Method**

Imaging findings were categorized according to the following grading systems. In assessment of MRA source images, re-sliced MRA source images (slice thickness/interslice gap (mm) =1.0/0 ) were generated from original images by using a workstation (Virtual Place Raijin version 3.1, Aze, Ltd, Tokyo, Japan) were used because various slice thicknesses of MRA source image were adopted among patients and might affect the evaluation. All images were observed on a computer viewer system (ViewR version1.09.15, Yokogawa Electric Corporation, Tokyo, Japan) with a 54cm class color LCD monitor (Radioforce R22, EIZO NANAO CORPORATION, Ishikawa, Japan).

**A) Conventional angiographic (CA) stage**

The severity of disease in the involved intracranial vessels in each patient was evaluated on conventional angiography, and the disease stage was determined according to Suzuki’s grading system (Table 1 on page 17). Stage 0: No stenosis of distal intracranial ICA; Stage 1: stenosis of distal intracranial ICA; Stage 2: basal moyamoya vessels can be detected based on Stage 1; Stage 3: discontinuity of MCA or ACA can be detected based on Stage 2; Stage 4: discontinuity or disappearance of posterior communicating artery (PCoA) can be detected combined with either or none of ACA/MCA disappearance based on Stage 3; Stage 5: disappearance of both ACA and MCA can be detected based on Stage 4; Stage 6: on the basis of Stage 5, no depiction of basal moyamoya vessels.
**B) MRA stage (score & grade)**

The steno-occlusive severity of intracranial vessels on MRA was evaluated according to Houkin’s grading system [3], and we modified it as follows (Figure 1 on page 6).

i) **Internal Carotid Artery (ICA) score:**

According to Houkin's grading system, it is difficult to distinguish "normal" from "stenosis" and "discontinuity" from "invisible". The ICA score was modified based on the appearances of middle cerebral artery (MCA), posterior communicating artery (PCoA) and anterior choroidal artery (AChA) continuity, as follows. Point 0: From the ICA tip (distal part to ophthalmic artery) to distal end of the horizontal segment of MCA (M1) is continuous. Point 1: From the ICA tip to distal end of M1 is discontinuous, but PCoA is continuous with posterior cerebral artery. Point 2: PCoA is discontinuous, but AChA is continuous with the inferior horn of the lateral ventricle. Point 3: AChA discontinuity on the basis of point 2.

ii) **Middle Cerebral Artery (MCA) score:**

As it is difficult to distinguish "discontinuity" from "invisible" using Houkin's grading system, we decided to add the visualization of an insular segment of MCA (M2) in compliance with the severity of the steno-occlusive change of M1. Point 0: No steno-occlusive change of M1. Point 1: Stenosis of M1 (the caliber of the distal part is dominant to that of the proximal part). Point 2: Discontinuity or disappearance of M1, but M2 can be visualized. Point 3: Regardless of M1, no visualization of M2 branches.

iii) **Anterior Cerebral Artery (ACA) score:**

It is difficult to distinguish a "normal infracallosal segment of ACA (A2)" from an "A2 signal decrease or loss" using Houkin's grading system. Meanwhile, the anterior falx artery (AFA) is one of the important collateral arteries that is frequently developed in conjunction with the progression of the ACA stenosis [4]. Accordingly, the appearances of AFA and A2 were adopted. Point 0: Signal intensity of A2 dominant or equal to AFA. Point 1: Signal intensity of the AFA dominant to A2. Point 2: Regardless of AFA, no visualization of A2 or A2 has the same signal intensity as the brain parenchyma.

iv) **Posterior Cerebral Artery (PCA) score:**

It is difficult to distinguish "normal P2" from "P2 signal decrease or loss" using Houkin’s grading system. We added visualization of the parietooccipital artery (POcA) because POcA is a primary distal branch originating from the ambient segment of PCA (P2). The PCA scores were evaluated depending on the visibility of P2 and POcA. Point 0: From P2 to POcA is continuous. Point 1: Signal shows discontinuity of P2 but POcA can be visualized. Point 2: regardless of P2, no visualization of POcA.
All scores were determined based on MRA source images using any window level (WL) and window width (WW) values. "Discontinuity", "disappearance" and "no visualization" were defined to mean that there was no vascular structure or a vascular structure with the same signal intensity as the brain parenchyma with any WL and WW. MIP images were also referentially used. The MRA scores were calculated from the summation of the ICA, ACA, MCA, and PCA scores, which ranged from 0 to 10 (ICA+MCA+ACA+PCA=3+3+2+2). MRA grading was defined according to Houkin's [3] system as follows: MRA score from 0 to 1= MRA grade 1; MRA score from 2 to 4= MRA grade 2; MRA score from 5 to 7= MRA grade 3; MRA score from 8 to 10= MRA grade 4.

**C) Other MR findings of MRA grading**

i) **Moyamoya vessel score** *(Figure 2 on page 8)*

The moyamoya vessel score was originally arranged ranging from 0 to 5 according to the regions where the collateral vessels were seen with any values of WL and WW on MRA source images. Those regions were the basal ganglia, anterior communicating artery (ACoA), MCA-ICA tip, PCoA-PCA, and basilar tip areas, where moyamoya vessels are frequently seen. Each region was given a score of 1 or 0, and the highest possible moyamoya vessel score was 5.

ii) **Ivy sign score**

It is well known that the "ivy sign" on FLAIR images is the result of the slow-flowing engorged pail convexity vessels and thickened arachnoids membrane which can be detected in MMD [5, 6]. Yoon’s 3 degrees of "absent", "equivocal" or "present" were adopted for evaluating the "ivy sign" [7]. As "equivocal" might be difficult to distinguish from "present", the following complementary definitions were assigned. Point 0: "absent" = the absence of ivy sign *(Figure 3A on page 9)*. Point 1: "equivocal" = ivy patterns with the same signal intensity as the brain parenchyma were dominantly observed (iso-dominant) *(Figure 3B on page 10)*. Point 2: "present" = high-intense ivy patterns were dominantly observed (high-dominant) *(Figure 3C on page )*.

iii) **CVA lesion**

Cerebrovascular attack (CVA) lesions were divided into small (less than 1 cm in maximal size), medium (1-3 cm in maximal size), and large lesions (over 3 cm in maximal size) according to the criteria proposed in the previous report [8].

**Statistical Analysis**

Relationships between Suzuki scores vs. MRA scores, MRA grading, ivy sign scores, and moyamoya vessel scores on every cerebral hemisphere in 13 patients (26 sides) were evaluated using Spearman's signed-rank test. Differences in Suzuki scores between
cases with or without CVA lesions of small, medium, and large CVA lesions were also analyzed. All analyses adopted a significance level of 0.05.

Images for this section:
**Fig. 1:** Modified MRA scores. Pictures of normal brain arteries and ICA, MCA, ACA, and PCA scores are listed. ICA point 1: from ICA tip to distal end of M1 is discontinuous, but PCoA is continuous. ICA point 2: PCoA is discontinuous, but AChA is continuous. ICA point 3: AChA discontinuity on the basis of point 2. MCA point 1: stenosis of M1 (caliber of the distal part is dominant to that of the proximal part). MCA point 2: discontinuity or disappearance of M1, but M2 can be visualized. MCA point 3: regardless of M1, no depiction of M2 branches or the signal intensity of M2 branches is the same as that of the parenchyma. ACA point 1: signal intensity of the AFA dominant to A2. ACA point 2: regardless of AFA, no depiction of A2 or A2 has the same signal intensity as the brain parenchyma. PCA point 1: signal shows discontinuity of P2 but POcA can be visualized. PCA point 2: regardless of P2, no visualization of POcA or POcA with the same signal intensity as the brain parenchyma. The MCA score = ICA + MCA + ACA + PCA (range from 0 to 10).
Fig. 2: Moyamoya vessel score ranges from 0 to 5 based on the five regions including the basal ganglia, ACoA, MCA-ICA tip, PCoA-PCA, and BA tip areas where the collateral vessels were frequently seen on MRA source images.
**Fig. 3**: Fig. 3A: MMD in a 36-year-old male. Transverse unenhanced FLAIR image shows no ivy patterns in leptomeninges (depict as "absent").
Fig. 4: Fig. 3B: MMD in a 44-year-old male. Transverse unenhanced FLAIR image shows iso-intense ivy patterns (arrows) in leptomeninges.
**Fig. 5**: Fig.5A: A 32-year-old female with MMD. The cerebral angiography revealed stenosis of the right internal carotid arteries associated with basal moyamoya vessels; MCA and ACA were also detected to be discontinuous.
Fig. 6: Fig.5B: The left side of the same patient. Stenosis of the right internal carotid arteries was associated with basal moyamoya vessels; MCA and ACA were also detected to be discontinuous.
Fig. 7: MRA images showed MCA discontinuity bilaterally (arrowheads), but bilateral PCoAs (double-headed arrows) were found to be continuous (ICA score =1, bilateral); bilateral PCAs were normal (arrows, PCA score =0)
Fig. 8: Fig 5D: Right M1 discontinuity with M2 branches (arrows, MCA score =2, right.) and a left M1 discontinuity without M2 branches (MCA score =3, left.) were visualized.
Fig. 9: Fig.5E: Signal intensity of the AFA (white arrow) was dominant to A2 (black arrow) (ACA score =1, bilateral). The MRA score of the right side was 4 (MRA grade 2), while that of the left side was 5 (MRA grade 3).

<table>
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<tr>
<th>SUZUKI STAGE</th>
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<td>Basal moyamoya vessels</td>
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Fig. 10: Table 1. Suzuki stage (from I to VI). Suzuki stage 1: stenosis of distal intracranial ICA; Stage 2: basal moyamoya vessels can be detected based on Stage 1; Stage 3: discontinuity of MCA or ACA can be detected based on Stage 2; Stage 4: discontinuity or disappearance of PCoA can be detected combined with either or none of ACA/MCA disappearance based on Stage 3; Stage 5: disappearance of both ACA and MCA can be detected based on Stage 4; Stage 6: on the basis of Stage 5, no depiction of basal moyamoya vessels.
Results

A good correlation was revealed between the conventional angiographic stage and the modified MRA score (rs=0.821, p<0.01) (Figure 4A on page 18), and MRA grading (rs=0.809, p<0.01) (Figure 4B on page 19).

Moyamoya vessel scores evaluated on MRA source images showed a significant correlation with Suzuki's CA stages (rs=0.667, p<0.01) (Figure 4C on page 20).

There was no significant correlation between ivy sign scores and CA stages and no significant differences in CA stages between cases with or without CVA lesions in any size (Figure 4D on page 21, E on page 22).

Figure 5(A-E) demonstrates a representative case of MMD and shows how to evaluate the MRA score. The patient was on Suzuki stage III in both sides (Figure 5A on page , B on page ), MRA images showed MCA discontinuity bilaterally (arrowheads), but bilateral PCoAs (double-headed arrows) were found to be continuous (ICA score =1, bilateral); bilateral PCAs were normal (arrows, PCA score =0) (Figure 5C on page ). Another slice of the source images showed a right M1 discontinuity with M2 branches visualized (arrows, MCA score =2, right.) and a left M1 discontinuity without M2 branches (MCA score =3, left.) (Figure 5D on page ). Signal intensity of the AFA (white arrow) was dominant to A2 (black arrow) (ACA score =1, bilateral). The MRA score of the right side was 4 (MRA grade 2), while that of the left side was 5 (MRA grade 3) (Figure 5E on page ). The ivy sign score of this patient was 2 bilaterally (Figure 3C on page 23). This patient had several small-sized CVA lesions bilaterally and a medium-sized CVA lesion in the left frontal lobe.

Images for this section:
Fig. 1: Fig. 4A: Suzuki stage correlated well with MRA score ($rs=0.821$, p
Fig. 2: Fig. 4B: Suzuki stage correlated well with MRA grade ($rs=0.809$, $p$
**Fig. 3:** Fig. 4C: Suzuki stage correlated well with moyamoya vessel score (rs=0.667, p...
**Fig. 4:** Fig. 4D: No significant correlation between Suzuki stage and ivy sign score.
**Fig. 5:** Fig. 4E: No significant differences in Suzuki stage between the presence and absence of small, medium and large CVA lesions.
Fig. 6: Fig. 3C: MMD in a 32-year-old female (the same case as in Figure 5). Transverse unenhanced FLAIR image reveals multiple areas of high signal intensity (arrows) in leptomeninges.
Conclusion

AChA and PCoA were adopted to evaluate the steno-occlusive changes of ICA in the modified MRA grading method because these two vessels were the main branches of the distal ICA. We considered those 2 branches to be relatively important for patients with MMD because the dilations of the AChA and PCoA were indicators with good sensitivity and specificity for predicting hemorrhagic events in MMD [9], and the presence of PCoA was one of the key ways to distinguish between Suzuki stages III and IV. Besides, we defined point 0 of ICA score as the continuity from distal ICA to distal end of M1 because it was sometimes difficult to find the division between ICA and MCA in cases of A1 disappearance.

As it was difficult to distinguish "discontinuity" from "invisible" with regard to the steno-occlusive changes of the MCA in Houkin's grading system, we added a visualization of M2 to evaluate the severity of the steno-occlusive change of M1. Because discontinuity represents the interruption of a vessel with visualization of its distal part, the absence of visualization of the distal part (M2) of the MCA indicates the severity of the disease.

As A1 (horizontal segment of ACA) is often hypoplastic on the basis of its reciprocal relation to the other side of ACA, we evaluated A2 in the present study according to Houkin's [3]. Furthermore, we added the AFA factor as a comparison because the ethmoid moyamoya and its branches spread into the forebrain, thus serving as a potential source for the supply of arterial blood, which might indicate the severity of the disease [4]. Also, because AFA is one of the collateral vessels which run parallel with the ACA, it can be clearly and easily identified on MRA due to its enlargement to provide additional blood flow in patients with MMD. In accordance with these facts, we adopted it as a comparative factor to evaluate the anterior cerebral circulation.

We evaluated the PCA score for the same reason we evaluated the MCA score. The quadrigeminal segment of PCA (P3) and the cortical segment of PCA (P4) were determined to describe the discontinuity or disappearance of P2. POcA and the calcarine artery are distal branches of P3 and P4, which are important collateral arteries for supplying blood from the posterior to the anterior circulation. Their stenoses demonstrate the severity of the restriction of PCA circulation. As POcA was more easily seen on MRA compared with the calcarine artery, POcA was evaluated as an indicator of stenotic severity in order to evaluate the posterior circulation in MMD before treatment in this study.

In this study, we evaluated the other MRI findings including ivy sign score, moyamoya vessel score and CVA score and analyzed their relationships with CA stage. Leptomeningeal high signal intensity on FLAIR images was defined as continuous linear or punctate high signal intensity along the cortical sulci and subarachnoid space, which can reflect cerebral flow in the same way as the "ivy sign" detected in MMD [9,10,11]. As a recent finding, unilateral hemispheric ivy proliferation correlated highly with the
existence of an ipsilateral decreased cerebrovascular reserve (CVR) associated with the development of leptomeningeal collaterals in patients with MMD [6]. In order to investigate the further meaning of "ivy sign" compared with CA, we used Yoon's three degree's grading method which was modified by using the cerebrospinal fluid intensity (a signal void intensity) and the parenchymal signal intensity as references for the purpose of more definitive evaluation. However, the "ivy sign" were found to have no relationship with Suzuki's stages. This might be acceptable because Suzuki's angiographic staging does not necessarily relate to the cerebral blood flow or CVR [3, 12].

The moyamoya vessel score was excluded from the MRA grading system following Houkin's because the development of basal moyamoya vessels is not linearly correlated with Suzuki's angiographic stages of this disease [13,14]. Still, we evaluated the moyamoya vessel score independently in comparison with Suzuki's stage and found a good correlation. Most of our cases were in Suzuki stages II to V, in which the amount of the moyamoya vessels can reflect the severity of the disease [1, 4,14].

CVA lesions were failed to find a significant correlation with Suzuki's stages. This may be suggesting that Suzuki's angiographic staging does not always relate to the cerebral blood flow or ischemic changes [3].

This study has several limitations. Firstly, the number of cases in this study was restricted because we chose patients who had undergone both 3.0-T MRA and conventional angiography in a preoperative state. Secondly, there was no patient with Suzuki's stage VI. It might be much rare for preoperative patients with advanced MMD to get a CA study because nowadays the revascularization surgery has been performed to patients before the disease progresses to the final stage. Thirdly, we compared between MRA scores and Suzuki stages of the MMD patients. PCA factor was not included in Suzuki's CA stage while PCA evaluation was included in our modified MRA score following Houkin's grading system. This may be a limitation of the study design considering that there are no other preferable CA grading methods for MMD which includes the degree of PCA steno-occlusive severity. Fourthly, we used modified staging systems for MRA and ivy sign score, and original staging for moyamoya vessel score. Validation studies for those grading systems should be performed with the sufficiently larger number of patients. Lastly, we cannot deny the possibility of overestimation of steno-occlusion on MRA evaluation, a phenomenon that has been previously reported [13,15].

However, all of the cases in this study had not undergone surgery, which made our evaluation of the vascular findings more objective. Our results demonstrate that the modified Houkin's MRA grading system on 3.0-T MR may be used as one of the vascular assessments of MMD.

1. MRA on 3.0-T MR could be used as one of the vascular assessments of MMD rather than conventional angiographic evaluation because of its noninvasive nature and visual clarity.
2. Moyamoya vessel findings on 3.0-T MR may reflect one aspect of Suzuki's grading system.

3. Only with Suzuki's grading systems, we cannot explain the causes of the "ivy sign" or CVA lesions.

4. Our study is the first to compare conventional angiography with 3.0-T MRA in MMD and may give the available information for evaluating MMD on 3.0-T MR.

References


**Personal Information**