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Assessment of Moyamoya Disease With 3.0-T Magnetic Resonance Angiography and Magnetic Resonance Imaging Versus Conventional Angiography

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Abstract

3.0-T magnetic resonance (MR) angiography and MR imaging were compared with conventional angiography for the evaluation of moyamoya disease in 13 preoperative patients (26 hemispheres) with moyamoya disease (4 males and 9 females aged 21–54 years). The correlation between MR angiography scores determined by modified Houkin's grading system (MRA score) and conventional angiography stages determined by Suzuki's grading system (CA stage) was analyzed. Other MR findings such as moyamoya vessel scores, ''ivy sign'' scores, and the presence of small, medium, and large cerebrovascular attack (CVA) lesions were compared with CA stages. MRA scores were significantly correlated with CA stages ($p < 0.01$). Moyamoya vessel scores correlated well with CA stages ($p < 0.01$). There was no significant correlation between ''ivy sign'' scores and CA stages, and no significant differences in CA stages with the presence and absence of CVA lesions of any size. 3.0-T MR angiography can be used as a vascular assessment in moyamoya disease with its priority of noninvasive nature and visual clarity compared with conventional angiography. The findings of 3.0-T MR angiography may reflect the stenoocclusive changes in moyamoya disease.

Key words: moyamoya disease, 3 tesla, magnetic resonance angiography, ivy sign, moyamoya vessel

Introduction

Moyamoya disease (MMD) is a chronic cerebrovascular disorder that is characterized by progressive occlusion of the distal part of the internal carotid artery (ICA) and the proximal part of the middle (MCAs) and anterior cerebral arteries (ACAs), which are the main branches within the circle of Willis. The term ''moyamoya'' in Japanese corresponds to the irregular vascular networks that look like a ''puff or spiral of smoke'' (cloud-like lenticulostriate and thalamostriate collaterals on angiography).[19] After the guidelines for diagnosing MMD with magnetic resonance (MR) imaging and MR angiography were published in 1997,[2] three-dimensional time-of-flight MR angiography has become widely accepted as a noninvasive diagnostic modality. The stenotic severity of intracranial vessels in MMD can be evaluated by 1.5-T MR angiography.[4,5,10,11,16]

The present study compared the use of 3.0-T MR angiography and MR imaging with conventional angiography for the identification of MMD.

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Materials and Methods

This study retrospectively evaluated 13 patients, 4 males and 9 females aged 21–54 years (mean 36 years) with MMD who were examined by both MR angiography (3.0 T) and conventional angiography

at our institution from December 2006 to June 2009. Diagnosis was based on the findings of conventional angiography. No patients underwent intracranial revascularization surgery. MR angiography and MR imaging were performed before or after conventional angiography within 1 month, and no clinical event occurred between the two examinations. Patients were initially suspected of 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. Two of the 13 patients had a family history of MMD.

MR imaging was performed using a 3.0-T unit (Magnetom Trio, A Tim System; Siemens AG, Erlangen, Germany). MR angiography was performed using the following parameters: repetition time (TR)/echo time (TE) 22/3.1 msec, flip angle 18 degrees, field of view (FOV) 200*166–180 mm, matrix 384*320–346, pixel spacing 0.5*0.5 mm, slice thickness/interslice gap 0.6–1.0/0 mm, number of slices 102–150, and no magnetization transfer contrast. Maximum-intensity projection reconstruction (MIP) images were generated. Fluid-attenuated inversion recovery (FLAIR) imaging was performed using a fast inversion recovery sequence with parameters as follows: TR/TE/inversion time 9000/63–86/2500 msec, flip angle 120–150 degrees, FOV 220*178–213 mm, matrix 384*312–372, pixel spacing 0.6*0.6 mm, slice thickness/interslice gap 6.0/1.2 mm, and number of slices 20.

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

All images were assessed on a computer viewer system (ViewR version 1.09.15; Yokogawa Electric Corporation, Tokyo) with a 54-cm color LCD monitor (Radioforce R22; Eizo Nanao Corp., Hakusan, Ishikawa). MR imaging and conventional angiography findings were interpreted by two observers (QJ with 2-year experience of radiology, TN with 7-year experience of neuroradiology) to establish a consensus. Imaging findings were categorized according to the following grading systems.

The severity of disease in the involved intracranial vessels was evaluated on conventional angiography, and the disease stage was determined according to Suzuki's grading system (CA stages I to VI)[19] (Table 1).

The steno-occlusive severity of intracranial vessels was evaluated on MR angiography according to Houkin's grading system[11] (MRA score) (Table 2). The related anatomical hallmarks were added to the definitions as Houkin's grading system presents some judgmental difficulties. All scores were mainly determined based on the MR angiography MIP images, but source images were also used referentially. Houkin's MR angiography grade (MRA grade) was defined as the sum of the ICA, ACA, MCA, and posterior cerebral artery (PCA) scores as follows: total MRA scores from 0 to 1, MRA grade 1; total MRA scores from 2 to 4, MRA grade 2; total MRA scores from 5 to 7, MRA grade 3; and total MRA scores from 8 to 10, MRA grade 4.

The moyamoya vessel score ranged from 0 to 5 according to the regions containing collateral vessels with any values of window level and window width on MR angiography source images (Fig. 1). The regions were the basal ganglia, anterior communicating artery, MCA-ICA tip, posterior communicating artery (PCoA)-PCA, and basilar tip areas, where moyamoya vessels are frequently seen.

Table 1 Suzuki's conventional angiography stage (CA stage)[19]

	CA stage					
	I	II	III	IV	V	VI
Stenosis of distal ICA	+	+	+	+	+	+
Basal moyamoya vessels	−	+	+	+	+	−
ACA or MCA discontinuity	−	−	+	+	+	+
PCoA discontinuity or disappearance	−	−	−	+	+	+
ACA disappearance	−	−	+/−	+/−	+	+
MCA disappearance	−	−	+/−	+/−	+	+

CA stage I, stenosis of distal intracranial internal carotid artery (ICA); stage II, basal moyamoya vessels can be detected based on stage I; stage III, discontinuity of middle cerebral artery (MCA) or anterior cerebral artery (ACA) can be detected based on stage II; stage IV, discontinuity or disappearance of posterior communicating artery (PCoA) can be detected combined with either or none of ACA/MCA disappearance based on stage III; stage V, disappearance of both ACA and MCA can be detected based on stage V; and stage VI, on the basis of stage V, no depiction of basal moyamoya vessels.

Table 2 Houkin's magnetic resonance angiography score (MRA score)[11]

	MRA score
i) ICA	
Normal	0
Stenosis of C_1	1
Discontinuity of C_1 signal	2
Invisible	3
ii) MCA	
Normal	0
Stenosis of M_1	1
Discontinuity of M_1 signal	2
Invisible	3
iii) ACA	
Normal A_2 and its distal	0
A_2 and its distal signal decrease or loss	1
Invisible	2
iv) PCA	
Normal P_2 and its distal	0
P_2 and its distal signal decrease or loss	1
Invisible	2
Total	0–10

i) Internal carotid artery (ICA) score: point 1, stenosis of posterior knee segment (C_1) of the distal ICA, or small caliber of C_1 compared to horizontal segment (M_1) of the middle cerebral artery (MCA); point 2, discontinuity of C_1 signal, or discontinuity of petrous segment (C_2) of ICA-insular segment (M_2) of MCA and C_2-infracallosal segment (A_2) of anterior cerebral artery (ACA); and point 3, invisible, or discontinuity of C_2-M_2, C_2-A_2, and posterior communicating artery. ii) MCA score: point 1, stenosis of M_1, or large caliber of the distal part compared to proximal part; point 2, discontinuity of M_1 signal, or C_2-M_2 and A_2-M_2 discontinuity; and point 3, invisible, or invisible M_2 branches. iii) ACA score: point 1, A_2 and its distal signal decrease or loss, or less signal intensity of A_2 compared to anterior falx artery; and point 2, invisible, or invisible A_2. iv) Posterior cerebral artery (PCA) score: point 1, ambient segment (P_2) of PCA and its distal signal decrease or loss, or P_2 discontinuity; and point 2, invisible, or invisible parietooccipital artery.

Each region was given a score of 1 or 0, and the highest possible moyamoya vessel score was 5.

The ''ivy sign'' on FLAIR images is the result of slow flow in the engorged pial convexity vessels and thickened arachnoid membrane which can be detected in MMD (Fig. 2).[13] Yoon's 3 degrees of ''absent,'' ''equivocal,'' or ''present'' were adopted for evaluating the ''ivy sign.''[22] As ''equivocal'' might be difficult to distinguish from ''present,'' the following complementary definitions were assigned. Point 0, ''absent'' = absence of ivy sign; point 1, ''equivocal'' = ivy patterns with the same signal intensity as the brain parenchyma were predominantly observed (iso-dominant); and point 2, ''present'' = high intensity ivy patterns were predominantly observed (high-dominant).

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 previously.[2]

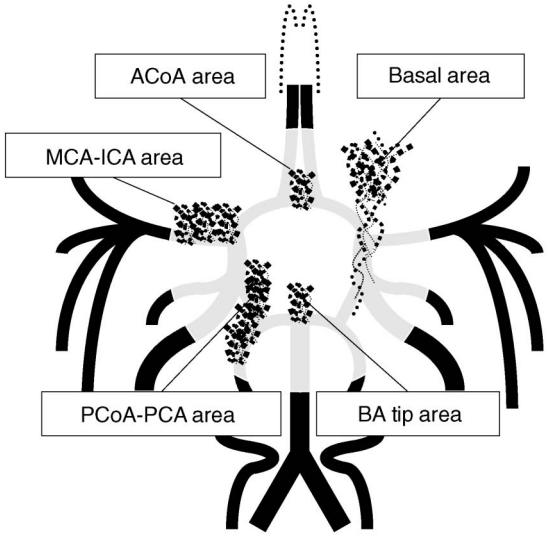

Fig. 1 Schematic illustration of moyamoya vessel score. Moyamoya vessel score ranges from 0 to 5 based on five regions including the basal ganglia, anterior communicating artery (ACoA), middle cerebral artery (MCA)-internal carotid artery (ICA) tip, posterior communicating artery (PCoA)-posterior cerebral artery (PCA), and basilar artery (BA) tip areas where collateral vessels are frequently seen in moyamoya disease.

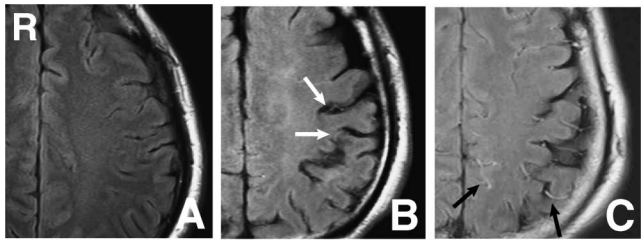

Fig. 2 A: Moyamoya disease in a 36-year-old male. Transverse fluid-attenuated inversion recovery (FLAIR) image showing no ivy patterns in leptomeninges (''absent''). B: Moyamoya disease in a 44-year-old male. Transverse FLAIR image showing iso-intense ivy patterns (*arrows*) in leptomeninges. C: Moyamoya disease in a 32-year-old female. Transverse FLAIR image revealing multiple areas of high signal intensity (*arrows*) in leptomeninges.

Relationships between CA stages and MRA scores, MRA grades, ivy sign scores, and moyamoya vessel scores for each cerebral hemisphere in the 13 patients (26 sides) were evaluated using Spearman's signed-rank test. Differences in CA stages between hemispheres with or without small, medium, and large CVA lesions were also analyzed. All analyses adopted a significance level of 0.05.

Results

A good correlation was revealed between the CA stage and the MRA score ($r_s = 0.876$, $p < 0.01$), and MRA grade ($r_s = 0.844$, $p < 0.01$), which indicated that patients with higher MRA score and grade had higher CA stage. Moyamoya vessel scores showed a significant correlation with CA stage ($r_s = 0.726$, $p < 0.01$), and most patients with higher moyamoya vessel scores (≥ 3) were in CA stage III or higher. 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 of any size, which might mean that CA stages were not necessarily correlated with the cerebral blood flow state since the ivy sign reflected the slow-flowing of the pial vessels and CVA lesions were caused by insufficient cerebral blood flow

Table 3 Summary of findings

Evaluation items and scores	CA stage			
	I	II	III	IV
MRA score				
1	1	1	0	0
2	1	0	1	0
3	0	1	0	0
4	0	0	4	0
5	0	0	4	1
6	0	0	5	2
7	0	0	0	2
8	0	0	0	3
MRA grade				
1 (MRA scores 0–1)	1	1	0	0
2 (MRA scores 2–4)	1	1	5	0
3 (MRA scores 5–7)	0	0	9	5
4 (MRA scores 8–10)	0	0	0	3
Moyamoya vessel score				
1	0	0	1	0
2	1	0	0	0
3	1	1	4	0
4	0	1	5	3
5	0	0	4	5
Ivy score				
0	1	0	1	0
1	1	0	3	3
2	0	2	10	5
Cerebrovascular attack lesions				
Smal				
(+)	2	2	11	6
(−)	0	0	3	2
Middle				
(+)	0	0	8	2
(−)	2	2	6	6
Large				
(+)	0	0	5	2
(−)	2	2	9	6

CA stage: Suzuki's conventional angiography stage, MRA score: Houkin's magnetic resonance angiography score, MRA grade: Houkin's magnetic resonance angiography grade.

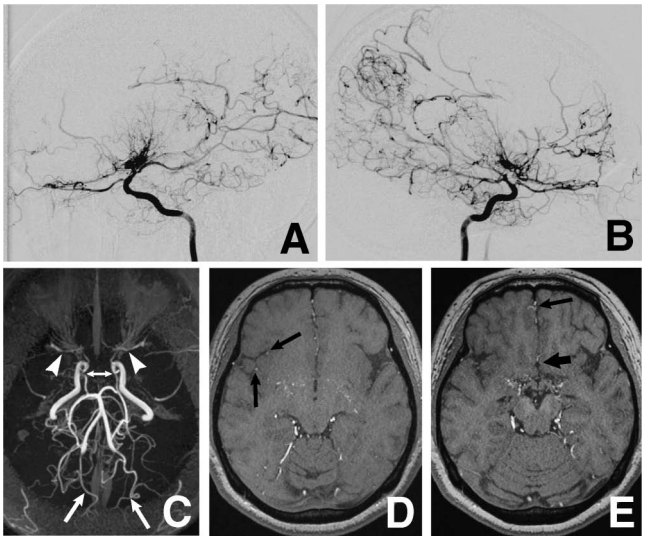

Fig. 3 Moyamoya disease in a 32-year-old female. A, B: Right (A) and left (B) carotid angiograms revealing stenosis of the bilateral internal carotid arteries (ICAs) associated with basal moyamoya vessels; middle cerebral artery (MCA) and anterior cerebral artery (ACA) were also discontinuous (Suzuki's conventional angiography stage III on both sides). C: Magnetic resonance (MR) angiography source image showing the bilateral MCAs were discontinuous (*arrowheads*), but the bilateral posterior communicating arteries (*double-headed arrows*) were continuous (ICA score 1, bilateral); and the bilateral posterior cerebral arteries (PCAs) were normal (*arrows*, PCA score 0). D, E: MR angiography source images showing a discontinuity of the right horizontal segment of the MCA (M_1) with branches of the insular segment of the MCA (M_2) (*arrows*, MCA score 2, right) and a discontinuity of the left M_1 without M_2 branches (MCA score 3, left) (D); and signal intensity of the anterior falx artery (*arrow*) dominant to A_2 (*thick arrow*) (ACA score 1, bilateral), indicating the Houkin's MR angiography score was 4 on the right side (MR angiography grade 2), and was 5 on the left (MR angiography grade 3) (E).

(Table 3).

Figure 3 demonstrates a representative case of MMD. The patient had CA stage III on both sides, and the MRA scores were 4 on the right and 5 on the left. Well-developed moyamoya vessels were detected in this patient except around the PCoA-PCA area (moyamoya vessel score 4). The ivy sign score of this patient was 2 bilaterally. This patient had several small CVA lesions bilaterally and a medium CVA lesion in the left frontal lobe.

Discussion

MR imaging and MR angiography are now widely accepted for the identification of MMD because of advantages of non-invasive examination, fewer complications, no requirement for contrast medium, and visual priority for detecting the secondary MR findings (cerebral infarction, white matter lesions, atrophy, and hemorrhage) compared to conventional angiography.[5,6,8,15,16] Depiction of small vessel segments was better with 3.0-T MR angiography than with 1.5-T MR angiography mainly because the signal-to-noise ratio and contrast-to-noise ratio at 3.0-T imaging was approximately twice that at 1.5-T MR imaging.[1,20,21,23] 3.0-T MR imaging with higher resolution could detect even smaller pathological vessels such as moyamoya vessels and can provide significantly greater angiographic details compared with 1.5-T MR imaging in MMD.[6] Medullary streaks, which were observed as linear structures crossing the centrum semiovale on high-resolution T_2-reversed imaging on 3.0-T MR imaging and appeared to represent the dilated medullary vessels in MMD, were significant in MMD patients with higher angiographical stage and cerebral hypoperfusion state.[7] Therefore, 3.0-T MR imaging has been proved to be superior to 1.5-T MR imaging in visualization ability. However, the radiological diagnosing usability has not been fully investigated by comparing 3.0-T MR angiography with conventional angiography in MMD. We comparatively assessed the findings of those two methods of examinations in preoperative patients in this study. We found a good correlation between the CA stage and the MRA score and grade, which supported the application of 3.0-T MR angiography in patients with MMD.

Other MR findings including ivy sign score, moyamoya vessel score, and CVA score were evaluated and their relationships with CA stage analyzed. 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 blood flow in the same way as the ''ivy sign'' detected in MMD.[3,14,22] Unilateral hemispheric ivy proliferation correlated highly with decreased ipsilateral cerebrovascular reserve (CVR) associated with the development of leptomeningeal collaterals in patients with MMD.[13] To investigate the further meaning of ''ivy sign'' compared with CA stage, we used Yoon's three degree grading method which was modified using the cerebrospinal fluid intensity (signal void intensity) and the parenchymal signal intensity as references for more definitive evaluation. However, the ''ivy sign'' was found to have no relationship with CA stage. This might be acceptable because CA stage does not necessarily relate to the cerebral blood flow or CVR.[9,11] The moyamoya vessel score was excluded from the MRA grade because the development of basal moyamoya vessels is not linearly correlated with CA stage of this disease.[10,18] Still, we found a good independent correlation between the moyamoya vessel score and CA stage. Most of our cases were in CA stages II to IV, in which the amount of the moyamoya vessels can reflect the severity of the disease.[17–19] CVA lesions had no significant correlation with CA stage, suggesting that CA stage does not always relate to the cerebral blood flow or ischemic changes.[11]

This study has several limitations. The number of cases was restricted because we chose patients who had undergone both 3.0-T MR angiography and conventional angiography during the preoperative period. Patients with advanced MMD rarely undergo preoperative cerebral angiography because nowadays revascularization surgery is performed before the disease progresses to the final stage. We compared moyamoya vessel score, ivy sign score, and CVA lesions to CA stage, but validation studies for those grading systems should be performed with a sufficiently large number of patients. Lastly, we cannot deny the possibility of overestimation of steno-occlusion on MR angiography, as previously reported.[10,12]

The present study demonstrated 3.0-T MR imaging has advantages in evaluating MMD. 3.0-T MR angiography could replace conventional angiography as the initial diagnostic tool as well as for follow up of MMD because of its noninvasive nature and visual priority for detecting secondary MR findings. Moyamoya vessel findings on 3.0-T MR angiography may reflect one aspect of CA stage. However, we cannot explain the causes of the ''ivy sign'' or CVA lesions with CA stage.

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