Clinical Features and Outcomes of Intracranial Aneurysm Associated with Moyamoya Disease

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\textbf{Background and Purpose} Moyamoya disease (MMD) is a rare form of intracranial steno-occlusive disease that can be associated with intracranial aneurysms. We evaluated the clinical features and outcomes of MMD-associated aneurysms while focusing on their locations.

\textbf{Methods} Between January 1998 and December 2018 there were 1,302 adult and pediatric patients diagnosed as MMD at a single institution. These patients included 38 with 44 MMD-associated aneurysms. The MMD-associated aneurysms were classified into two groups based on their locations: major-artery aneurysms and non-major-artery aneurysms. The clinical and radiological data for patients with MMD-associated aneurysms were reviewed retrospectively.

\textbf{Results} The 44 MMD-associated aneurysms comprised 28 in major arteries and 16 in non-major arteries. All of the major-artery aneurysms were initially unruptured lesions, and follow-up angiography showed that 23 (82.1\%) had an improved or stable status and 5 (17.9\%) had a worse status. The non-major-artery aneurysms comprised 10 ruptured and 6 unruptured lesions, and follow-up angiography showed that 11 (68.8\%) had improved or were stable and 5 (31.2\%) had worsened. At the latest follow-up, there were four cases of unfavorable outcome: two initial hemorrhagic insults, one treatment-related morbidity, and one repeated-hemorrhage case.

\textbf{Conclusions} MMD-associated aneurysms occurred in 3.3\% of the MMD cohort in this study, of which 63.6\% were major-artery aneurysms and 36.4\% were non-major-artery aneurysms. The major-artery group included 17.9\% that became angiographically worse, while 31.2\% were growing or hemorrhaging in the non-major-artery group.

\textbf{Key Words} moyamoya disease, aneurysm, pseudoaneurysm, outcome.

INTRODUCTION

Moyamoya disease (MMD) is a rare form of cerebrovascular disease of unknown etiology. MMD has a low reported prevalence of 1.61–16.1 per 100,000, but it is a major cause of stroke in pediatric and young-adult age groups.\textsuperscript{3} MMD is characterized by progressive non-atherosclerotic steno-occlusion of intracranial arteries along with their collateral networks,\textsuperscript{2} and these structural changes can be vulnerable to the formation of intracranial aneurysms. These characteristics reportedly result in 3.6–12.9\% of MMD patients showing accompanying aneurysms,\textsuperscript{1,4} which is much higher than the rate seen in the general population. Above all, these aneurysms are frequently associated with hemorrhagic events in MMD patients.

MMD-associated aneurysms can theoretically develop in any intracranial vessels, such as the circle of Willis, moyamoya vessels, and other collateral networks. These sites of MMD-associated aneurysms are classified into two artery groups: major and nonmajor arteries. Major-artery aneurysms are located within the circle of Willis, while non-major-artery aneurysms are located in peripheral arteries distal to the circle of Willis, in moyam-
MMD-associated aneurysms were known to vary with their location. Kwak et al.\(^7,8\) reported that the hemorrhagic rate was higher for peripheral artery aneurysms (6/14) than for major-artery aneurysms (3/13). Konishi et al.\(^9\) reported that in their MMD patients, all aneurysms within moyamoya vessels (5/5) were ruptured lesions, while only one aneurysm within the circle of Willis (1/2) was ruptured.

These variations indicate that anatomical and clinical differences should be considered when attempting to ensure the appropriate management of MMD-associated aneurysms. To the best of our knowledge, no previous studies have compared MMD-associated aneurysms according to their locations. Moreover, most previous studies have had cross-sectional designs, and the long-term clinical courses of MMD-associated aneurysms were not well documented.

The present study evaluated and compared the clinical features and outcomes of MMD-associated aneurysms according to their locations.

**METHODS**

**Patient population**

This study performed a retrospective review of prospectively collected MMD data in a single institute. The Institutional Review Board at the institute approved this study and waived the requirement to obtain informed consents from patients due to its retrospective design (IRB No. 4-2018-0064).

Between January 1998 and December 2018, 1,302 adult and pediatric patients were diagnosed as bilateral or unilateral MMD at a single institution. All of them underwent either magnetic resonance angiography (MRA) or biplane digital subtraction angiography (DSA), and the diagnosis of MMD was based on Japanese guidelines established by the Research Committee on Spontaneous Occlusion of the Circle of Willis (Moyamoya Disease).\(^10,11\) No other vasculopathy responsible for moyamoya syndrome was included.

MMD-associated aneurysms were concomitantly diagnosed in 43 (3.3%) of the 1,302 MMD patients. After excluding 5 patients with 6 aneurysms from this study due to the clinical and radiological follow-up lasting for ≤12 months, 38 patients with 44 MMD-associated aneurysms were finally included.

All clinical and radiological data were obtained from electronic medical records and a prospectively registered MMD and aneurysm database, and were reviewed retrospectively.

**Classification of MMD-associated aneurysms**

MMD-associated aneurysms were classified into two groups based on the regional relationship between the parent artery of the aneurysm and the circle of Willis.\(^5,6,12\) Major-artery aneurysm was defined as an MMD-associated aneurysm located in the arteries of the circle of Willis. Non-major-artery aneurysm was defined as an aneurysm located in the peripheral arteries distal to the circle of Willis, in moyamoya vessels, or in other collateral networks.

**Treatment of MMD-associated aneurysms**

In accordance with the protocol that is normally applied at our institute, a definite treatment (either surgical or endovascular) was considered for ruptured aneurysms at the time of diagnosis. Regular follow-up was recommended for unruptured aneurysms, but if a lesion became enlarged or ruptured during follow-up, additional treatment would also be considered. Treatment planning was discussed by a multidisciplinary team, and the final treatment decision was based on considerations of its technical feasibility and risk.

After the procedure, angiographic and clinical outcomes were assessed by a single neuroradiologist and a single neurosurgeon based on consensus. Treatment-related complications were defined as the development of any deterioration or illness due to the procedure. Treatment-related morbidity was defined as the development of any new deficit due to treatment-related complications that remained present at discharge. Treatment-related mortality was defined as patient death due to treatment-related complications during admission or clinical follow-up.

**Follow-up and outcome measurements**

Clinical and angiographic follow-up data were available for all of the included patients. Routine clinical follow-ups were conducted at 1, 6, 12, 24, and 48 months for both ruptured and unruptured aneurysms, while routine angiographic follow-ups were conducted at 12, 24, and 48 months. Additional follow-ups were performed when deemed necessary based on the clinical situation of each patient.

The clinical follow-up was independently conducted by a board-certified neurosurgeon, and clinical outcomes were assessed using the modified Rankin Scale (mRS) at the latest clinical follow-up. An unfavorable outcome was defined as an mRS score of 3 or more. Angiographic follow-up was conducted either MRA or DSA. The imaging data were reviewed retrospectively by two investigators independently. When these two investigators arrived at different diagnoses, another neuroradiologist reviewed the data and then the three investigators together decided on the final result based on consensus. Follow-up angiographic results were categorized into 1) improved or stable (spontaneously regressed or size unchanged) and 2) worse (increased size or hemorrhaging).
**Statistical analysis**
This study included the demographics and results of patients with MMD-associated aneurysms, and so only descriptive statistics are presented without any comparison with other types of disease or treatment. All data are presented as mean±standard-deviation values for continuous variables and as number and percentage values for categorical variables.

**RESULTS**

**Characteristics of patients and MMD-associated aneurysms**
The demographics of the patients are summarized in Table 1. The 38 patients (10 males and 28 females aged 40.2±12.6 years) with 44 MMD-associated aneurysms comprised 10 who initially presented with aneurysmal hemorrhage (all in nonmajor arteries), 6 with nonaneurysmal hemorrhage, 9 with cerebral infarction, 7 with transient ischemic attack, and 6 with incidental findings. One of the 10 patients with aneurysmal hemorrhage experienced a ruptured pseudoaneurysm in the distal anterior choroidal artery that was initially treated by endovascular embolization using N-butyl cyanoacrylate (Cordis Neurovascular, Miami Lakes, FL, USA), and

![Clinical courses of cases of moyamoya disease-associated aneurysms](image)

Fig. 1. Clinical courses of cases of moyamoya disease-associated aneurysms. In the major-artery group (n=28, mean follow-up of 68.1 months), 5 (17.9%) aneurysms showed growing and 23 showed an improved or stable status (1 spontaneously regressed and 22 were unchanged). All five growing lesions were treated, and one case of procedure-related complication occurred. In the non-major-artery group (n=16, mean follow-up of 56.4 months), 10 initially ruptured lesions were identified. Five (31.3%) lesions worsened during the follow-up: three repeated hemorrhages from ruptured aneurysms, one new hemorrhage, and one growing from an unruptured aneurysm. *One treatment-related complication (modified Rankin Scale score=3), †Including one initially treated.

Table 1. Demographic characteristics of patients with MMD-associated aneurysms

| Demographic characteristic | Value (n=38) |
|----------------------------|-------------|
| Sex, male:female           | 10:28       |
| Age, years                 | 40.2±12.6   |
| Laterality of MMD          |             |
| Bilateral involvement      | 30 (78.9)   |
| Unilateral involvement     | 8 (21.1)    |
| Initial presentation       |             |
| Incidental                 | 6 (15.8)    |
| Ischemia*                  | 16 (42.1)   |
| Hemorrhage†                | 16 (42.1)   |

Data are n, mean±standard-deviation or n (%) values.
*9 cerebral infarctions and 7 transient ischemic attacks, †10 aneurysmal and 6 nonaneurysmal hemorrhages.
MMD: moyamoya disease.
there were no procedure-related complications. The remaining nine ruptured non-major-artery aneurysms were conservatively managed and followed up due to technical inaccessibility or the expectation of complications (Fig. 1).

The 44 MMD-associated aneurysms comprised 28 (63.6%) major-artery aneurysms and 16 (36.4%) non-major-artery aneurysms. All of the major-artery aneurysms were unruptured lesions, whereas 10 ruptured and 6 unruptured lesions were identified in the non-major-artery aneurysms (Fig. 1). The major-artery aneurysms comprised 9 in the basilar artery (BA), 10 in the intradural segment of the internal carotid artery (ICA), 5 in the anterior communicating artery, 3 in the proximal posterior cerebral artery (PCA), and 1 in the middle cerebral artery. The non-major-artery aneurysms comprised seven in the posterior choroidal artery, three in the anterior choroidal artery, and six in moyamoya vessels.

The major-artery aneurysms comprised 27 saccular types and 1 fusiform type, while the non-major-artery aneurysms comprised 6 saccular types and 10 pseudoaneurysms. The sizes of the major-artery and non-major-artery aneurysms at the time of diagnosis were $3.6 \pm 2.0$ mm and $3.4 \pm 1.9$ mm, respectively, while their Suzuki stages were $2.4 \pm 1.0$ and $3.1 \pm 0.5$, respectively. The characteristics of the MMD-associated aneurysms are summarized in Table 2.

### Clinical courses and follow-up outcomes of MMD-associated aneurysms

During the follow-up lasting a mean of 62.5 months (range 14–186 months), 34 (77.2%) aneurysms had improved or were stable (including an initially embolized aneurysm) and 10 (22.8%) had worsened. The natural courses and follow-up results are summarized in Fig. 1.

Follow-up angiography in the major-artery group ($n=28$) showed 23 (82.1%) aneurysms with an improved or stable status (1 spontaneously regressed and 22 were unchanged; Fig. 2) and 5 (17.9%) that had worsened (all growing). These five growing lesions (three at the top of the BA, one at the

### Table 2. Characteristics of moyamoya disease-associated aneurysms

| Aneurysm characteristic | Major artery ($n=28$) | Nonmajor artery ($n=16$) |
|-------------------------|-----------------------|--------------------------|
| Rupture                 | 0 (0.0)               | 10 (62.5)                |
| Location                |                       |                          |
| BA                      | 9 (32.1)              | -                        |
| Proximal PCA            | 3 (10.7)              | -                        |
| ICA intradural segment  | 10 (35.7)             | -                        |
| Acom                    | 5 (17.9)              | -                        |
| MCA                     | 1 (3.6)               | -                        |
| Posterior choroidal artery | -                    | 7 (43.8)                |
| Anterior choroidal artery | -                    | 3 (18.7)                |
| Moyamoya vessels        | -                     | 6 (37.5)                |
| Type                    |                       |                          |
| Saccular                | 27 (96.4)             | 6 (37.5)                 |
| Fusiform                | 1 (3.6)               | 0 (0.0)                  |
| Pseudoaneurysm          | 0 (0.0)               | 10 (62.5)                |
| Size at diagnosis, mm   | $3.6 \pm 2.0$          | $3.4 \pm 1.9$            |
| Suzuki stage at diagnosis | $2.4 \pm 1.0$        | $3.1 \pm 0.5$            |

Data are $n$ (%) or mean±standard-deviation values.

Acom: anterior communicating artery, BA: basilar artery, ICA: internal carotid artery, MCA: middle cerebral artery, PCA: posterior cerebral artery.

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**Fig. 2.** A 40-year-old female with ischemic-type unilateral moyamoya disease. A: Frontal projection of the left carotid angiogram shows a small aneurysm in the right A1 (black arrow) and another aneurysm in the supraclinoid segment of the left internal carotid artery (black arrowhead). B: Angiogram obtained at the 29-month follow-up shows spontaneous regression of the A1 aneurysm (black arrow) and an unchanged left aneurysm (black arrowhead).
junction between the BA and superior cerebellar artery, and one in the proximal PCA) were treated by coiling (Fig. 3). The lesions at the top of the BA and at the BA–superior cerebellar artery junction were simultaneously treated in one patient, but a procedure-related complication occurred (in-stent thrombosis). This complication was managed by administering an intra-arterial infusion of tirofiban, but the latest clinical outcome was unfavorable (mRS score=3).

In the non-major-artery group (n=16), 11 (68.8%) aneurysms had improved or were stable (5 spontaneously regressed and 6 unchanged; Fig. 4) and 5 (31.2%) had worsened (4 hemorrhage and 1 growing) at the follow-up. One case of repeated hemorrhage was successfully treated using the endovascular method, while the remaining four worsened lesions were conservatively managed and followed up for 29–72 months, during which no further events occurred.

At the latest clinical follow-up there were four cases (10.5%) of unfavorable outcome: two initial hemorrhagic insults (mRS score=3 and 5 in nonmajor arteries), one case of treatment-related morbidity (mRS score=3 in a major artery), and one repeated-hemorrhage case (mRS score=5 in a nonmajor artery). The treatment details for the MMD-associated aneurysms and treatment-related complications are summarized in Table 3.

DISCUSSION

This study found that 1) MMD-associated aneurysms were present in 3.3% of the MMD cohort, 2) all of the major-artery aneurysms were initially unruptured and ruptured aneurysms that predominated in nonmajor arteries (0% versus 62.5%), and 3) 22.8% of the aneurysms (17.9% in major ar-

Fig. 3. A 43-year-old female with a BA aneurysm. A: Image obtained in the initial MRA shows a 6-mm aneurysm at the top of the BA (white arrow). B: MRA image obtained at the 10-year follow-up shows a growing aneurysm (from 6 mm to 11 mm) (white arrow). C and D: The control angiogram obtained after stent-assisted coiling shows complete occlusion of sac (black arrows). BA: basilar artery, MRA: magnetic resonance angiography.
The gradual occlusion of the ICA associated with MMD progression alters the hemodynamic stress around the circle of Willis. Such a hemodynamic burden can directly affect major arteries and lead to aneurysm formation, which are morphologically similar to common saccular lesions. However, in contrast to common saccular aneurysms, MMD-associated aneurysms usually develop in the posterior circulation, particularly in the BA. One retrospective analysis of 111 cases of MMD-associated aneurysm found that 58.9% of major-artery aneurysms were located in the posterior circulation. In the present study, the major-artery aneurysms were commonly located in the posterior circulation (42.9%, 12/28). Above all, the specific characteristics of MMD could affect the natural course of major-artery aneurysms, which differs from that of common saccular lesions. A study of small unruptured aneurysms found angiographic worsening in 4.5% of cases from the general population (10 growing and 7 hemorrhaging). On the other hand, there are also some reports of the spontaneous disappearance of anterior circulation aneurysms in MMD, while Ito et al. reported two cases of exacerbated aneurysms in the posterior circulation. These results might be due to both a progressive occlusion of the ICA and altered hemodynamic stress in MMD. The present study found that 17.9% of major-artery aneurysms showed angiographic growing during the follow-up, all of which were in the posterior circulation, while all of the an-

![Fig. 4. A 23-year-old female with intraventricular hemorrhage. A and B: Lateral projection of the left carotid angiogram (A) and 3-D volume-rendered reconstruction image (B) show a pseudoaneurysm (arrows) in the posterior choroidal artery and that the diameter of the PCA is normal (arrowheads). C and D: Follow-up angiogram (C) and 3-D volume-rendered reconstruction image (D) show spontaneous regression of the aneurysm (arrows) and narrowing of the PCA (arrowheads). PCA: posterior cerebral artery.](image-url)
eurysms in the anterior circulation had improved or were stable at follow-up. Unfortunately, applying open surgery to these aneurysms in the posterior circulation is challenging because they are located deeply and tiny fragile basal collaterals of MMD usually form around them. Thus, endovascular embolization can be a useful alternative treatment modality. However, considering the vasculopathy nature of MMD, endovascular treatment needs to be performed with great care by experienced neurointerventionalists. In the present study, we also experienced one case of in-stent thrombosis after applying stent-assisted coiling to an aneurysm at the top of the BA. Meanwhile, in the major-artery group, 82.1% of MMD-associated aneurysms were angiographically improved or stable, especially for those in the anterior circulation. Thus, careful consideration is required when determining the optimal treatment for these aneurysms.

Meanwhile, moyamoya vessels and other collateral networks in MMD have critical histological defects and also reductions in wall thicknesses due to characteristics such as fragmented internal elastic lamina and tunica media.21,22 These structural features are related to repeated hemorrhagic or ischemic events in MMD,23 and they can also contribute to the formation of microaneurysms in the small peripheral arteries, particularly in posterior or anterior choroidal arteries, and lenticulostriate arteries.24,25 As a result, such non-major-artery aneurysms are more fragile and prone to rupture than major-artery aneurysms.26 In the present study, 62.5% (10/16) of the non-major-artery aneurysms were initially ruptured lesions, while no such lesions appeared in the major-artery aneurysms. Moreover, long-term follow-up revealed angiographic worsening of 31.2% (5/16) and 17.9% (5/28) of the non-major-artery and major-artery aneurysms, respectively. Although non-major-artery aneurysms and their parent arteries are smaller than major-artery aneurysms, the frequent occurrence of repeated hemorrhage can have fatal outcomes27 because they are commonly located near the periventricular area or deep nuclei.28 Thus, definite treatments can be considered for non-major-artery aneurysms with the aim of preventing hemorrhage. Revascularization surgery might be effective for the treatment of non-major-artery aneurysms,29,30 which alters the intracranial hemodynamics and accelerates the steno-occlusion of pathogenic MMD vessels; however, this intervention requires an interval between the surgery and aneurysm obliteration. Alternatively, endovascular embolization has been successfully applied to such lesions. However, some lesions are endovascularly inaccessible due to the small diameter or tortuosity of the parent vessels, with the rate of inaccessibility reportedly varying from 12.5% to 73.7%.25,26 Non-major-artery aneurysms can spontaneously disappear,26,31 and this occurred in 31.2% of

| Pt. | Aneurysm group | Aneurysm | Aneurysm size, mm | Treatment method | Diagnosis-to-treatment time, months | Angiographic result | Treatment-related complications | Preop. mRS score | Postop. mRS score | In-stent thrombosis† |
|-----|----------------|----------|-------------------|-----------------|-----------------------------------|---------------------|--------------------------------|------------------|------------------|---------------------|
| 1*  | Major          | Unruptured→growing BA→growing BA | 5−8 | Stent-assisted coiling | 90 | Complete | In-stent thrombosis† | 0 | 3 | - |
| 2   | Major          | Unruptured→growing BA→growing BA→superior cerebellar artery | 1→2 | Stent-assisted coiling | 90 | Complete | Complete | 0 | 0 | - |
| 3   | Major          | Unruptured→growing BA→growing BA→proximal posterior cerebellar artery | 5−7 | Simple coiling | 53 | Complete | Complete | 0 | 1 | - |
| 4   | Major          | Unruptured→growing BA→distal anterior choroidal artery | 1→11 | Stent-assisted coiling | 0 | Complete | None | 0 | 0 | - |
| 5   | Nonmajor       | Initially ruptured BA→growing Moyamoya vessel | 1→4 | NBCA embolization | 13 | Complete | Complete | None | None | - |
| 6   | Nonmajor       | Ruptured→repeated hemorrhage Moyamoya vessel | 1→4 | NBCA embolization | 13 | Complete | Complete | None | None | - |

*Simultaneous coiling applied to two separate aneurysms in one patient.†Managed using intra-arterial tirofiban.

BA: basilar artery, mRS: modified Rankin Scale, NBCA: N-butyl cyanoacrylate, Postop.: postoperative, Preop.: preoperative, Pt.: patient number.
our non-major-artery aneurysms. For this reason, some authors have suggested conservative treatment for non-major-artery aneurysms in MMD. However, the optimal follow-up strategies and outcomes remain unclear since most published experiences have related to case reports. Therefore, further studies are needed in larger populations with MMD-associated aneurysms to assess the long-term clinical courses and outcomes.

Due to the limitations inherent in the retrospective design of this study, selection bias may have affected the demographics of the included patients and the percentage values obtained. To lessen such selection bias, patients were extracted from a large MMD cohort using electronic medical records. The smallness of the sample could also be another limitation. However, MMD is a rare disease and, to the best of our knowledge, this study involved the largest sample gathered in a single center.

In conclusion, MMD-associated aneurysms occurred in 3.3% of the MMD cohort in this study, with 63.6% in major arteries and 36.4% in non-major arteries. Angiographic worsening was observed in 17.9% of the aneurysms in the major arteries, particularly in the posterior circulation, while 31.2% of those in non-major arteries were growing or hemorrhaging.

Author Contributions
Conceptualization: Dong-Seok Kim, Keun Young Park. Data curation: all authors. Formal analysis: Sunghan Kim, Keun Young Park. Investigation: Sunghan Kim, Chang Ki Jang, Eun-Kyung Park, Kyu-Won Shim, Dong-Seok Kim, Keun Young Park. Methodology: Sunghan Kim, Eun-Kyung Park, Kyu-Won Shim, Dong-Seok Kim, Keun Young Park. Supervision: Keun Young Park. Writing—original draft: Sunghan Kim, Keun Young Park. Writing—review & editing: Chang Ki Jang, Eun-Kyung Park, Kyu-Won Shim, Dong-Seok Kim, Joonho Chung, Yong Bae Kim, Jae Whan Lee.

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Conflicts of Interest
The authors have no potential conflicts of interest to disclose.

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