The clinical characteristics and treatment considerations for intracranial aneurysms associated with middle cerebral artery anomalies: a systematic review

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Research article

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Abstract

Background: As a result of its low occurrence, most of the studies on intracranial aneurysms associated with MCA anomalies were presented as case reports or small case series. In this study, a systematic review on this specific entity was conducted.

Methods: A PubMed search of the published studies was performed on April 6th, 2019 for patients who had intracranial aneurysms associated with MCA anomalies. The languages included in this study were English, Chinese, and Japanese.

Results: Finally, 58 articles reporting 67 patients including 1 case in our center were included. The identified patients (37 females, 55.2%) aged from 4 to 81 (49.85 ± 15.22) years old. Fifty (50/67, 74.6%) patients presented with hemorrhagic stroke either from the MCA anomalies associated aneurysms or other sources. Sixty-three aneurysms (63/67, 94.0%) were saccular, 3 (4.5%) were dissecting or fusiform, and 1 (1.5%) was pseudoaneurysm. Thirty-two (32/65, 49.2%) patients had other concurrent cerebrovascular anomalies. Fifty-six (83.6%) patients underwent open surgeries, 8 (11.9%) patients underwent endovascular treatment, and 3 (4.5%) patients were conservatively managed. Fifty-six (56/61, 91.8%) patients achieved a good recovery.

Conclusions: The pathophysiological genesis of MCA anomalies associated aneurysms is still obscure to us. The inflicted patients tend to have other concurrent cerebrovascular anomalies, which denotes that congenital defect in cerebrovascular development might play a role in this process. Open surgery is, hitherto, the mainstay of treatment for this specific entity. Most of the affected patients could experience a good recovery after treatment.

Background

Middle cerebral artery (MCA) is the largest and most important branch of the internal carotid artery. Compared to its counterparts of the posterior circulation, MCA has a lower incidence of vascular anomalies [1]. Generally speaking, MCA anomalies include accessory MCA (ac-MCA), duplicate MCA (d-MCA), d-MCA origin, MCA fenestration, and twig-like MCA (Figure 1). In rare circumstances, the MCA anomalies can be associated with intracranial aneurysms [2-4]. As a result of its low occurrence, most of the studies on intracranial aneurysms associated with MCA anomalies were presented as case reports or small case series. Hence, large-scale investigation on this rare entity in a single center is unrealistic. In this study, we would like to perform a systematic review on this specific entity to further elucidate its demographic, clinical, therapeutic, and prognostic characteristics.

Methods

A PubMed search of the published studies was performed on April 6th, 2019 for patients who had intracranial aneurysms associated with MCA anomalies. The languages included in this study were English, Chinese, and Japanese. The algorithm used in this search was (((((accessory middle cerebral
artery [Title/Abstract] OR duplicate middle cerebral artery [Title/Abstract] OR duplicated middle cerebral artery [Title/Abstract] OR duplicate middle cerebral artery origin [Title/Abstract] OR duplicated middle cerebral artery origin [Title/Abstract] OR fenestration of middle cerebral artery [Title/Abstract] OR fenestrated middle cerebral artery [Title/Abstract] AND aneurysm [Title/Abstract]. Only articles of which the full text or enough information could be obtained were included in this study. Reference lists of the identified articles were also manually searched for additional studies. Glasgow Outcome Scale was used for the outcome assessment. A Glasgow Outcome Scale score $\geq 4$ was defined as good recovery. An aneurysm $< 10$ mm was defined as small aneurysm.

**Definition of intracranial aneurysm associated with MCA anomalies**

Intracranial aneurysms located at the beginning or on the trunk of the abnormal MCAs were considered as in association with MCA anomalies. Aneurysms having no direct anatomical neighborhood with the MCA anomalies were excluded in the final analysis.

**Location of ac-MCA and the associated aneurysm**

Based on their sites of origin along the anterior cerebral artery (ACA), the ac-MCA are divided into 3 types: 1) originating from the A1 segment of the ACA, 2) originating from the anterior communicating artery (AComA) or the A1-A2 junction, 3) originating from the A2 segment. The locations of aneurysms were at the beginning or the trunk of the ac-MCA (Figure 1A-C).

**Location of d-MCA associated aneurysm**

The locations of aneurysms were at the beginning or on the trunk of the d-MCA (Figure 1D).

**Location of MCA fenestration associated aneurysm**

The locations of aneurysms were proximal to the fenestration, in the fenestration, or distal to the fenestration (Figure 1E).

**D-MCA origin aneurysm**

The locations of aneurysms were at the beginning of any branch of the duplicate origins or on the fused common trunk (Figure 1F).

**Results**

**General information**

The PubMed search identified 113 records. Fifty-nine records were excluded based on titles and abstracts screening. After assessing the full text of the remaining 54 articles, 5 were further excluded. A manual searching of the reference lists of the remaining 49 articles was performed, which yielded 9 additional
articles. Finally, 58 articles reporting of 67 patients including 1 case in our center were included for the
analysis. Searching strategy is presented in Figure 2.

The identified patients (37 females, 55.2%) aged from 4 to 81 (49.85 ± 15.22) years old. Fifty (50/67, 74.6%) patients presented with hemorrhagic stroke either from the MCA anomalies associated aneurysms or other sources. The MCA anomalies associated aneurysms were located at the left side in 32 (32/66, 48.5%) patients. Sixty-three aneurysms (63/67, 94.0%) were saccular, 3 (4.5%) were dissecting or fusiform, and 1 (1.5%) was pseudoaneurysm. Thirty-two (32/65, 49.2%) patients had other concurrent cerebrovascular anomalies in addition to MCA anomalies associated aneurysms. Fifty-six (83.6%) patients underwent open surgeries, 8 (11.9%) patients underwent endovascular treatment, and 3 (4.5%) patients were conservatively managed. Ten (10/62, 16.1%) patients experienced procedure-related complications. Fifty-six (56/61, 91.8%) patients achieved a good recovery.

Accessory MCA aneurysm

Nineteen studies reporting of 20 patients including 1 case in our center were identified (Table 1) [4-22]. The patients aged from 4 to 73 (48.65 ± 16.70) years old, with a male to female ratio of 1:1. Eighteen (18/20, 90%) patients presented with intracranial bleeding from ac-MCA associated aneurysms or other sources. The sizes of aneurysms were below and above 10 mm in 18 (90%) and 2 (10%) patients, respectively. Seventeen (85%) aneurysms were saccular, 2 (10%) were dissecting, 1 (5%) was pseudoaneurysm. The left to right ratio of aneurysm allocation was 1:1. The locations of ac-MCAs were A1, A1-A2 junction, and A2 in 16 (16/19, 84.2%), 2 (2/19, 10.5%), and 1 (1/19, 5.3%) patient, respectively. Of the 20 aneurysms, 14 (70%) were located at the origin of ac-MCA, 6 (30%) were on the trunk. Nine (9/18, 50%) patients had other concurrent cerebrovascular anomalies. With respect to the treatment, 14 (70%) patients underwent microsurgical clipping of the aneurysms, 4 (20%) (3 coiling, 1 glue embolization) underwent endovascular treatment, 1 (5%) underwent resection of the pseudoaneurysm and distal ac-MCA, and 1 (5%) underwent aneurysm wrapping. Procedure-related complications occurred in 1 (1/19, 5.3%) patient. Eighteen (18/20, 90%) patients experienced good recovery.

Duplicate MCA aneurysm

Twenty-seven studies reporting of 34 patients were finally included (Table 2) [3, 7, 23-47]. The patients aged from 20 to 76 (50.79 ± 13.72) years old, with a male to female ratio of 0.62:1 (13:21). Twenty (20/34, 58.8%) patients presented with intracranial bleeding from d-MCA aneurysms or other sources. The sizes of aneurysms were below and above 10 mm in 33 and 1 patients, respectively. All of the aneurysms were saccular except a fusiform one. The left to right ratio of aneurysm allocation was 1.2:1 (18:15). Of the 34 aneurysms, 32 (94.1%) were located at the origin of d-MCA, 2 were on the trunk. Nine (9/18, 50%) patients had other concurrent cerebrovascular anomalies. Of the 34 patients, 18 (52.9%) have concurrent cerebrovascular anomalies. Twenty-seven (79.4%) patients underwent microsurgical clipping of the aneurysms, 3 (8.8%) patients underwent endovascular coiling, 1 (2.9%) underwent trapping of the aneurysm and simultaneous superficial temporal artery-d-MCA anastomosis, and 3 (8.8%) patients were
conservatively followed up. Procedure-related complications occurred in 7 (22.6%) patients. Twenty-six (89.7%, 26/29) patients experienced good recovery.

**MCA fenestration aneurysm**

Twelve studies reporting of 12 patients were identified, aging from 14 to 81 (49.3 ± 13.1) years old (Table 3) [2, 48-58]. The male to female ratio was 1:1. All of the patients were admitted for intracranial bleeding. All of the aneurysms were smaller than 10 mm except for 1 the size of which could not be determined. All of the aneurysms were saccular. Nine (9/12, 75%) of the aneurysms were located at the right side. All of the fenestrations were located on M1 segment of the MCAs. The aneurysms were located proximal to, in, and distal to the fenestration in 5 (41.7%), 4 (33.3%), and 3 (25%) patients, respectively. Concurrent cerebrovascular anomalies were identified in 5 (41.7%) patients. With respect to the treatment, 10 patients underwent surgical clipping, 1 underwent aneurysm wrapping, and 1 underwent coiling. Two (2/11, 18.2%) patients experienced procedure-related complications. All of the patients experienced good recovery except for 1 patient the outcome of whom was not provided.

**Duplicate MCA origin aneurysm**

d-MCA origin aneurysm was only identified in a 49-year man incidentally, who was admitted for vertigo [59]. No other cerebrovascular anomaly was reported. The saccular unruptured d-MCA origin aneurysm was microsurgically clipped. The postoperative course was uneventful and no neurological deficit was reported.

**Illustrative case**

A 59-year old man was admitted for sudden onset of headache 2 days before. He was a smoker and denied history of any chronic diseases. He was alert on admission. Physical examination was unremarkable except for neck rigidity. Head computed tomography (CT) revealed subarachnoid hemorrhage of modified Fisher grade 2 (Figure 3 A-B). Further CT angiography showed the A1 segments of the bilateral ACAs gave rise to their respective ac-MCAs (Figure 3 C). A saccular aneurysm was noted at the origin of the left ac-MCA (Figure 3 C-D). No other cerebrovascular anomaly was identified. After discussion between the neurosurgical and neuro-interventional members and sufficient negotiation with the patient's legal relatives, endovascular coiling of the aneurysm was planned.

Preprocedural digital subtraction angiography also confirmed the findings on CT angiography (Figure 4 A-B). An Echelon-10 (Medtronic, Irvine, CA) microcatheter was advanced into the left ACA directed by a 0.010-in guidewire. The tip of the microcatheter was introduced into the aneurysm. The aneurysm was satisfactorily coiled using 3 detachable coils with preservation of the distal ACA and ac-MCA (Figure 4 C-D). He experienced an uneventful postprocedural recovery and was discharged the next day without neurological deficit. Follow-up CT angiography 1 year later revealed no recurrence of the aneurysm.

**Discussion**
According to Padget’s description, at 34-36 days of the embryonal stage (12-14 mm), multiple plexiform arterial twigs develop just distally to the anterior choroidal artery [60]. The plexiform arterial twigs would evolve into the MCA and lateral striate arteries through subsequent fusion and regression. Hypothetically, failure of this process can lead to diverse variations of the MCA (e.g. ac-MCA, d-MCA, MCA fenestration, d-MCA origin, and twig-like MCA) [1]. Generally speaking, the incidence of any MCA anomalies is very low [1, 61-63]. However, in even rarer circumstances, the MCA anomalies could be associated with intracranial aneurysms [2-4]. As a result of the low incidence of MCA anomalies, the reported cases of MCA anomalies associated intracranial aneurysms were all presented as case reports. Hence, the true incidence of MCA anomalies associated aneurysms in patients with MCA anomalies is still unknown.

According to this study, 49.2% of the patients had other concurrent cerebrovascular anomalies in addition to MCA anomalies associated aneurysms, which implies that this specific subset of patients might have congenital defect in cerebrovascular development and be prone to cerebrovascular anomalies.

According to Teal et al., anomalous arteries originating from the ACAs and coursing in parallel to or in close relationship with MCA were defined as ac-MCAs. And those arising from ICAs were considered d-MCAs [64]. The ac-MCA could be subdivided into 3 types based on their sites of origin (Figure 1A-C). Type I originates from the A1 segment of the ACA, type II originates from the A1-A2 junction (including AComA), and type III originates from the A2 segment. According to Kai et al., the d-MCA was further divided into 2 types (type A and type B) (Figure 1D) [34]. Those d-MCAs arising from the top of ICA bifurcation were defined as type A, and those between the anterior choroidal artery and the ICA top were type B. However, an exceptional case had been reported by Tutar et al., of which the d-MCA originated from the petrous portion of ICA [65]. In another case, a d-MCA originated from the ICA about 10 mm proximal to the ICA bifurcation and an ipsilateral fetal-type posterior cerebral artery originated from the d-MCA [27]. In our systematic review of the literature, ac-MCAs originated from the A1 segment in 84.2% (16/19) of the patients with ac-MCA associated aneurysms. And the d-MCA originated between the anterior choroidal artery and ICA bifurcation in all but 1 patient.

In contrast to the tendency for endovascular treatment for other intracranial aneurysms, 83.6% of the reported patients with MCA anomalies associated aneurysms underwent open surgeries, and only 11.9% of the patients underwent endovascular treatment. This phenomenon is still obscure to us. In our opinion, the reasons are multi-factorial. Firstly, as a result of the technical constraint, earlier cases had to undergo open surgeries. And then due to the unconventional locations and low incidence, medical practitioners are prone to the seemingly safer open surgical approaches. Thirdly, due to the specific local angioarchitecture, endovascular treatment might be more difficult. However, according to our study of the past reports, the outcome in the patients undergoing endovascular treatment was not inferior to that in patients undergoing open surgeries. And most of the cases with endovascular treatment were reported recently [20, 39, 46, 47, 56]. We believe that with the advancement in endovascular technology, more and more patients with MCA anomalies associated aneurysms would undergo endovascular treatment.

Due to rarity of the studied issue, the data of this review was extracted from retrospective case reports or small case series. The conclusions drawn from the reported cases might be affected by the inherent bias
of the evaluated case reports. Some anatomical, clinical, therapeutic, and prognostic details might be missed due to the different reporting customs. Statistical analysis is inappropriate for this kind of study. No comparative study between endovascular treatment and open surgery could be performed at present.

**Conclusions**

MCA anomalies have a low incidence of occurrence. Their associated intracranial aneurysms are even rarer that only sporadic reports were presented. The pathophysiological genesis of this specific entity is still obscure to us. The patients with MCA anomalies associated aneurysms tend to have other concurrent cerebrovascular anomalies, which denotes that congenital defect in cerebrovascular development might play a role in this process. Open surgery is, hitherto, the mainstay of treatment for this specific entity. Most of the affected patients could experience a good recovery after treatment.

**Declarations**

**Ethics approval**

This study was approved by the institutional review board of The First Hospital of Jilin University, and the participant gave his/her informed consent before inclusion in the study.

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**Availability of data and materials**

The datasets used and analyzed during the current study are available from the corresponding author on reasonable request.

**Authors’ Contributions**

Conception and design: JY, KH. Acquisition of data: KH, GL. Analysis and interpretation of data: HL, KX. Drafting of the article: KH, GL. Critical revision of the article: JY, KX. All of the authors have read and approved the final manuscript.

**Competing interests**

The authors declare that they have no competing interests.

**Consent for publication**

Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images. A copy of the written consent is available for review by the editor of this journal.
Abbreviations

ACA, anterior cerebral artery
AComA, anterior communicating artery
ac-MCA, accessory middle cerebral artery
CT, computed tomography
d-MCA, duplicate middle cerebral artery
GOS, Glasgow Outcome Scale
ICA, internal carotid artery
MCA, middle cerebral artery

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Tables
Table 1. Aneurysms associated with accessory MCA
| Case | Study | Age/sex | Presentation | Size (mm) | Morphology | Side | Location of ac-MCA and aneurysm | Accompanying intracranial anomalies | Treatment | Procedure-related complications | Outcome (GOS) |
|------|-------|---------|--------------|-----------|------------|------|--------------------------------|----------------------------------|-----------|---------------------------------|--------------|
| 1    | Waga et al., 1977<sup>5</sup> | 51/F | SAH | Small | Saccular | L | A1, origin | NA/N | Clipping | NA/N | 1 |
| 2    | Handa et al., 1984<sup>6</sup> | 55/F | SAH | Small | Saccular | R | A1, origin | NA/N | Clipping | No | 5 |
| 3    | Fuwa et al., 1984<sup>7</sup> | 57/M | SAH | Small | Saccular | L | A1, origin | Right ICA-PComA aneurysm | Clipping | No | 5 |
| 4    | Miyazaki et al., 1984<sup>8</sup> | 42/M | SAH | Small | Saccular | L | A1, origin | No | Clipping | No | 5 |
| 5    | Kubara et al., 1990<sup>9</sup> | 73/F | SAH | 4x6 | Saccular | R | A1, origin | No | Clipping | Hydrocephalus | 5 |
| 6    | Han et al., 1994<sup>10</sup> | 34/F | SAH | 4x5 | Saccular | L | A1, origin | No | Clipping | No | 5 |
| 7    | Sugita et al., 1995<sup>11</sup> | 53/M | Visual disturbance | Giant | Saccular | R | A1, origin | No | Clipping | No | 5 |
| 8    | Otawara et al., 1997<sup>12</sup> | 66/F | SAH | Small | Dissecting | R | A1, origin | Ipsilateral A1 dissecting aneurysm | Wrapping | No | 2 |
| 9    | Georgopoullos et al., 1999<sup>13</sup> | 32/F | SAH, IVH, ICH | Small | Saccular | L | A1, trunk | No | Clipping | No | 4 |
| 10   | Fujiwara et al., 2003<sup>14</sup> | 30/M | SAH | 3x5 | Saccular | R | A1, origin | No | Clipping | No | 5 |
| 11   | Kang | 38/M | SAH | 4x5 | Saccular | L | A1 | No | Clipping | No | 5 |
|   |   |   |   |   |   |   |
|---|---|---|---|---|---|---|
|   |   |   |   |   |   |   |
| 12 | Lee et al., 2009<sup>1</sup> 5 | 59/F | SAH | 7.3×4.8 | Saccular | L | A1, origin | Contralateral ac-MCA at A1 | Coil | No | 5 |
| 13 | Wakahayashi et al., 2011<sup>2</sup> 2 | 36/F | SAH | 3×6 | Saccular | L | A1, origin | Ipsilateral ac-MCA at A2 | Clipping | No | 5 |
| 14 | Lee et al., 2011<sup>2</sup> 1 | 56/M | SAH, IVH | Small | Lobular pseudoaneurysm | R | ACA, trunk | Right MMD involving ICA, ACA, and MCA | Resection of aneurysm and distal ac-MCA | No | 5 |
| 15 | Nomura et al., 2015<sup>4</sup> | 64/M | CI | Small | Saccular | L | A1, origin | No | Clipping | No | 5 |
| 16 | Teramoto et al., 2015<sup>1</sup> 7 | 68/M | ICH | 7 | Saccular | R | A2, trunk | Ipsilateral M1 stenosis | Clipping | No | 5 |
| 17 | Parthasarathy et al., 2015<sup>1</sup> 8 | 4/F | SAH | 8.8×2.1 | Fusiform dissecting | R | A1-A2 junction, trunk | Contralateral d-MCA | Aneurysm and parent artery occlusion with glue | No | 5 |
| 18 | Kheyreddin et al., 2017<sup>1</sup> 9 | 37/F | SAH and ICH | 25 | Saccular | R | A1, trunk | No | Clipping | No | 5 |
| 19 | Ren et al., 2018<sup>2</sup> 0 | 59/M | ICH | Small | Saccular | R | A1-A2 junction, trunk | Ipsilateral d-MCA origin | Palliative coiling | No | 5 |
| 20 | Present case | 59/M | SAH | 6.5 × 3.0 | Saccular | L | A1, origin | Contralateral ac-MCA | Coil | No | 5 |
Table 2. Aneurysms associated with duplicate MCA
| Case | Study | Age/sex | Presentation | Size (mm) | Morphology | Side | Location of aneurysm | Accompanying intracranial anomalies | Treatment | Postoperative complication | Outcome (GOS) |
|------|-------|---------|--------------|-----------|------------|------|---------------------|-----------------------------------|-----------|--------------------------|---------------|
| 1    | Stable et al., 1970 | 31/F | SAH | Small | Saccular | R | Origin | An aneurysm at the bifurcation of the left ICA | Clipping | Hydrocephalus | NA/NM |
| 2    | In et al., 1981 | 29/F | SAH | Small | Saccular | R | Origin | No | Clipping | No | 5 |
| 3    | Fuwa et al., 1984 | 46/F | SAH | Small | Saccular | R | Origin | No | Clipping | No | 5 |
| 4    | Takan o et al., 1988 | 74/M | Head trauma | 6 | Saccular | R | Origin | No | Clipping | Hydrocephalus | NA/NM |
| 5    | Dong et al., 1991 | 50/M | SAH | Small | Saccular | L | Origin | ACA fenestration, ac-MCA | Clipping | No | 5 |
| 6    | Takahashi et al., 1994 | 51/F | SAH | Small | Saccular | L | Origin | Contralateral carotid-ophthalmic aneurysm, d-MCA sharing common trunk with fetal PCA | Clipping | No | 5 |
| 7    | | 54/M | SAH | Small | Saccular | L | Origin | No | Clipping | Vasospasm | 3 |
| 8    | Koyamae et al., | 28/M | SAH | Small | Saccular | R | Origin | No | Clipping | No | 5 |
|   |   |   |   |   |   |   |   |   |
|---|---|---|---|---|---|---|---|---|
| 9 | Nomura et al., 2002 | 63/F | Incidental | Small | Saccular | L | Origin | No | Clippi ng | No | 5 |
| 10 | Tabuse et al., 2002 | 34/F | SAH | Small | Saccular | R | Origin | No | Clippi ng | No | 5 |
| 11 | Imaizumi et al., 2002 | 52/M | SAH | Small | Saccular | L | Origin | Contralateral ICA-PCom A aneurysm | Clippi ng | No | 5 |
| 12 | Uchino et al., 2004 | 45/F | SAH, ICH | Small | Saccular | L | Trunk | Bilateral ac-MCAs | Clippi ng | No | 5 |
| 13 | Hori et al., 2005 | 67/M | SAH, ICH | Small | Saccular | R | Origin | Ipsilateral ICA-PCom A aneurysm | Clippi ng | Aphasia, hydrocephalus | 3 |
| 14 | Kai et al., 2006 | 49/M | Incidental | Small | Saccular | L | Origin | BA tip aneurysm | Clippi ng | No | 5 |
| 15 | Kaliaperuma et al., 2007 | 63/F | Vertigo | Small | Saccular | L | Origin | No | Clippi ng and STA-d-MCA anastomosis | No | 5 |
| 16 | Miyahara et al., 2009 | 39/F | SAH | <10 | Saccular | L | Origin | No | Clippi ng | No | 5 |
| 17 | Miyahara et al., 2009 | 56/F | Incidental | Small | Saccular | R | Origin | 3 aneurysms at other locations | Clippi ng | No | 5 |
| 18 |   | 58/M | Vertigo | 7 | Saccular | R | Origin | Ipsilateral ICA- | Clippi ng | No | 5 |
| Case | Authors | Age | Gender | Event | Side | Origin | Location | Treatment | Type | Grad |
|------|---------|-----|--------|-------|------|--------|----------|-----------|-------|------|
| 19   | Otani et al., 2010 | 66/F | SAH | 6 | Saccular | R | Origin | Ipsilateral ac-MCA | Clipping | No | 5 |
| 20   | Kimura et al., 2010 | 60/F | Incidental | 4 | Saccular | L | Origin | No | Clipping | No | 5 |
| 21   | Takahashi et al., 2011 | 62/F | SAH | 4.0 x 4.2 | Saccular | L | Origin | Ipsilateral AChA aneurysm | Coiling | No | 5 |
| 22   | Labored et al., 2012 | 34/M | Incidental | 10 | Fusiform | L | Trunk | No | Trapping of the aneurysm and STA-MCA anastomosis | Cranial flap infection | 5 |
| 23   | Renne et al., 2013 | 52/F | Recurrent headache | 2 | Saccular | L | Origin | Ipsilateral supraclinoid ICA fenestration, AComA aneurysm | Clipping | No | 5 |
| 24   | Elsharkawy et al., 2012 | 62/M | Epilepsy | 12 | Saccular | L | Origin | No | Clipping | No | 5 |
| 25   |  | 55/F | SAH | 3 | Saccular | L | Origin | Cerebral ICA-PComA aneurysm | Clipping | No | 5 |
| 26   |  | 49/F | Migraine and double vision | 1 x 2 | Saccular | R | Origin | Cerebral ICA bifurcation aneurysm | Conservative management | NA/NM | NA/NM |
| 27   |  | 37/M | Incidental | 1 | Saccular | L | Origin | No | Conservative | NA/NM | NA/NM |
| No. | Authors            | Gender | Age | Location     | Side | Origin | Management  | Comments                      |
|-----|--------------------|--------|-----|--------------|------|--------|-------------|-------------------------------|
| 28  | Kim et al., 2015   | F      | 61  | Headache     | R    | Origin | Clipping    | No                            |
| 29  | Iida et al., 2015  | F      | 41  | SAH          | R    | Origin | Clipping    | No                            |
| 30  | Iida et al., 2015  | F      | 76  | SAH, ICH     | R    | Origin | Clipping    | Vasospasm, hydrocephalus     |
| 31  | Miyouhi et al., 2016 | F | 60 | SAH          | L    | Origin | Clipping    | Temporary aphasia            |
| 32  | Hayashi et al., 2017 | M  | 41  | SAH          | NA/NM| Origin | Coiling     | No                            |
| 33  | Mori et al., 2018  | M      | 62  | Alcoh abuse  | L    | Origin | Conserve     | NA/NM                        |
| 34  | Tsang et al., 2018 | F      | 20  | SAH          | R    | Origin | Coiling     | No                            |

**Abbreviations:** ACA, anterior cerebral artery; AChA, anterior choroidal artery; BA, basilar apex; F, female; GOS, Glasgow Outcome Scale; ICA, internal carotid artery; ICH, intracerebral hemorrhage; L, left; M, male; MCA, middle cerebral artery; NA/NM, not applicable or not mentioned; PCA, posterior cerebral artery; PComA, posterior communicating artery; R, right; SAH, subarachnoid hemorrhage; STA, superficial temporal artery

Table 1. Aneurysms associated with MCA fenestration
| Case | Study                     | Age/sex | Presentation | Size (mm) | Morphology | Side | Location of MCA fenestration and aneurysm | Accompanying intracranial anomalies | Treatment      | Procedure-related complication | Outcome (GOS) |
|------|---------------------------|---------|--------------|-----------|------------|------|-------------------------------------------|-----------------------------------|---------------|---------------------------------|---------------|
| 1    | Ueda et al., 1983         | 65/M    | ICH          | NA/N      | Saccular   | R    | M1, ipsilateral MCA bifurcation           | No                                | Clipping      | NA/N                           | 4             |
| 2    | Ueda et al., 1984         | 45/F    | SAH          | Small     | Saccular   | R    | M1, proximal to fenestration             | Multiple intracranial aneurysms, PTA | Clipping      | No                             | 5             |
| 3    | Kalia et al., 1991        | 49/M    | SAH          | Small     | Saccular   | R    | M1, in fenestration                      | AComA fenestration, multiple intracranial aneurysms, AVM | Wrapping      | No                             | 5             |
| 4    | Deruty et al., 1992       | 52/F    | SAH          | Small     | Saccular   | R    | M1, in fenestration                      | AComA aneurysm                    | Clipping      | No                             | 5             |
| 5    | Nakamura et al., 1994     | 36/F    | SAH          | Small     | Saccular   | L    | M1, proximal to fenestration             | No                                | Clipping      | No                             | 5             |
| 6    | Schmieder et al., 1997    | 14/M    | SAH          | Small     | Saccular   | L    | M1, proximal to fenestration             | No                                | Clipping      | No                             | 5             |
| 7    | Nussbaum et al., 2009     | 75/F    | SAH          | 5         | Saccular   | R    | M1, distal to fenestration               | No                                | Clipping      | No                             | 5             |
| 8    | Sim et al., 2010          | 32/M    | SAH          | 6         | Saccular   | R    | M1, in fenestration                      | Contralateral MCA                 | Clipping      | No                             | 5             |
|   | Author(s)                      | Year | Age | Sex | Size | Location | Side | Procedure | Adjacent Anatomy | Complications | Outcome | Grade |
|---|--------------------------------|------|-----|-----|------|----------|------|------------|------------------|---------------|----------|-------|
| 9 | Yamaguchi et al., 2010         | 81/F | SAH | Small | Saccular | R | M1, in fenestration | Contralateral MCA aneurysm | Coilung | No | 5     |
| 10| Tabuchi et al., 2014           | 47/F | SAH | Small | Saccular | R | M1, proximal to fenestration | No | Clipping | Hydrocephalus | 5        |
| 11| Sharifi et al., 2015           | 52/M | SAH | Small | Saccular | L | M1, distal to fenestration | Multiple intracranial aneurysms | Clipping | Bacterial meningitis | 5     |
| 12| Xue et al., 2019               | 43/M | SAH | 2.5  | Saccular | R | M1, proximal to fenestration | No | Clipping | No | 5     |

Abbreviations: AComA, anterior communicating artery; AVM, arteriovenous malformation; F, female; GOS, Glasgow Outcome Scale; ICH, intracerebral hemorrhage; L, left; M, male; MCA, middle cerebral artery; NA/NM, not applicable or not mentioned; PTA, persistent trigeminal artery; R, right; SAH, subarachnoid hemorrhage

Figures
Figure 1

The accessory MCA (brown branch) can originate from the A1 segment (A, Type 1), A1-A2 junction (B, Type 2), and A2 segment (C, Type 3) of the anterior cerebral artery. The accessory MCA associated aneurysms (black dots) can locate at the beginning or the trunk of the accessory MCA. The duplicate MCA (brown branch) can originate from the ICA bifurcation or between the AchA and MCA (D). The duplicate MCA associated aneurysms (black dots) can locate at the beginning or the trunk of the duplicate MCA. A fenestration can occur on the M1 segment of an MCA (E). The MCA fenestration associated aneurysms (black dots) can be proximal to, in, or distal to the fenestration. When duplicate MCAs fuse into one single trunk, it is called duplicate MCA origin (F). The duplicate MCA origin associated aneurysms (black dots) can locate at the beginning of each branch or the fused trunk.

Abbreviations: AchA, anterior choroidal artery; ICA, internal carotid artery; MCA, middle cerebral artery

Figure 2

Flow chart of searching strategy
Figure 3

Head CT shows subtle SAH (A-B). CTA reveals two ac-MCAs originate respectively from the A1 segment of the bilateral ACAs (C, black arrow) and an aneurysm originates from the beginning of the left ac-MCA (C-D, asterisk and white arrow). Abbreviations: ACA, anterior cerebral artery; ac-MCA, accessory middle cerebral artery; CT, computed tomography; CTA, CT angiography
Figure 4

Three dimensional (A) and plain (B) angiogram of the left ICA in AP view shows an ac-MCA arises from the A1 segment of ACA and a saccular aneurysm is also noted at the origin of ac-MCA. Angiogram of the left ICA in AP view shows the aneurysm is successfully coiled (C-D). Abbreviations: ACA, anterior cerebral artery; ac-MCA, accessory middle cerebral artery; AP, anteroposterior; ICA, internal carotid artery

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