Cerebral Arterial Variations Associated with Moyamoya Disease Diagnosed by MR Angiography

AKIRA UCHINO¹, NAOKO SAITO¹, MASAHIRO TAKAHASHI², HIROKI KURITA², SHOICHIRO ISHIHARA²

¹Department of Diagnostic Radiology, Saitama Medical University International Medical Center; Hidaka, Saitama, Japan

²Department of Neurosurgery, Saitama Medical University International Medical Center; Saitama, Japan

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SUMMARY – *Moyamoya disease is a rare progressive cerebrovascular steno-occlusive disease associated with different variations of the cerebral arteries. We evaluated the types and prevalence of such variations among patients with moyamoya disease. In our institution during the past seven years, we diagnosed 72 patients (24 male, 48 female; aged 6 to 75 years, mean, 42 years) with moyamoya disease by magnetic resonance (MR) angiography using either a 3-Tesla or one of two 1.5-T imagers and a standard time-of-flight technique without contrast media. An experienced neuroradiologist retrospectively reviewed the images. There were 15 cerebral arterial variations in 13 of 72 patients with moyamoya disease (18.1%), including four basilar artery fenestrations, three ophthalmic arteries arising from the middle meningeal artery, two intracranial vertebral artery fenestrations, two persistent first cervical intersegmental arteries, two persistent trigeminal arteries, one extracranial origin of the posterior inferior cerebellar artery, and one persistent stapedia artery. Although our number of patients was small, moyamoya disease was frequently associated with variations of the cerebral arteries, especially fenestrations in the vertebrobasilar system and persistent trigeminal artery.*

Introduction

Moyamoya disease is a rare cerebrovascular steno-occlusive disease initially reported in Japan¹. Its characteristic angiographic feature is stenosis or occlusion of the terminal segments of the bilateral internal carotid arteries (ICAs) and the proximal segments of both the middle and anterior cerebral arteries. Because moyamoya disease is most prevalent in Japan and some patients have a family history of the disease, genetic factors were long suspected to play an important role in its etiology, and the genes responsible have been recently identified².

Several reports describe cases of moyamoya disease associated with a persistent trigeminal artery or its variants³⁻⁸ or other cerebral arterial variations, such as persistent hypoglossal artery⁹ and basilar artery fenestration¹⁰. To re-

assess the relationship between moyamoya disease and such variations of the cerebral arteries, we retrospectively reviewed magnetic resonance (MR) angiographic images of patients with moyamoya disease who were examined in our institution.

Patients and Methods

We conducted our study in accordance with the policies for exemption set by our internal institutional review board. Because the study was retrospective, we did not obtain informed consent from patients.

From April 1 2007 through March 31 2014, we diagnosed 72 patients (24 male, 48 female; aged 6 to 75 years, mean, 42 years) with moyamoya disease by MR angiography using either

Figure 1 Case 1. An 11-year-old boy with moyamoya disease. Anteroposterior projection of magnetic resonance angiography shows a basilar artery fenestration at its proximal segment (arrow).

Figure 2 Case 5. A 67-year-old woman with moyamoya disease. Caudocranial projection of magnetic resonance angiography shows bilateral ophthalmic arteries arising from the middle meningeal arteries (arrows).

a 3-Tesla or one of two 1.5-T imagers. Most patients were examined more than once. All MR angiograms were obtained using standard time-of-flight technique without contrast media.

The first author, an experienced neuroradiologist, retrospectively reviewed MR angiographic images using a picture archiving and communication system (PACS) (SYNAPSE, Fujifilm Medical, Tokyo, Japan), paying special attention to detect variations in cerebral arteries. In patients who underwent serial examinations, the most recently obtained images were reviewed. Thus, 60 images were obtained by a 3-T scanner, and the remaining 12 were acquired using a 1.5-T imager.

Results

Table 1 summarizes results. There were 15 cerebral arterial variations in 13 of 72 patients with moyamoya disease (18.1%), including four basilar artery fenestrations (5.6%, Figure 1), three ophthalmic arteries arising from the middle meningeal artery (2.1% per artery, Figure 2), two intracranial vertebral artery fenestrations (2.8%, Figure 3), two persistent first cervical intersegmental arteries (2.8%, Figure 3),

two persistent trigeminal arteries (2.8%, Figure 4), one extracranial origin of the posterior inferior cerebellar artery, and one persistent stapedial artery (Figure 5A,B). Ten of the 48 female patients (20.8%) and three of the 24 male patients (12.5%) demonstrated cerebral arterial variations.

Discussion

Moyamoya disease is frequently associated with variations of the cerebral arteries, especially with persistent trigeminal artery or its variants³⁻⁸. Kwak and Kadoya³ initially reported two such cases, and Komiyama's group⁷ reported a 10.7% incidence of such association on cerebral angiography. Although our number of patients was small, two of the 72 patients (2.8%) with moyamoya disease had a persistent trigeminal artery. In the general population, the incidence of persistent trigeminal artery on MR angiography is reported as 0.29 to 0.34%^{11,12}. Therefore, the incidence of persistent trigeminal artery is at least ten times higher among patients with moyamoya disease.

We found a high incidence of associated arterial fenestration in the vertebrobasilar sys-

Table 1 Cerebral arterial variations associated with moyamoya disease.

Case Number	Age (years)	Sex	Arterial variations	Figure
1	11	M	Basilar artery fenestration	Figure 1
2	26	F	Basilar artery fenestration	
3	40	M	Basilar artery fenestration	
4	57	F	Basilar artery fenestration*	
5	67	F	Bilateral ophthalmic arteries from middle meningeal artery	Figure 2
6	55	F	Left ophthalmic artery from middle meningeal artery	
7	53	F	Right intracranial vertebral artery fenestration	
8	45	F	Left intracranial vertebral artery fenestration, Left persistent first cervical intersegmental artery	Figure 3
9	72	F	Left persistent first cervical intersegmental artery	
10	15	F	Left persistent trigeminal artery	Figure 4
11	20	M	Right persistent trigeminal artery	
12	39	F	Left extracranial origin of the posterior inferior cerebellar artery	
13	42	F	Right persistent stapedial artery	Figure 5

*An aneurysm was seen at the proximal end of the fenestration.

tem, an observation not reported previously. For example, we observed four fenestrations of the basilar artery in 72 patients (5.6%) on MR angiography compared to 2.07% in the general population¹³, an almost twofold incidence among patients with moyamoya disease. Ohba and colleagues¹⁰ reported a case of unilateral moyamoya disease associated with ruptured aneurysm at the basilar artery fenestration; among our four patients, one aneurysm was found at the fenestration. Steno-occlusive lesions of the carotid system produce hemodynamic stress in the vertebrobasilar system that causes aneurysms to form¹⁴. We observed two fenestrations of the intracranial vertebral artery (2.8%) in our study, and an incidence of 0.54% is reported in the general population¹³. Our observation of two persistent first cervical intersegmental arteries and one extracranial origin of the posterior inferior cerebellar artery was consistent with frequencies noted in the general population¹⁵.

There tended to be a female predominance (female:male, 20.8:12.5%) but without statistical significance between genders, probably due to our small number of cases.

We observed no variations in either the middle or anterior cerebral arteries. The presence of steno-occlusive lesions in the area of observation probably prevented detection of variations in the middle or anterior cerebral arteries. If MR angiography was performed in the

early stage of moyamoya disease, such variations as duplications and fenestrations might be found in these two arteries. We diagnosed three ophthalmic arteries arising from the middle meningeal artery (2.1% per artery), which was comparable to the 1.45% per artery reported in the general population¹⁶.

In the case of occlusion of the proximal ICA, anastomosis from the middle meningeal artery to the ophthalmic artery may play an important role for the collateral circulation to the distal ICA. In contrast, the terminal segment of the ICA is occluded in advanced stages of moyamoya disease. Thus, the ophthalmic artery arising from the middle meningeal artery has no clinical significance in moyamoya disease. We experienced a case of persistent stapedial artery in which the middle meningeal artery arose from the petrous segment of the ICA, an extremely rare variation¹⁷.

Moyamoya disease is a progressive steno-occlusive disease of the cerebral arteries usually diagnosed during childhood or young adult life. Its congenital origin may account for the variations in cerebral arteries that frequently occur in the disease.

Our study has several limitations. It was a retrospective clinical study performed in one institution; case materials were limited in number; and only one author analyzed images. A larger number of MR angiographic images of patients with moyamoya disease should be

Figure 3 Case 8. A 45-year-old woman with moyamoya disease. Anteroposterior projection of magnetic resonance angiography shows fenestration of the left vertebral artery (VA) at its V4 segment (long arrow). The left V3-V4 junction is lower than that of the right VA, indicative of persistent first cervical intersegmental artery (short arrow).

Figure 4 Case 10. A 15-year-old girl with moyamoya disease. Anteroposterior projection of magnetic resonance angiography shows lateral-type left persistent trigeminal artery (arrow).

analyzed in a multicenter study to verify our findings.

Conclusions

We report the incidence of variations of the cerebral arteries in patients with moyamoya disease. Moyamoya disease was frequently as-

sociated with such variations, especially fenestrations in the vertebrobasilar system and persistent trigeminal artery.

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Figure 5 Case 13. A 42-year-old woman with moyamoya disease. A) Left anterior oblique projection of magnetic resonance angiography shows the right middle meningeal artery arising from the petrous internal carotid artery, indicative of a persistent stapedia artery (arrow). B) Partial maximum intensity projection image of the right carotid system clearly demonstrates this anomalous artery (arrow).

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- Akira Uchino, MD, PhD
 Department of Diagnostic Radiology
 Saitama Medical University International Medical Center
 1397-1 Yamane, Hidaka, Saitama 350-1298, Japan
 Tel.-Fax: +81-42-984-4520
 E-mail: auchino@saitama-med.ac.jp