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Case Report

Endovascular treatment of unruptured aneurysm arising from duplicate origin of the middle cerebral artery – A case report and literature review

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ABSTRACT

Background: Duplicate origin of the middle cerebral artery (MCA) is a rare variation of MCA, often mislabeled as the fenestration of the M1 segment of MCA.

Case Description: The authors treated an unruptured aneurysm, 8 mm in diameter, associated with a duplicate origin of MCA in a 42-year-old woman who underwent magnetic resonance imaging for transient vertigo. Clipping surgery was inapplicable due to the lack of space to insert clip blades between the neck and two origins of MCA. Under stent-assisted maneuver, the aneurysm sac was successfully obliterated using three coils, resulting in Raymond-Roy class 1 occlusion status. Digital subtraction angiography performed 3 months after the embolization showed complete obliteration of the aneurysm. So far, only 11 patients with aneurysms associated with duplicate origin of MCA have been reported. We performed a literature review of this very rare combination. The size of aneurysms ranged from 2 to 8 mm, with a mean of 5.2 mm. The neck of the aneurysm is mainly located at the corner between the inferior limb and the internal carotid artery. Ours is the youngest and has the largest aneurysm.

Conclusion: Aneurysm can arise from duplicate origin of MCA, for which stent-assisted coiling may be an appropriate treatment modality.

Keywords: Aneurysm, Coil embolization, Duplicate origin of middle cerebral artery, Endovascular treatment, Stent-assisted coiling

INTRODUCTION

Anatomical variations of proximal middle cerebral artery (MCA) include duplicate MCA, accessory MCA, early bifurcation of MCA, fenestrated MCA, and duplicate origin of MCA (DOMA).[6,13-17] The latter two are relatively rare, reportedly seen in only 0.11% and 0.09–0.3% of 1.5-T magnetic resonance angiography (MRA).[14-16] Moreover, DOMA has been often mislabeled as fenestrated MCA.[14,15] Fenestration of the MCA is a short slit-like division of the M1 or proximal M2 segment of MCA.[16] While DOMA is the condition in which two proximal MCA segments, superior and inferior limbs, arise separately from the terminal segment of the

internal carotid artery (ICA) and fuse to form an arterial ring succeeding to the distal M1 segment of MCA.[14,15] DOMA is supposedly formed by distal fusion of the duplicate MCA or accessory MCA in embryonic development. The anterior temporal artery often arises from the inferior limb.[14,15]

The authors hereby report a rare case of aneurysm arising from the proximal portion of DOMA, treated by endovascular coil embolization. A literature review of this rare combination is also performed.

CASE REPORT

An otherwise healthy 42-year-old woman suffering transient vertigo underwent magnetic resonance imaging (MRI), which did not show any causative condition for the symptom, whereas MRA found an aneurysm, 8 mm in the largest diameter, at the origin of the left MCA [Figure 1a]. Considering her age, the size of the aneurysm, and the irregular aneurysmal shape, she was subjected to further examination and possible treatment. Digital subtraction angiography (DSA) revealed a DOMA forming a 4 × 4 mm arterial ring composed of superior thick and inferior thin limbs. Moreover, the aneurysm arose from the corner between the ICA and the inferior limb [Figures 1b-d]. The irregular-shaped domes of aneurysm protruded both anteriorly and posteriorly from the ring. Aneurysm size was 8 × 4 × 4 mm, the largest in the anteroposterior diameter. Anterior temporal artery arose from the distal end of the inferior limb near the fusion point of two limbs, and the lenticulostriate artery arose from the superior limb. Due to the lack of space to insert blades of the aneurysm clip between the aneurysmal neck and the two limbs and the relatively wide diameter of the neck, a 4 mm stent-assisted coiling strategy was chosen to treat this aneurysm.

Before the treatment, the patient was given 2 weeks-long dual antiplatelet treatment with prasugrel hydrochloride 3.75 mg/day and acetylsalicylic acid 100 mg/day.

Under general anesthesia, a 9 Fr sheath was inserted into the right femoral artery. Through the sheath, the tip of an 8 Fr guiding catheter was put into the left cervical portion of the ICA, and then, a 6 Fr intermediate catheter was introduced into the cavernous portion of the ICA, through which microcatheters were advanced distally. Microcatheter-1 for stenting was advanced to M1-portion of the MCA through the superior limb. Through microcatheter-2, which was put into the aneurysmal sac, a Target 360 SOFT ® 5 mm × 15 cm coil (Stryker, USA) for framing was advanced into the sac. After the second loop of the framing coil was deployed, the tip of the coil was found inadvertently advanced into the inferior limb and then into the superior limb through the distal fusion point of the ring [Figures 2a and b]. The coil was pulled out, and the retrial achieved initial framing with three loops. To prevent the dislodgment of the coil, a Neuroform Atlas ® 4 mm × 21 mm stent (Stryker, USA) was deployed from the distal M1 segment of MCA through the superior limb of DOMA to C2 segment of ICA, just distal to the origin of the ophthalmic artery, through the microcatheter-1. After additional loops securing framing, the coil was

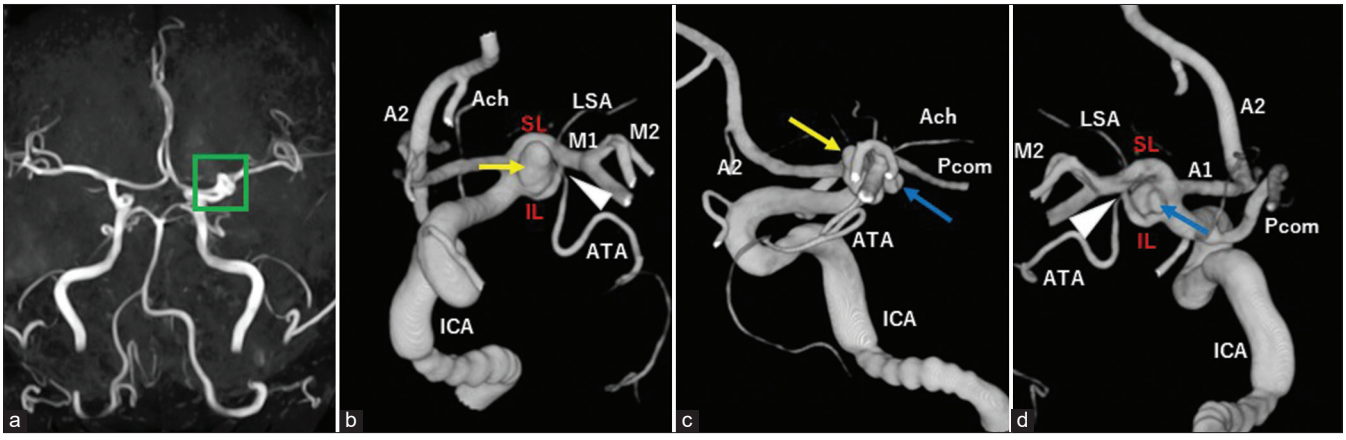

Figure 1: Imaging studies before treatment. (a) Anteroposterior view of MRA, (b) anteroposterior view of 3DR-DSA, (c) left-right view of 3DR-DSA, and (d) posteroanterior view of 3DR-DSA. MRA suggested an aneurysm (square in a) at the origin of the left MCA. The aneurysm with irregular configuration protruded anteriorly and posteriorly through the ring composed of two limbs of DOMA on 3DR-DSA images (b-d). Square: Supposed aneurysm on MRA: Magnetic resonance angiography, ICA: internal carotid artery, ATA: Anterior temporal artery, Ach: Anterior choroidal artery, LSA: lenticulostriate artery, SL: superior limb of DOMA, IL: inferior limb of DOMA, Pcom: posterior communicating artery, Arrowhead: fusing point of two limbs of DOMA, Yellow arrow: Anterior projection of aneurysm, Blue arrow: posterior projection of aneurysm, 3DR-DSA: Three-dimensional reconstruction image of digital subtraction angiography, MCA: Middle cerebral artery, DOMA: Duplicate origin of middle cerebral artery. A1 and A2: A1 and A2 portion of anterior cerebral artery, M1 and M2: M1 and M2 portion of middle cerebral artery.

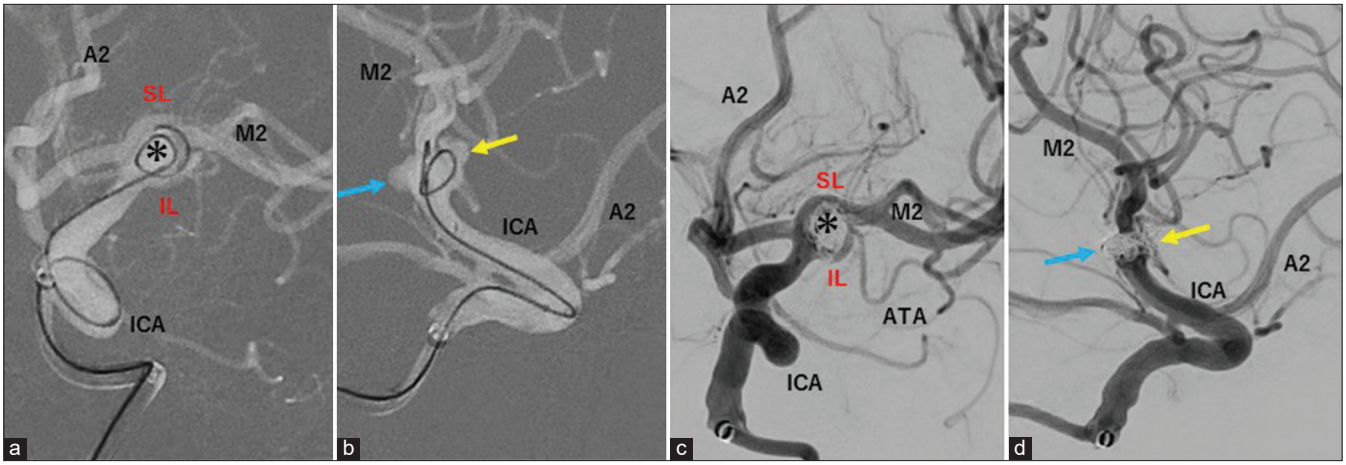

Figure 2: DSA of the left internal carotid artery during coil embolization. (a) Anteroposterior view of road mapping image during embolization, (b) right-left view of road mapping image during embolization, (c) anteroposterior view of DSA after the embolization, and (d) right-left view of DSA after the embolization. The tip of the coil inadvertently migrated into the upper limb through the lower limb of DOMA at the initial stage of framing (a and b). Complete obliteration of the aneurysm was achieved (c and d). Asterisk: Aneurysmal sac. ICA: Internal carotid artery, SL: Superior limb of DOMA, IL: Inferior limb of DOMA, ATA: Anterior temporal artery, Yellow arrow: Anterior projection of aneurysm, Blue arrow: Posterior projection of aneurysm, DOMA: Duplicate origin of middle cerebral artery, DSA: Digital subtraction angiography. A2: A2 portion of anterior cerebral artery, M2: M2 portion of middle cerebral artery.

detached. Two more coils, Optimax Complex Soft ® 4 mm × 7 mm (Balt LLC, USA) and Optima Complex-10 Super Soft® 2 mm × 4 mm (Balt LLC, USA), were inserted into the sac through the jailed microcather-2, which eventually led to complete occlusion of the aneurysm (Raymond-Roy class 1) [Figures 2c and d].

Diffusion-weighted MRI 1 day after the procedure revealed a scattering of asymptomatic small ischemic lesions in the cortical area irrigated by the left MCA. She was discharged 8 days after the procedure without sequela.

DSA performed 3 months after the embolization showed that the aneurysm remained completely occluded [Figures 3a and b].

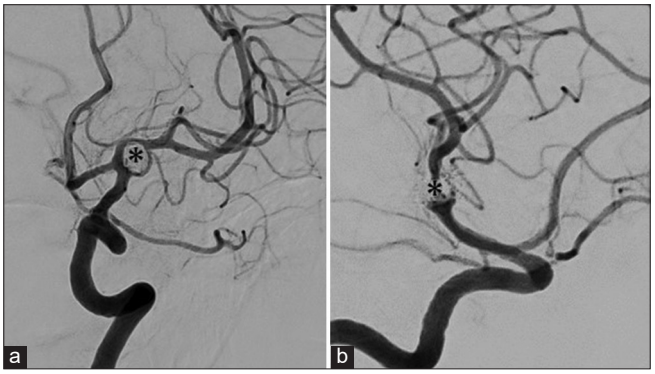

Figure 3: DSA of the left internal carotid artery performed 3 months after the coil embolization. (a) Anteroposterior view, and (b) right-left view. The aneurysmal sac was completely obliterated, demonstrating Raymond-roy occlusion class I. Asterisk: Aneurysmal sac, DSA: Digital subtraction angiography.

LITERATURE REVIEW

So far as we know, there are 11 reported patients, including ours, eight women and three men, with an aneurysm arising from DOMA [Table 1].[3,5,7,9-12,18-20] The age ranged from 42 to 81 years old with a mean of 54.8 ± 13.4 (Standard deviation [SD]) years. Our patient is the youngest. Aneurysm is located on the right side in seven and on the left side in four patients. The reported size of aneurysms in eight patients ranged from 2 to 8 mm, with a mean of 5.2±2.2 (SD) mm. Ours is the largest. Five were ruptured, and six were unruptured aneurysms. The superior limb was generally larger than the inferior limb except for three cases with similar calibers. The aneurysm neck is located at the junction between the inferior limb and ICA in six, including ours, inferior limb per se in one, superior limb per se in two, distal fusion point in one, and not mentioned in one patient. The anterior temporal artery branched from the inferior limb in four and the perforating artery branched from the superior limb in three patients. Five DOMA-associated aneurysms were accompanied by or more intracranial aneurysms.

For the treatment, clipping was performed in eight, coiling was performed in two, and clipping was initially attempted, but eventually, coiling was performed in another patient. Except for two patients with complications after clipping, one with slight dysphasia and another with hemiplegia and aphasia, outcomes of the treatment were generally favorable.

Table 1: Reported intracranial aneurysm associated with duplicate origin of middle cerebral artery (DOMA)

Author, published year	Reported as	Age / Sex	Side	Size of aneurysm	State of aneurysm	Caliber of two limbs	Location of neck	Branching from two limbs	Treatment	Outcome	Reasons for screening or comorbidities
Ueda T, 1984[18]	MCA fenestration	45/M	Rt.	n.m.	unruptured	SL＞IL	n.m.	n.m.	clip	slight dysphasia	multiple intracranial aneurysms
Nussbaum ES, 2009[7]	MCA fenestration	75/F	Rt.	5 mm	ruptured	SL≒IL	DF	ATA from IL	clip	no deficit	headache
Yamaguchi S, 2010[20]	MCA fenestration	81/F	Rt.	small	ruptured	SL＞IL	IL	n.m.	coil	recovered well	ruptured left MCA aneurysm
Rennert J, 2013[9]	MCA duplication	52/F	Lt.	2 mm	unruptured	SL＞IL	IL-ICA	ATA from IL	clip	no deficit	unruptured A-com aneurysm
Tabuchi S, 2014[12]	MCA fenestration	47/F	Rt.	small	ruptured	SL＞IL	IL-ICA	n.m.	clip	good condition	SLE, chronic renal failure
Hirayama K, 2018[3]	DOMA	49/M	Lt.	7 mm	unruptured	SL＞IL	IL-ICA	perforator from SL	clip	no deficit	transient vertigo
Xue X, 2019[19]	MCA fenestration	43/F	Rt.	2.5 mm	ruptured	SL＞IL	IL-ICA	n.m.	clip	no deficit	severe headache
Iwata M, 2020[5]	DOMA	59/M	Rt.	4 mm	unruptured	SL＞IL	IL-ICA	n.m.	clip #→coil	no deficit	left ruptured IC-PC aneurysm
Stojanović N, 2020[11]	segmental duplication of MCA	56/F	Lt.	6 mm	ruptured	SL≒IL	SL	perforator from SL	clip	hemiplegia & aphasia	multiple intracranial aneurysms
Shiozaki E, 2022[10]	segmental duplication of MCA	55/F	Rt.	7 mm	unruptured	SL≒IL	SL	ATA from IL	clip	no deficit	hypertension
Present case, 2024	DOMA	42/F	Lt.	8 mm	unruptured	SL＞IL	IL-ICA	ATA from IL & perforator fom SL	coil	no deficit	transient vertigo

M: male, F: female, MCA: middle cerebral artery, DOMA: duplicate origin of middle cerebral artery, n.m.: not mentioned, SL: superior limb, IL: inferior limb, DF: distal fusion point, IL-ICA: junction between inferior limb and ICA, Rt.: right, Lt. left
#: failed attempt of clipping, ATA: anterior temporal artery, A-com: anterior communicating artery, SLE: systemic lupus erythematosus, IC-PC: internal carotid artery-posterior communicating artery

DISCUSSION

In general, anatomical variations of the MCA are caused by a failure in the formation of a single MCA trunk through the coalescing primitive plexiform arterial network, which typically appears during the 34–36 days of the embryonic stage.[8,17] The failure results in the formation of two main trunks. When the smaller trunk arises distal to the larger MCA trunk, it is designated as an accessory MCA; when the smaller trunk arises proximal to the larger MCA trunk, it is designated as a duplicated MCA.[17] DOMA is speculated to be the result of the distal fusion of accessory MCA or duplicated MCA into the main MCA trunk.[15] Moreover, a failure in the fusion of the plexiform arterial network caused by early branching of the temporal artery may result in MCA fenestration.[2]

In 2020, Hou *et al*. found 67 patients with aneurysms reported to be associated with one of the four types of MCA anomalies, including 20 aneurysms associated with accessory MCA, 34 with duplicated MCA, and 12 with MCA fenestration.[4] By that time, only one aneurysm was reported to be associated with DOMA, according to the review.[4]

Thus, this is a rare case of an aneurysm arising from DOMA. The aneurysm was the largest, 8 mm, among ever-reported patients with such a combination. The size, patient's age, and irregular shape of the two domes urged us to treat this unruptured aneurysm. The authors chose the endovascular coiling technique due to the lack of space between the aneurysm neck and two limbs. With the aid of a protective stent of the parent arteries, complete obliteration of the aneurysm was accomplished. During the attempt of initial framing, the tip of the coil was found to be in the superior limb. The happening meant the tip inadvertently migrated into the inferior limb and went into the superior limb through the distal fusion point of two limbs, verifying the existence of the distal communication of the two limbs.

Our extensive review found 11 reported cases of DOMA-associated aneurysms, including ours.[3,5,7,9-12,18-20] The paucity may be, in part, due to the general lack of awareness of this variation. Actually, out of the 11 patients, five were reported as fenestration in MCA and three as MCA duplication. Among the reported 10 DOMA-associated aneurysms with a description of the neck location, six aneurysms arise from the junction between the inferior limb and ICA. Congenital structural weakness of the arterial wall on this region and/or the hemodynamic stress associated with unusual arterial branching from ICA may explain the preference for aneurysmal origin within the ring of DOMA. Because the number of reported DOMA-associated aneurysms is still very low, the authors cannot determine that the special location of DOMA is the preferential site of aneurysmal formation. It is noteworthy that the review by Hou *et al*. found concurrent cerebrovascular anomalies in 49.2% of the total 67 patients with aneurysms associated with MCA anomalies.[4] Fujimoto *et al*. showed that 13 (38.2%) out of 32 reported aneurysms associated with duplicated MCA had coexisting intracranial aneurysms.[1] Our review of DOMA-associated aneurysms also revealed a high incidence of coexisting intracranial aneurysms, 45.5% (5/9). However, it might be overly simplistic to conclude that these findings suggest a shared congenital background for both aneurysm formation and MCA anomalies. Future large population-based studies on DOMA may reveal the prevalence of aneurysms on it, the preferential site of aneurysmal formation, and possible mechanisms for how this type of aneurysm arises. It is also noteworthy for the safe treatment of this special entity of aneurysm that arterial branches such as the anterior temporal artery and perforating artery toward basal ganglia arise from limbs of DOMA in half of cases. Only three patients, including ours, underwent endovascular treatment.[5,20] The optimal treatment strategy, including the role of stent-assisted coiling for this uncommon combination, should be elucidated through future studies.

CONCLUSION

The authors presented a rare case of an aneurysm associated with DOMA, successfully treated with coil embolization. This report will raise awareness of DOMA, preventing it from being mislabeled as other types of anatomical MCA variations. In the future, larger case series will enhance our understanding of the true nature of this combination and inform appropriate management strategies.

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Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

REFERENCES

1. Fujimoto K, Hashimoto H, Uchiyama Y, Maekawa H, Shida Y, Nakagawa I. Duplicated middle cerebral artery aneurysms treated by coil embolization; a report of two cases and literature review. J Stroke Cerebrovasc Dis 2021;30:105773.
2. Gailloud P, Albayram S, Fasel JH, Beauchamp NJ, Murphy KJ. Angiographic and embryologic considerations in five cases of middle cerebral artery fenestration. AJNR Am J Neuroradiol 2002;23:585-7.
3. Hirayama K, Nakamura H, Yoshimura S, Ozono K, Fukuda Y, Hiu T, *et al*. Unruptured aneurysm arising from the proximal end of the duplicate origin of the middle cerebral artery. No Shinkei Geka 2018;46:901-9.
4. Hou K, Xu K, Liu H, Li G, Yu J. The clinical characteristics and treatment considerations for intracranial aneurysms associated with middle cerebral artery anomalies: A systematic review. Front Neurol 2020;11:564797.
5. Iwata M, Kawaguchi S, Manaka H. A case of unruptured cerebral aneurysm arising from duplicate origin of the middle cerebral artery. No Shinkei Geka 2020;48:515-20.
6. Komiyama M, Nakajima H, Nishikawa M, Yasui T. Middle cerebral artery variations: Duplicated and accessory arteries. AJNR Am J Neuroradiol 1998;19:45-9.
7. Nussbaum ES, Defillo A, Janjua TM, Nussbaum LA. Fenestration of the middle cerebral artery with an associated ruptured aneurysm. J Clin Neurosci 2009;16:845-7.
8. Padget DH. The development of the cranial arteries in the human embryo. Contr Embryol Carneg Instn 1948;32:205-61.
9. Rennert J, Ullrich WO, Schuierer G. A rare case of supraclinoid internal carotid artery (ICA) fenestration in combination with duplication of the middle cerebral artery (MCA) originating from the ICA fenestration and an associated aneurysm. Clin Neuroradiol 2013;23:133-6.
10. Shiozaki E, Kawahara I, Uchida D, Morofuji Y, Ono T, Haraguchi W, *et al*. Unruptured cerebral aneurysms with the segmental duplicated middle cerebral artery formed a fenestrated structure at origin. Surg Neurol Int 2022;13:33.
11. Stojanović NN, Kostić A, Mitić R, Berilažić L. Correlation between multiple cerebral aneurysms and a rare type of segmental duplication of the middle cerebral artery. BMC Neurol 2020;20:3.
12. Tabuchi S, Yoshioka H. Ruptured aneurysm at the fenestration of the middle cerebral artery detected by magnetic resonance angiography in a patient with systemic lupus erythematosus and renal failure: A case report. J Med Case Rep 2014;8:30.
13. Teal JS, Rumbaugh CL, Bergeron RT, Segall HD. Anomalies of the middle cerebral artery: Accessory artery, duplication, and early bifurcation. Am J Roentgenol 1973;118:567-75.
14. Uchino A, editor. Variation of proximal middle cerebral artery (MCA). Atlas of the supraaortic craniocervical arterial variations: MR and CT angiography. Singapore: Springer-Verlag; 2022. p. 99-107.
15. Uchino A, Saito N, Okada Y, Nakajima R. Duplicate origin and fenestration of the middle cerebral artery on MR angiography. Surg Radiol Anat 2012;34:401-4.
16. Uchino A, Takase Y, Nomiyama K, Egashira R, Kudo S. Fenestration of the middle cerebral artery detected by MR angiography. Magn Reson Med Sci 2006;5:51-5.
17. Uchiyama N. Anomalies of the middle cerebral artery. Neurol Med Chir (Tokyo) 2017;57:261-6.
18. Ueda T, Goya T, Kinoshita K, Wakuta Y, Mihara K. Multiple anomalies of cerebral vessels. A case of multiple aneurysms associated with fenestration of the middle cerebral artery and persistent primitive trigeminal artery. No Shinkei Geka 1984;12:531-6.
19. Xue X, Li Y, Yin Y, Yuan X, Xiao H, Wei L. Aneurysms arising from the fenestrated middle cerebral artery. J Craniofac Surg 2019;30:e485-7.
20. Yamaguchi S, Ito O, Suzuki S. Coil embolization of a ruptured aneurysm arising from a middle cerebral artery fenestration--case report. Neurol Med Chir (Tokyo) 2010;50:213-6.

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