

Exacerbation of Major Artery Aneurysms after Revascularization in Patients with Moyamoya Disease: Two Case Reports

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Abstract

The natural history of aneurysms of the major arteries after revascularization in moyamoya disease has yet to be documented. At our institute, we treated two patients with moyamoya disease-associated aneurysms involving major arteries of the posterior cerebral circulation. The aneurysms became enlarged at an early stage after revascularization, necessitating coil embolization. Although cerebral blood flow was improved in the anterior circulation, revascularization for these patients did not decrease hemodynamic stress in the posterior circulation and was not able to attenuate aneurysmal growth. Therefore, patients with moyamoya disease-associated aneurysms of the major arteries should be carefully monitored after revascularization.

Key words: moyamoya disease-associated aneurysm, hemodynamic stress, revascularization, coil embolization

Introduction

Cerebral aneurysms associated with moyamoya disease are generally categorized into two groups: (1) peripheral artery aneurysms occurring in the fragile moyamoya vessels, which is a part of the collateral circulation and (2) major artery aneurysms in the circle of Willis.¹⁾ Surgical revascularization decreases hemodynamic stress in collateral vessels that spontaneously develop in moyamoya disease, thereby helping to obliterate peripheral artery aneurysms.²⁾ However, no studies have focused on the effects of revascularization on aneurysms of the major arteries. In this report, we describe two patients with moyamoya disease-associated aneurysms of the major arteries that exacerbated after revascularization.

Case Reports

I. Case 1

A 48-year-old woman was diagnosed with moyamoya disease after experiencing a transient ischemic attack with left hemiparesis (Fig. 1A–C). Cerebral angiography revealed a saccular aneurysm at the bifurcation of the

basilar artery and superior cerebellar artery (Fig. 1D). Single photon emission computed tomography (SPECT) revealed that mean cerebral blood flow (CBF) at rest and after acetazolamide stress testing was 42.6 mL/100 g/min and 72.9 mL/100 g/min, respectively. Cerebrovascular reactivity (CVR) to acetazolamide was 71.1%. The patient underwent direct revascularization involving anastomosis of the right superficial temporal artery (STA) to the middle cerebral artery (MCA). Seven months after revascularization, cerebral angiography revealed that the aneurysm had become enlarged (Fig. 1E). The angiography also showed that the donor vessels were able to sufficiently supplement circulation to the region supplied by the right MCA and that the posterior circulation was unchanged (Fig. 1–H). SPECT after revascularization revealed that mean CBF at rest and after acetazolamide stress testing was 42.9 mL/100 g/min and 80.9 mL/100 g/min, respectively, and CVR to acetazolamide was 88.6%.

The enlarged aneurysm was an indication for repair, which was carried out by coil embolization under general anesthesia (Fig. 1I).

II. Case 2

A 39-year-old woman developed intraventricular hemorrhage and was diagnosed with moyamoya disease (Fig. 2A–C).

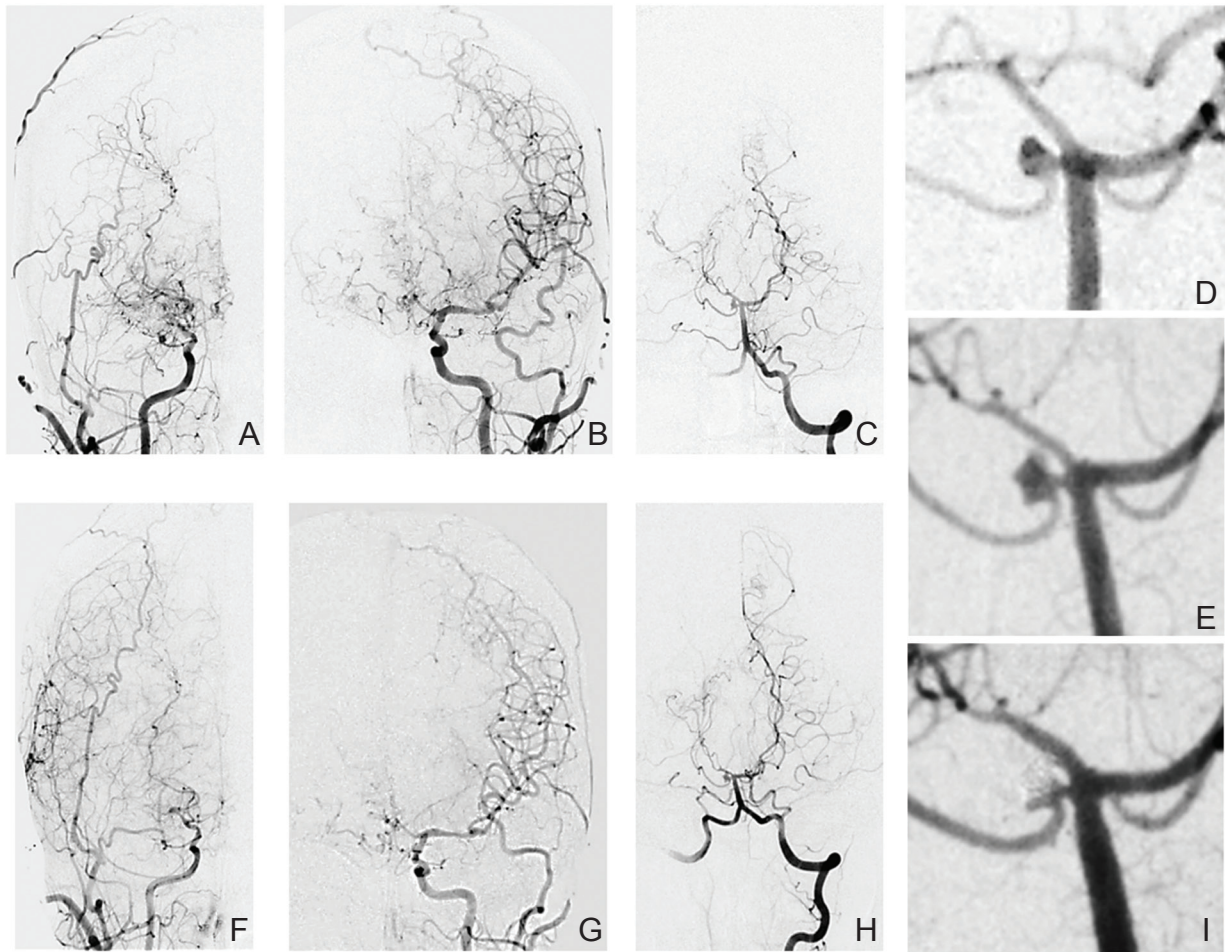


Fig. 1 (Case 1) Cerebral angiography before revascularization (A: right carotid artery, B: left carotid artery, C: vertebral artery) revealing a poorly depicted bilateral anterior cerebral artery (ACA) and right middle cerebral artery (MCA). Significant collateral vessels from the posterior cerebral artery to the territory of the ACA and MCA were found. A saccular aneurysm before revascularization had no bleb (D). The aneurysm after revascularization had multiple blebs (E). Cerebral angiography after revascularization (F: right carotid artery, G: left carotid artery, H: vertebral artery) found blood flow to the MCA territory by vessel anastomosis and unchanged posterior circulation. Coil embolization was performed and the aneurysm was obliterated (I).

Computed tomographic angiography (CTA) revealed peripheral artery aneurysms at the right anterior choroidal artery and a saccular aneurysm at the right P1 portion of the posterior cerebral artery (PCA; Fig. 2D). SPECT revealed that mean CBF at rest was 39.3 mL/100 g/min. SPECT further indicated that CBF was decreased bilaterally in the anterior circulation. The patient underwent direct bilateral revascularization consisting of STA-MCA anastomosis. SPECT performed 8 months later revealed that mean CBF at rest was 50.3 mL/100 g/min and that CBF of the anterior circulation was increased while CBF of the posterior circulation was unchanged. Nine months after revascularization, the patient was admitted for sudden-onset headache. Computed tomography revealed subarachnoid hemorrhage without intraventricular hemorrhage (Fig. 2E). Cerebral angiography and CTA showed that the donor vessels were able to provide sufficient supplemental

blood flow to the region supplied by both MCAs; however, it also revealed that the saccular aneurysm at the right P1 portion had become enlarged and that the peripheral artery aneurysms were unchanged (Fig. 2F–I). Coil embolization was performed under general anesthesia (Fig. 2J, K).

Discussion

In adults, moyamoya disease is occasionally (3–14% of patients) associated with cerebral aneurysms, which are typically classified as peripheral artery aneurysms or major artery aneurysms, regarded as pseudoaneurysms and true aneurysms, respectively.^{3,4} Cerebral aneurysms occur in the posterior circulation in approximately 50–60% of patients with moyamoya disease,⁴ and are most likely to occur here owing to the gradual reduction in perfusion of

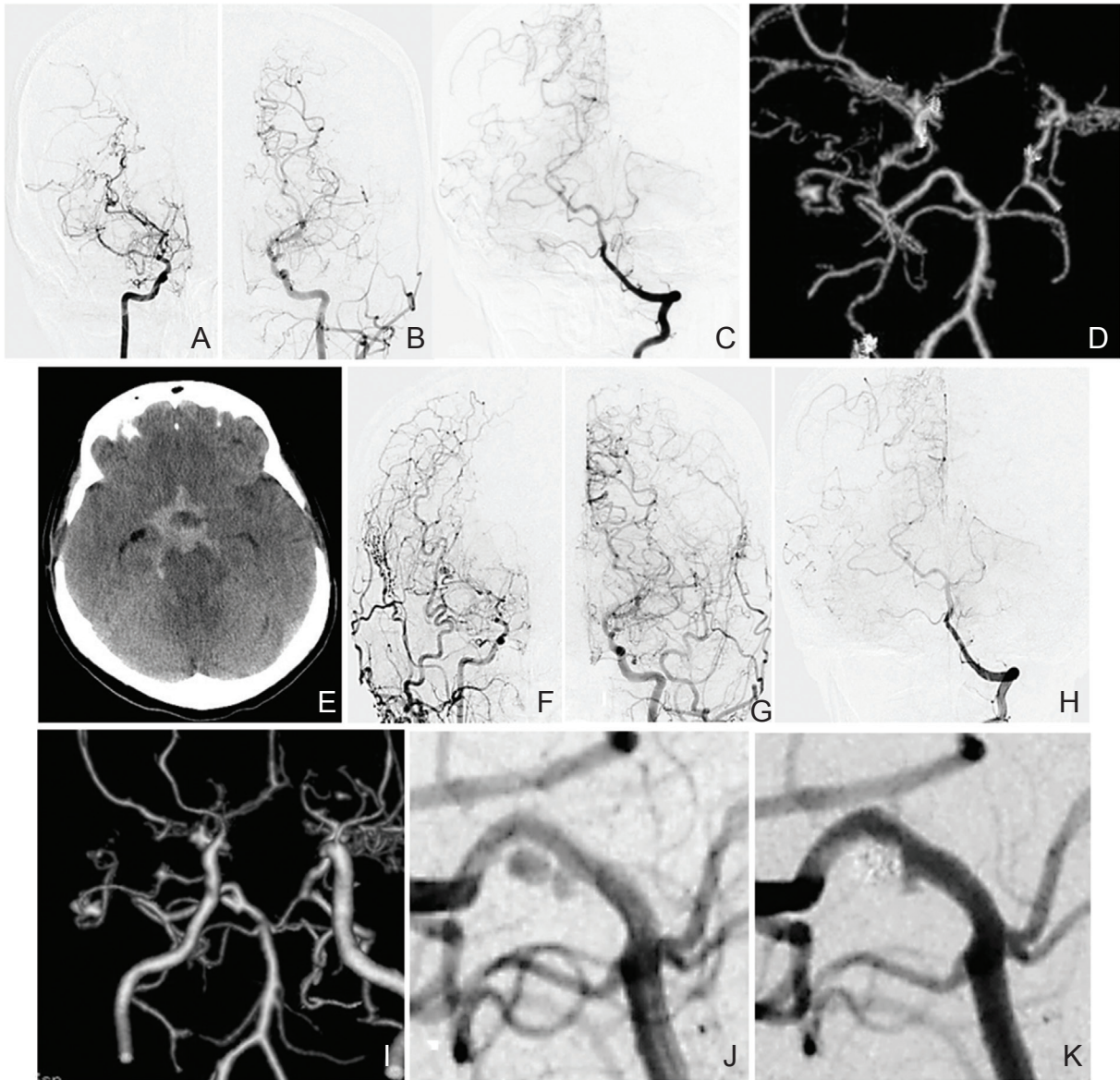


Fig. 2 (Case 2) Cerebral angiography before revascularization (A: right carotid artery, B: left carotid artery, C: vertebral artery) revealing a poorly depicted bilateral anterior cerebral artery (ACA) and middle cerebral artery (MCA). Significant collateral vessels from the posterior cerebral artery to the territory of the ACA and MCA were found. Computed tomography upon initial admission revealed multiple aneurysms (posterior cerebral artery and posterior choroidal artery) (D). Computed tomography revealed a subarachnoid hemorrhage (E). Cerebral angiography after revascularization (F: right carotid artery, G: left carotid artery, H: vertebral artery) revealed blood flow in the territory of the MCA by bilateral vessel anastomosis and unchanged posterior circulation. The aneurysm of the posterior cerebral artery was enlarged and the aneurysm of the posterior choroidal artery was unchanged (I, J). Coil embolization was performed and the aneurysm was obliterated (K).

the internal carotid artery system (due to chronic stenosis and occlusion in the anterior circulation), which increases hemodynamic stress in the posterior circulation.⁵⁾

Cerebral aneurysms can also be related to certain vascular diseases, such as arteriovenous malformation and occlusion of the internal carotid artery.^{6,7)} The formation and regression of flow-related aneurysms is associated with hemodynamic changes in blood flow. Therefore, the

flow-related aneurysm can regress spontaneously subsequent to the treatment of vascular diseases. In moyamoya disease, surgical revascularization has the potential to improve cerebral circulation and decrease hemodynamic stress in collateral vessels, thereby helping to obliterate peripheral artery aneurysms.²⁾ However, the natural history of aneurysms of the major arteries after revascularization is not known. It has previously been reported that the

posterior circulation is involved in 30–58% of patients with moyamoya disease.^{8,9} Further, progressive steno-occlusive changes in the PCA after revascularization were noted in 47.4% of patients with moyamoya disease.¹⁰ The mean period to progressive steno-occlusive changes in the PCA was 18.4 months in patients who underwent revascularization compared with 43.7 months in patients who did not.^{8,10} This finding suggests that revascularization might accelerate these changes and hemodynamic stress in the posterior circulation generally decreases over a relatively short period of time after revascularization. Possible mechanisms of progressive steno-occlusive changes in the PCA after revascularization include the following: hemodynamic stress on the vascular endothelium, regional heterogeneity of sympathetic innervation of the intracranial arterioles, and reduced collateral blood supply due to reduced demand.^{10,11} Patients with progressive steno-occlusive changes in the PCA are characterized by decreased leptomeningeal collateral flow and decreased CBF in the posterior circulation.¹⁰ In patients with aneurysms of the major arteries in the posterior circulation, decrease of hemodynamic stress and progressive steno-occlusive changes in the posterior circulation could prompt the size of the aneurysms to stabilize.

Hemodynamic stress in the posterior circulation generally decreases over a relatively short period of time after revascularization. Patients in this study did not exhibit progressive steno-occlusive changes in the posterior circulation, although both patients displayed improved CVR or CBF at rest. Revascularization for these patients would not decrease hemodynamic stress in the posterior circulation, and aneurysmal growth would continue. Therefore, moyamoya disease-associated aneurysms of the major arteries should be monitored carefully in patients in whom the hemodynamic stress of the posterior circulation does not decrease after revascularization because the aneurysms may exacerbate.

Cerebral aneurysms with moyamoya disease are characterized by abnormally developed blood vessels and the presence of collateral circulation, which makes direct surgical clipping difficult and causes high disability rates.¹² We used coil embolization to treat the aneurysms because, unlike surgical clipping, it does not interfere with collateral flow. Coil embolization for moyamoya disease-associated aneurysms is regarded safe and effective.^{1,13}

Conflicts of Interest Disclosure

The authors declare that they do not have professional or financial affiliations that may have biased the results.

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