Rapidly Growing Distal Choroidal Artery Aneurysm Re-rupture Following Revascularization for Hemorrhagic Moyamoya Disease: A Case Report

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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 five years and 19%-36% within ten 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, which originate from collateral vessels, or saccular aneurysms, which 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 artery, thalamic perforating artery, 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 that rapidly developed an MMD-associated pseudoaneurysm after direct revascularization. The patient recovered after endovascular coiling, and secondary revascularization was performed.

Case presentation

A 45-year-old female was referred to our hospital with headache, nausea, and vomiting, followed by mental changes in the morning. She was diagnosed with MMD ten years previously. Initially, non-contrast 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). For emergency cerebrospinal fluid (CSF) diversion, the patient was referred to the neurosurgery department, and her consciousness recovered immediately after extraventricular drainage. Digital subtraction angiography (DSA) revealed bilateral MMD and the prominent development of a choroidal anastomosis (ChA) originating from the left lateral posterior choroidal artery (LPChA). The ChA involved a small pseudoaneurysm, which was considered the culprit lesion for the bleeding (Figure 1b-c). The postcontrast high-resolution vessel wall image (HR-VWI) demonstrated strong wall enhancement of the pseudoaneurysm, indicating the rupture point of the aneurysm.

A superficial temporal artery-middle cerebral artery (STA-MCA) bypass was performed to reduce the ChA and banish the pseudoaneurysm (Figure 1d). Immediate post-bypass 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 defect was completed. Postoperative magnetic resonance imaging (MRI) revealed multiple microbleeds, T2 white matter changes in the left frontotemporal area, and external brain herniation via the craniectomy site (Figure 2a). A neurological examination revealed motor aphasia and paresis of the right hand. Post-bypass 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 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 non-patent 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 the medullary tributaries distal to the aneurysm (Figure 3e). Coil embolization was then carefully conducted under fluoroscopic observation with five coils (Axium Prime 8 mm x 20 cm, ev3, Irvine, USA / Target(r) 360 ULTRA 5 mm x 10 cm, 4.5 mm x 10 cm, 4 mm x 10 cm, Stryker, Cork, Ireland / Target(r) HELICAL NANO 2.5 mm x 4 cm, Stryker, Cork, Ireland). During coil embolization, we attempted to preserve the flow of the medullary tributaries distal to the aneurysm due to the risk of LPChA territory infarction (Figure 3f). The patient's immediate postoperative period 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, the disappearance of the ChA, or patent bypass (Figure 3g-h). Strict blood pressure control was achieved, and the clinical symptoms gradually improved within a week. The patient was discharged after an additional two-week course of conservative management. The right hand paresis had fully resolved by the three-month follow-up, but mild aphasia persisted. At the four-month follow-up, MRI showed signal intensity normalization of the T2 white matter high-signal lesion (Figure 2e). At the six-month follow-up, patent flow of the bypass was confirmed by cerebral angiography (Figure 3h).

Discussion and conclusion

Peripheral aneurysms, usually originating from distal fragile arteries, which include 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 the hemorrhage, especially from 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, the distal choroidal artery, and the lenticulostriate arteries. Previous studies have suggested that surgically increased bypass blood flow could decrease the 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. We hypothesized that the swollen brain tissue in the left cerebral hemisphere compresses and diminishes the blood flow of 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 the STA-MCA bypass.

Another hypothesis was inferred from Lee's paper [11]. Lee et al. reported the 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, inducing hemodynamic instability of the aneurysm's parent artery [11]. In our cases, massive hydration of over 3 L˜6 L of normal saline per day was perfused for two days after surgery, and over 3 L of hydration persisted for three days, which might have caused hemodynamic instability of the aneurysm. Simultaneously, the patient developed pulmonary edema with bilateral pleural effusion owing 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; 2) The amount of intraventricular hemorrhage increased further during rebleeding, even after revascularization surgery. With the choroidal collaterals remaining after post-embolization angiography to prevent rebleeding, we decided to conduct a second operation that removed the flow tracts and reduced choroidal anastomosis. The treatment strategy has not been standardized for posterior choroidal artery aneurysm cases 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 with intraventricular aneurysms [12]. Nevertheless, many studies have reported difficulty in treating PChoA pseudoaneurysms owing to the deep location, tortuosity, and fragility of the parent vessel, which makes surgical targeting and endovascular superselection of the parent vessel challenging [13].

From this case report, it is clear that the 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, when treating MMD-associated aneurysms. Treatment of MMD-associated aneurysms should be individualized based on the characteristics of the patient's aneurysm and their 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 fluid management.

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Figure legends

Figure 1a-d: Axial view of the initial computed tomography (a) showing left dominant intraventricular hemorrhage and hydrocephalus with a small amount of subarachnoid hemorrhage along both frontal sulci. Diffuse brain swelling can also be seen. Left internal carotid artery (ICA) digital subtraction angiography (DSA) (b) with an anterioposterior view showing occlusion of the supraclinoid ICA and basal and leptomeningeal collateral vessels representing Moyamoya disease. A pseudoaneurysm about 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 at the left posterior choroidal artery (red arrow). (d) The frontal branch of the superficial temporal artery was selected as the donor artery for the first revascularization surgery (yellow arrow).

Figure 2a-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 at 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 post-bypass intraoperative findings (c-d) showing severe brain edema and patent anastomosis. T2 weighted image from four months after the second bypass surgery. Follow-up MRI (e) showing resolved edema and herniated brain tissue at the left frontoparietal lobe. Five months after the second bypass surgery, follow-up basal brain SPECT (f) reveals recovered luxury perfusion at the left frontotemporal lobe.

Figure 3a-h: Cerebral angiogram (Figure a-d) after rebleeding attack. Left internal carotid artery (a) and vertebral artery (b) digital subtraction angiography (DSA) showing an increased size of the pseudoaneurysm compared with preoperative DSA (Figure 1b-c) to about 13 mm at the longest diameter (red arrow). Left external carotid artery angiogram (c-d) showing non-patent superficial temporal artery-middle cerebral artery anastomosis (dotted white arrow) after the first bypass surgery. Selective lateral posterior choroidal angiogram and stand-alone coiling of the re-ruptured pseudoaneurysm (e-f) preserving the lateral choroidal anastomosis branches were performed. Postoperative six-month follow-up angiography (g-h) showing total obliteration of the pseudoaneurysm, the disappearance of the medullary tributaries from the left lateral posterior choroidal artery, and patent anastomosis (dotted black arrow).