Received: 26 August 2023 | Revised: 21 February 2024 | Accepted: 22 February 2024

DOI: 10.1002/ccr3.8605

Clinical Case Reports Open Access WILEY

CASE REPORT

# Rapidly growing distal choroidal artery aneurysm re-rupture following revascularization for hemorrhagic Moyamoya disease: A case report

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**Key Clinical Message**

Intracranial hemorrhage is the leading cause of neurological deficits and poor prognosis in adult patients with Moyamoya disease (MMD). Intracranial hemorrhage is occasionally accompanied by MMD-associated aneurysm and requires additional treatment. To date, direct or indirect bypass surgery or endovascular treatment, such as coil embolization, has been adopted and has achieved successful outcomes. The rapid growth of MMD-associated aneurysms and rebleeding after direct bypass surgery via superficial temporal artery-middle cerebral artery (STA-MCA) anastomosis has rarely been reported. We report a case of a rapidly growing fragile arterial pseudoaneurysm in a patient with MMD. A 45-year-old female was admitted with a headache and decreased mental status. Radiological evaluation, including distal subtraction angiography, revealed intraventricular hemorrhage with a left posterior choroidal artery pseudoaneurysm. Within 4 days after revascularization surgery via STA-MCA direct bypass, the size of the pseudoaneurysm rapidly increased and rebleeding occurred, requiring coil embolization. After endovascular therapy and a second STA-MCA bypass surgery, the patient recovered well and was discharged 8 days later. Follow-up radiological imaging revealed an obliterated pseudoaneurysm without rebleeding or complications. In this case, the rapid growth of an MMD-associated pseudoaneurysm was observed after revascularization surgery because of temporary hemodynamic instability. This report raises questions regarding the causes and management of unstable postbypass hemodynamics.

**KEYWORDS**

aneurysm, endovascular therapy, intraventricular hemorrhage, Moyamoya disease, revascularization

## 1 | INTRODUCTION

Moyamoya disease (MMD) is a chronic and progressive cerebrovascular occlusive disease involving the end of the internal carotid artery (ICA) and the main branches within the circle of Willis (COW).[1,2] In Korea and Japan, more than half of the adult patients with MMD present with hemorrhagic stroke.[2,3] The risk of recurrent hemorrhage in MMD is estimated at 11%–25% within 5 years and 19%–36% within 10 years, with each subsequent hemorrhage increasing the risk of poor neurological outcomes.[4] The incidence of MMD-associated aneurysms is estimated to be 3%–14%, although the frequency of aneurysmal rupture as the cause of hemorrhagic MMD remains uncertain.[5,6] MMD-associated aneurysms can be classified as peripheral aneurysms that originate from collateral vessels or saccular aneurysms that originate from major intracranial arteries.[7]

The treatment strategies for MMD-associated peripheral aneurysms remain controversial because of their deep location, tortuosity, and fragility.[7] Peripheral aneurysms originate mainly from the lenticulostriate, thalamic perforating, or choroidal arteries.[8] Previous reports have described aneurysms treated with close observation, direct clipping, endovascular coiling, or direct revascularization.[6]

Herein, we report the case of a patient with Moyamoya disease who rapidly developed an MMD-associated pseudoaneurysm after direct revascularization. The patient recovered after endovascular coiling and secondary revascularization was performed.

## 2 | CASE HISTORY/ EXAMINATION

A 45-year-old female was referred to our hospital with a headache, nausea, and vomiting, followed by mental changes in the morning. She had been diagnosed with MMD 10 years previously. Initially, noncontrast computed tomography (CT) of the brain revealed a left-dominant intraventricular hemorrhage (IVH) with hydrocephalus, scant subarachnoid hemorrhage along both frontal sulci, and diffuse brain parenchymal swelling (Figure 1A). The patient was referred to the neurosurgery department for emergency cerebrospinal fluid (CSF) diversion, and her consciousness recovered immediately after extraventricular drainage. Digital subtraction angiography (DSA) revealed bilateral MMD and prominent development of choroidal anastomosis (ChA) originating from the left lateral posterior choroidal artery (LPChA). The ChA involved a small pseudoaneurysm, which was considered to be the culprit lesion for bleeding (Figure 1B,C). Postcontrast high-resolution vessel wall image (HR-VWI) demonstrated strong wall enhancement of the pseudoaneurysm, indicating the rupture point of the aneurysm.

## 3 | INVESTIGATIONS AND TREATMENT

A superficial temporal artery-middle cerebral artery (STA-MCA) bypass was performed to reduce the ChA and banish the pseudoaneurysm (Figure 1D). Immediate

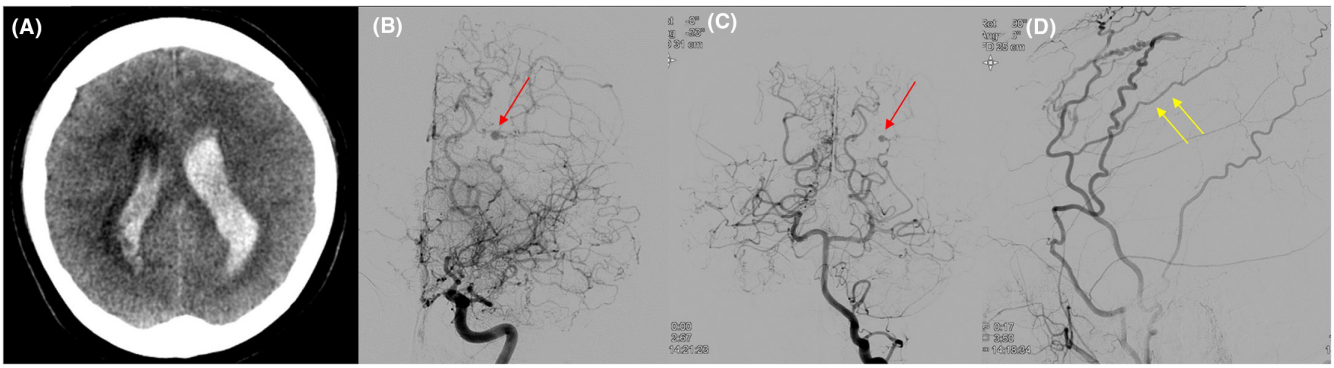


**FIGURE 1** (A–D) Axial view of the initial computed tomography scan (A) showing left-dominant intraventricular hemorrhage and hydrocephalus with a small amount of subarachnoid hemorrhage along both frontal sulci. Diffuse brain swelling is also observed. Left internal carotid artery (ICA) digital subtraction angiography (DSA) (B) with an anteroposterior view showing occlusion of the supraclinoid ICA and basal and leptomeningeal collateral vessels, representing Moyamoya disease. A pseudoaneurysm of approximately 4.5 mm in size can also be seen at the distal branch of the left posterior choroidal artery (red arrow). Left vertebral artery DSA (C) also shows a 4 mm-sized pseudoaneurysm in the left posterior choroidal artery (red arrow). (D) The frontal branch of the superficial temporal artery is selected as the donor artery for the first revascularization surgery (yellow arrow).

postbypass indocyanine green angiography confirmed the patency of the anastomosis; however, extensive brain swelling was observed in the surgical field 20 min after the bypass. The surgical procedure for the bony defects was completed. Postoperative magnetic resonance imaging (MRI) revealed multiple microbleeds, T2 white matter changes in the left frontotemporal area, and external brain herniation at the craniectomy site (Figure 2A). Neurological examination revealed motor aphasia and paresis in the right hand. Postbypass brain single-photon emission computed tomography (SPECT) revealed hyperperfusion in the left MCA territory (Figure 2B). Intensive blood pressure control was achieved with the administration of a hyperosmolar agent to prevent hemorrhage expansion and the progression of hyperperfusion syndrome.

The patient complained of severe headache, nausea, and vomiting postoperatively. Brain CT revealed a left-dominant IVH, suggesting rebleeding from the pseudoaneurysm. The second DSA revealed significant growth of the pseudoaneurysm and nonpatent flow of the bypass (Figure 3A–D). The patient underwent emergency coil embolization for a ruptured pseudoaneurysm under general anesthesia. A 6Fr guiding catheter (DA-XB Envoy, CERENOVUS, Le Locle, Switzerland) was placed into the distal cervical left vertebral artery (V3), and a microcatheter (Excelsior® SL-10® pre-shaped 45, Stryker, Cork, Ireland), advanced over an 0.010 inch microguidewire (Synchro®, Stryker, Salt lake city, USA) into the distal left LPChA. The resulting super-selective angiogram revealed a 13 mm pseudoaneurysm in the distal choroidal portion of the left LPChA and medullary tributaries distal to the aneurysm (Figure 3E). Coil embolization was then carefully conducted under fluoroscopic observation with five coils (*Axium*™ Prime 8 mm × 20 cm, ev3™, Irvine, USA/Target® 360 ULTRA 5 mm × 10 cm, 4.5 mm × 10 cm, 4 mm × 10 cm, Stryker, Cork, Ireland/

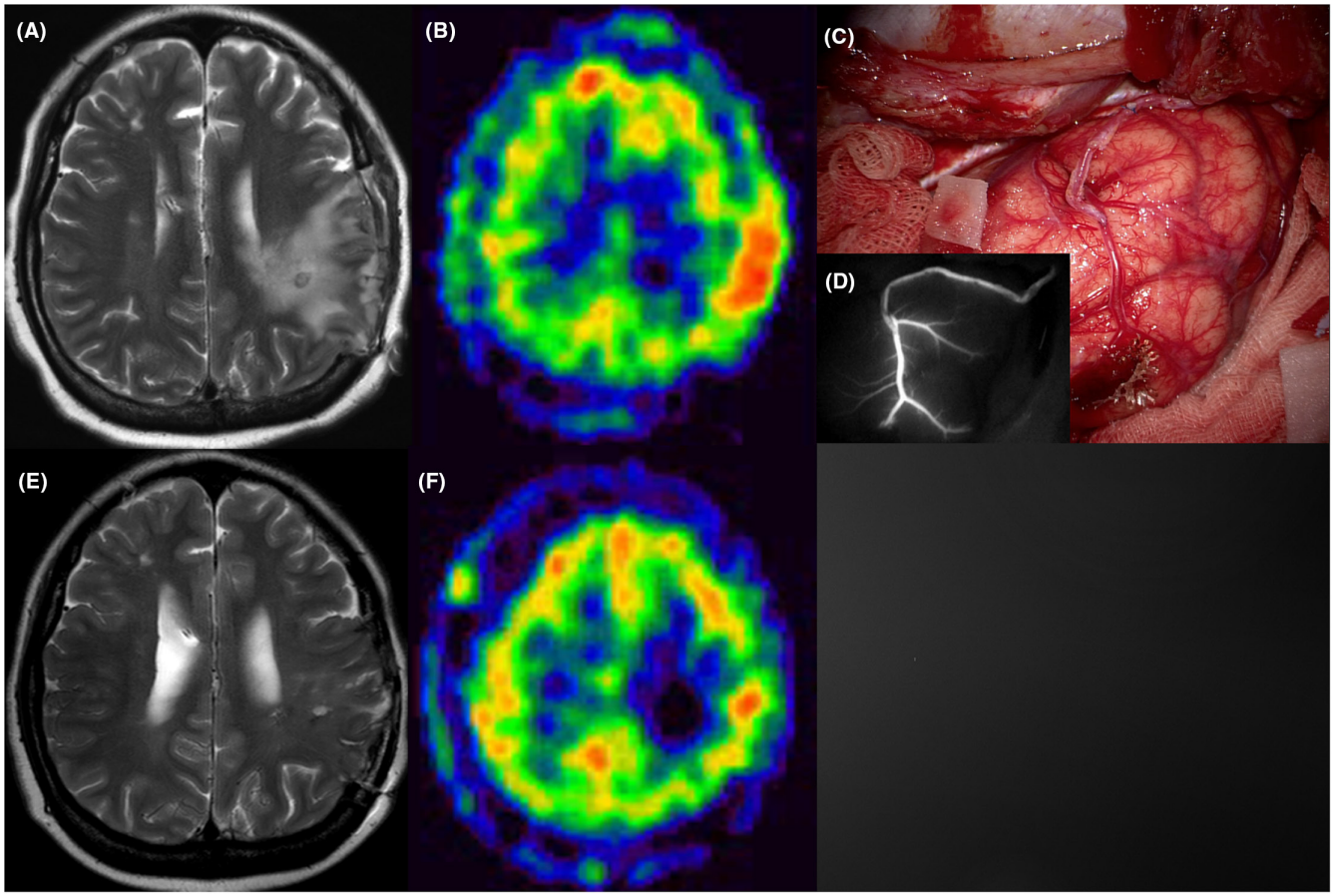


**FIGURE 2** (A–F) T2-weighted brain magnetic resonance imaging (MRI) after the first superficial temporal artery-middle cerebral artery anastomosis bypass (A) showing perilesional edema with herniated brain tissue via the craniectomy site in the left frontoparietal lobe. Basal brain single-photon emission computed tomography (SPECT), (B) showing luxury perfusion in the herniated and swollen left frontotemporal lobe. Immediate postbypass intraoperative findings (C, D) showing severe brain edema and patent anastomosis. T2-weighted image 4 months after the second bypass surgery. Follow-up MRI (E) showing resolution of edema and herniated brain tissue in the left frontoparietal lobe. Five months after the second bypass surgery, follow-up basal brain SPECT (F) reveals recovery of luxury perfusion in the left frontotemporal lobe.

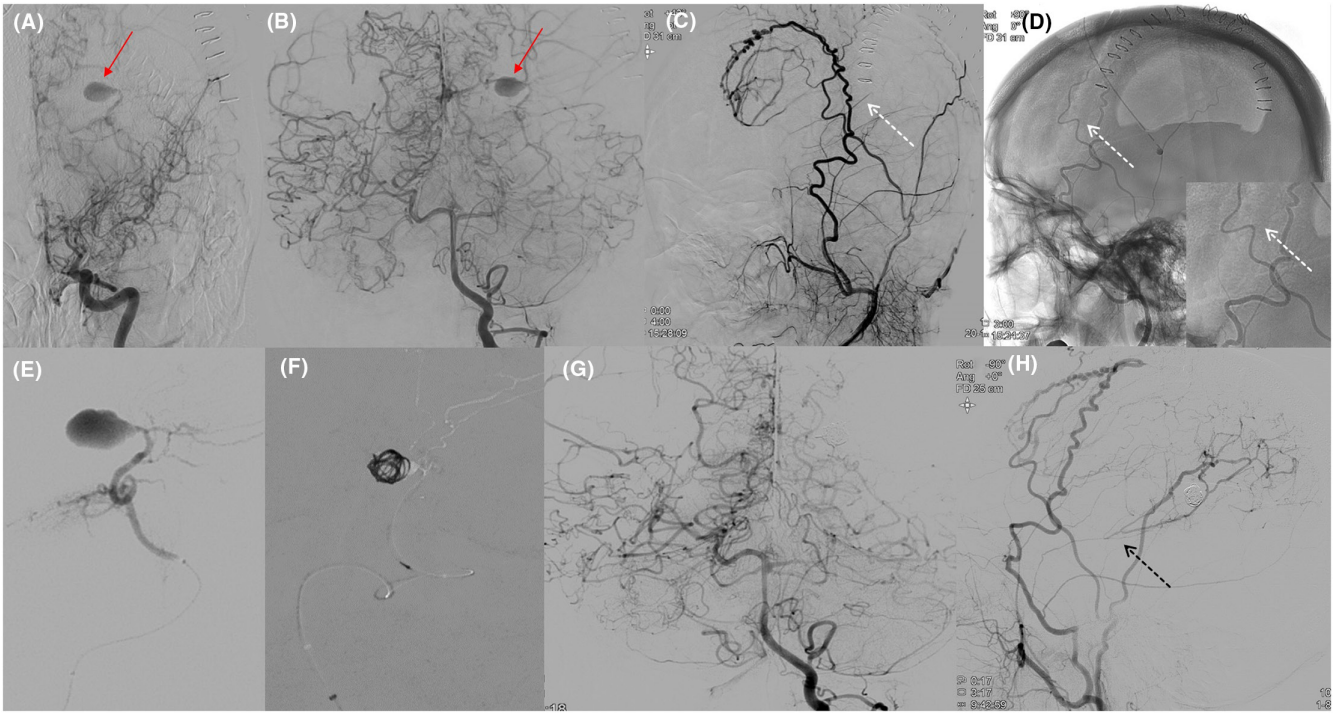


**FIGURE 3** (A–H) Cerebral angiogram (A–D) after a rebleeding attack. Left internal carotid artery (A) and vertebral artery (B) digital subtraction angiography (DSA) showing an increased pseudoaneurysm size compared with preoperative DSA (Figure 1B,C) to approximately 13 mm at the longest diameter (red arrow). Left external carotid artery angiogram (C, D) showing nonpatent superficial temporal artery-middle cerebral artery anastomosis (dotted white arrow) after the first bypass surgery. Selective lateral posterior choroidal angiography and stand-alone coiling of the re-ruptured pseudoaneurysm (E, F) preserving the lateral choroidal anastomosis branches were performed. Postoperative 6-month follow-up angiography (G, H) showing total obliteration of the pseudoaneurysm, disappearance of the medullary tributaries from the left lateral posterior choroidal artery, and patent anastomosis (dotted black arrow).

Target® HELICAL NANO™ 2.5 mm × 4 cm, Stryker, Cork, Ireland). During coil embolization, we attempted to preserve the flow of the medullary tributaries distal to the aneurysm because of the risk of infarction in the LPChA territory (Figure 3F). The patient's immediate postoperative course was uneventful. The patient underwent a second STA-MCA bypass to minimize the medullary tributaries of the ChA (Figure 2C,D), and the postoperative course was uneventful. Follow-up DSA revealed no recanalization of the pseudoaneurysm, disappearance of the ChA, or patent bypass (Figure 3G,H).

## 4 | OUTCOME AND FOLLOW-UP

Strict blood pressure control was achieved, and clinical symptoms gradually improved within a week. The patient was discharged after an additional 2 weeks of conservative management. The right-hand paresis had fully resolved by the 3-month follow-up, but mild aphasia persisted. At the 5-month follow-up, MRI and PET-CT showed normalization of the T2 white matter high-signal lesion and luxury perfusion in the left frontotemporal lobe (Figure 2E,F). At the 6-month follow-up, the patent flow of the bypass was confirmed by cerebral angiography (Figure 3H).

## 5 | DISCUSSION AND CONCLUSION

Peripheral aneurysms, usually originating from distal fragile arteries, including moyamoya vessels, such as the lenticulostriate and thalamoperforating arteries, and the anterior/posterior choroidal arteries, are more likely to disappear.[8] However, these aneurysms have also been disposed to re-rupture in patients with MMD and hemorrhagic manifestations.[9] Patients with IVH and multiple moyamoya collateral vessels often have periventricular collaterals responsible for hemorrhage, especially from the posterior choroidal artery (PChoA) collaterals, called choroidal anastomoses. These choroidal collateral vessels have an outflow tract of the distal cortical M4 branches of the MCA, which is associated with hemodynamic burden after bypass surgery.[10] Revascularization surgery, either direct STA-MCA bypass or indirect encephaloduroarteriosynangiosis, reduces the hemodynamic burden of the

aneurysm's parent artery, distal choroidal artery, and lenticulostriate arteries. Previous studies have suggested that surgically increased bypass blood flow could decrease flow in the parent artery, causing MMD-associated aneurysms to regress spontaneously.[7,11]

However, the current case showed rapid growth of the pseudoaneurysm after direct bypass surgery. There are several reasons for the growth of MMD-associated aneurysms. The patient's brain tissue was already showing signs of inflammation and edema due to posthemorrhagic hydrocephalus at the time of the first surgery.[12] We hypothesized that the swollen brain tissue in the left cerebral hemisphere compresses and diminishes the blood flow to the adjacent cortical M4 branches of the left MCA, obstructing the outflow tract. Consequently, the compensatory ipsilateral distal PChoA blood flow increases, causing the preexisting pseudoaneurysm to grow rapidly. This hypothesis is supported by the reduction in choroidal collaterals after the second direct revascularization surgery, resulting in a new outflow tract via STA-MCA bypass.

Another hypothesis was inferred from Lee's paper.[11] Lee et al. reported rapid growth and regression of a preexisting unruptured pseudoaneurysm in the left choroidal artery after indirect revascularization. They suggested that the reason for the fluctuation in aneurysm size was massive hydration after surgery, which induced hemodynamic instability of the aneurysm's parent artery.[11] In our cases, massive hydration of over 3–6 L of normal saline per day was perfused for 2 days after surgery, and over 3 L of hydration persisted for 3 days, which might have caused hemodynamic instability of the aneurysm. Simultaneously, the patient developed pulmonary edema with bilateral pleural effusion due to fluid overload.

There are two reasons for the selection of endovascular embolization rather than surgical options: (1) our patient had a distal choroidal collateral artery large enough to pass through the microcatheter wire and (2) the amount of intraventricular hemorrhage increased further during rebleeding, even after revascularization surgery. With the choroidal collaterals remaining after postembolization angiography to prevent rebleeding, we decided to conduct a second operation to remove the flow tracts and reduce the choroidal anastomosis. The treatment strategy has not been standardized for posterior choroidal artery aneurysms and should be individualized based on the patient's situation. Multiple treatment options are available including surgical clipping, direct resection, revascularization, and endovascular embolization. Recently, endoscopic clipping was reported to be safe and less invasive in cases of intraventricular aneurysms.[13] Nevertheless, many studies have reported difficulties in treating PChoA pseudoaneurysms because of the deep location, tortuosity, and fragility of the parent vessel, which makes surgical targeting and endovascular superselection of the parent vessel challenging.[14]

From this case report, it is clear that treatment of posterior choroidal pseudoaneurysms can be challenging, and multiple treatment options should be considered to prevent rebleeding. Furthermore, this case demonstrates the importance of maintaining hemodynamic control, including postsurgical cerebral perfusion and hydration, in treating MMD-associated aneurysms. The treatment of MMD-associated aneurysms should be individualized based on the characteristics of the patient's aneurysm and hemodynamic conditions. Further studies and case reports are needed to identify the best combination of treatment options for each type of posterior choroidal aneurysm and for fluid management.

## AUTHOR CONTRIBUTIONS

**Jeong Taek Yoon:** Data curation; resources; writing – original draft; writing – review and editing. **Kyung Mi Lee:** Conceptualization; supervision; writing – review and editing. **Jiwook Ryu:** Conceptualization; data curation; supervision; writing – review and editing. **Ju In Park:** Writing – review and editing.

## ACKNOWLEDGMENTS

None.

## FUNDING INFORMATION

This research did not receive any specific grants from funding agencies in the public, commercial, or not-for-profit sectors.

## CONFLICT OF INTEREST STATEMENT

The authors declare that they have no competing interests.

## DATA AVAILABILITY STATEMENT

Data openly available in a public repository that issues datasets with DOIs.

## ETHICS STATEMENT AND CONSENT TO PARTICIPATE

All procedures performed in the study involving human participants were in accordance with the ethical standards of the institution and/or national research committee and the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. This study was approved by the Institutional Review Board of Kyung Hee University Hospital, and written informed consent was obtained from the patient.

## CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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**How to cite this article:** Yoon JT, Lee KM, Ryu J, Park JI. Rapidly growing distal choroidal artery aneurysm re-rupture following revascularization for hemorrhagic Moyamoya disease: A case report. *Clin Case Rep*. 2024;12:e8605. doi:10.1002/ccr3.8605