Ruptured Intracranial Aneurysms Associated With Moyamoya Disease
—Three Case Reports—

Yoshikazu ARAI,1 Ken MATSUDA,1 Makoto ISOZAKI,1
Tsuyoshi NAKAJIMA,1 and Ken-ichiro KIKUTA1

1Department of Neurosurgery, Faculty of Medical Sciences, University of Fukui, Fukui

Abstract

Three cases of ruptured intracranial aneurysm associated with moyamoya disease are presented. Endovascular treatments were performed successfully in two patients with major artery aneurysms. One patient with a collateral aneurysm was managed conservatively and follow-up angiography 1 year later demonstrated spontaneous disappearance of the aneurysm. Our experience suggests that although aneurysms associated with moyamoya disease show differences in evolution and location, endovascular treatment of major artery aneurysms is safe and effective, and peripheral aneurysms which cannot be directly accessed for surgery or endovascular embolization may be treated conservatively.

Key words: subarachnoid hemorrhage, moyamoya disease, intracranial aneurysm, endovascular embolization, Guglielmi detachable coil

Introduction

Moyamoya disease is a slowly progressive occlusive disease of the major vessels involving the internal carotid arteries, and is associated with intracranial aneurysms in 3–15% of cases.2–5 Although direct surgical clipping has been advocated for the treatment of such aneurysms,2,4,6,12 such procedures are difficult and hazardous. Dural and arachnoid incisions may disturb collateral flow through anastomotic vessels and intraoperative compression of ischemic cerebral cortex or temporary clipping of the parent artery may result in irreversible brain damage. In recent years, endovascular treatments have been used to treat such directly challenging aneurysms.7,10 We describe three cases of ruptured aneurysm associated with moyamoya disease presenting with subarachnoid hemorrhage (SAH). Endovascular treatment was performed for two cases, while the remaining case was treated conservatively.

Case Reports

Case 1: A 73-year-old woman, who had experienced right putaminal hemorrhage with left hemiparesis 4 years earlier and was diagnosed with moyamoya disease at that time, presented with severe headache and vomiting, and was transferred to our hospital from the local hospital. Computed tomography (CT) demonstrated SAH and old post-putaminal hemorrhage (Fig. 1A). Cerebral angiography revealed evidence of moyamoya disease with abnormal

Fig. 1 Case 1. A: Axial computed tomography scan at presentation demonstrating subarachnoid hemorrhage in the prepontine cistern and left sylvian fissure, and old post-putaminal hemorrhage on the right. B, C: Bilateral internal carotid angiograms, anteroposterior view, showing occlusion at the origin of the right internal carotid artery and stenosis at the proximal segment of the left anterior cerebral artery. D, E: Left vertebral angiograms, anteroposterior view, showing a saccular aneurysm at the tip of the basilar artery (D) and complete occlusion of the aneurysm after embolization (E).
moyamoya vessels (Fig. 1B, C). In addition, a saccular aneurysm (diameter 3 mm × 2.5 mm) was observed at the tip of the basilar artery (Fig. 1D) and was thought to be the source of the SAH. Endovascular embolization was performed using Guglielmi detachable coils (GDCs), and the aneurysm was completely occluded (Fig. 1E). The patient was subsequently discharged with only the pre-existing left hemiparesis.

Case 2: A 54-year-old woman experienced right thalamic hemorrhage and was diagnosed with moyamoya disease. Cerebral angiography showed a saccular aneurysm (diameter 3 mm × 5.5 mm) at the left P2 portion of the posterior cerebral artery (PCA). One year later, she presented with sudden onset of severe headache and vomiting. CT revealed SAH around the brainstem, particularly at the left ambient cistern (Fig. 2A). Cerebral angiography found evidence of moyamoya disease (Fig. 2B, C) and no aneurysms except the previous PCA aneurysm, which showed no changes in size (Fig. 2D). This aneurysm was considered the cause of SAH and was completely embolized using GDCs with preservation of the PCA (Fig. 2E). The patient was subsequently discharged in a neurologically intact condition. No further bleeding occurred during the follow-up period (45 months).

Case 3: A 58-year-old man experienced right thalamic hemorrhage 12 years earlier and was diagnosed with moyamoya disease. He presented to our hospital 3 days after suffering sudden onset of severe headache. CT revealed a small amount of SAH in the right Sylvian fissure (Fig. 3A). Cerebral angiography demonstrated bilateral occlusion of the internal carotid artery at its terminal with abnormal moyamoya vessels (Fig. 3B, C) and a 4 mm × 3.5 mm aneurysm arising from a prominent posterior thalamoperforating artery at the tip of the basilar artery (Fig. 3D). We considered this as the cause of SAH and attempted endovascular embolization, but were unable to introduce a microcatheter into the perforator because of severe stenosis of the origin. The patient was therefore followed up conservatively. This aneurysm had disappeared without re-rupture at follow-up examination 1 year later (Fig. 3E).

Discussion

Cerebral aneurysms associated with moyamoya disease can be generally classified into peripheral artery aneurysms in the fragile moyamoya vessels or collateral circulation, and major artery aneurysms in the circle of Willis.\(^1,3-5,8,10\) The former are considered to be pseudoaneurysms, with rupture frequently causing intraventricular or intracerebral hemorrhage. However, these pseudoaneurysms can gradually reduce in size and may completely disappear.\(^1,3\) The latter are true aneurysms, occurring in the posterior circulation in about 50–60% of cases associated with moyamoya disease.\(^3,6\) Increased hemodynamic stress in the posterior circulation due to oc-
clusion or stenosis of the internal carotid artery may contribute to the development of this type of aneurysm, and the resultant increase in blood flow often causes enlargement and rupture of these aneurysms with time.

Spontaneous disappearance of true aneurysms is extremely rare. Most authors have advocated clipping of a saccular aneurysm in the circle of Willis and major vessels in these patients. However, in many cases, clipping had to be abandoned because of technical difficulties due to the abundance of collateral vessels in the operative field, or outcomes have not been good due to postoperative stroke. Endovascular therapy with GDCs represents a good option for cases that are difficult to treat by direct surgery. In our Cases 1 and 2, endovascular embolization was successfully performed, and no procedure-related complications were encountered. In Case 3, we attempted endovascular embolization for the aneurysm, but were unable to introduce a microcatheter into the perforator. Endovascular occlusion of aneurysms on collateral vessels using glue has recently been reported for patients with moyamoya disease, and indicated that, if a fragile aneurysm on collateral vessels was considered as the cause of hemorrhage, then aggressive treatment by endovascular n-butyl-cyanoacrylate injection is warranted. However, this treatment remains controversial, as the safety of occluding the parent artery of the aneurysm and its branches is unclear. Conservative management may sometimes represent a treatment option. Several reports have described spontaneous disappearance of small aneurysms on collateral vessels in patients who did not exhibit repeated intracranial bleeding, such as in our Case 3. However, this phenomenon does not always happen and careful follow up is needed. Several authors have proposed surgical revascularization, which could reduce preexisting collateral vessels or hemodynamic stress on those vessels, and may reduce hemorrhagic risk.

Our present experience suggests that although aneurysms associated with moyamoya disease show differences in evolution and location, endovascular treatment for major artery aneurysms is safe and effective. However, the vertebrobasilar system is the only source of blood supply for the brain in most patients with moyamoya disease. Parent artery occlusion due to coil migration, mechanical spasm, thrombo-embolism, or dissection during catheter manipulation would thus cause serious problems. Therefore, endovascular treatment needs to be performed by skilled interventionists, and long-term follow up with cerebral angiography is necessary because of the higher recurrence rate following embolization compared to complete clipping.

References


Address reprint requests to: Yoshikazu Arai, MD, PhD, Department of Neurosurgery, Faculty of Medical Sciences, University of Fukui, 23–3 Matsukashimaizuki, Elheiji-cho, Yoshida-gun, Fukui 910–1193, Japan.

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