

Case Report

Long-term effect of surgical revascularization on silent microbleeds in adult moyamoya disease: A case report

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Abstract

Background: Recent development of magnetic resonance (MR) imaging has shown that silent microbleeds can be observed in a certain subgroup of adult patients with moyamoya disease. The patients with microbleeds are at higher risk for hemorrhagic stroke. However, the beneficial effects of surgical revascularization have not been established in asymptomatic patients with moyamoya disease. The authors present a case that underwent surgical revascularization for asymptomatic moyamoya disease because the number of silent microbleeds increases on serial MR examinations.

Case Description: A 61-year-old female was referred to our hospital because of nonspecific headache. T2-weighted MR imaging revealed silent microbleeds in the corpus callosum. She was diagnosed as moyamoya disease on cerebral angiography. She was conservatively followed up, however, *de novo* microbleeds developed in the right temporal and frontal lobes on follow-up MR imaging 6 months later. Superficial temporal artery to middle cerebral artery (STA-MCA) anastomosis and indirect bypass were performed on the right side to prevent hemorrhagic stroke. Postoperative course was uneventful. Follow-up cerebral angiography performed 10 months after surgery showed a marked development of surgical collateral through both direct and indirect bypass and the diminishment of moyamoya vessels. For the last 7 years after surgery, she is free from any cerebrovascular events, and serial MR examinations revealed no further development of *de novo* microbleeds.

Conclusions: Surgical revascularization may be useful to reduce the moyamoya vessels and prevent cerebrovascular events in a certain subgroup of patients with asymptomatic moyamoya disease, although its universal benefits on asymptomatic moyamoya disease have not been established yet.

Key Words: Moyamoya disease, prognosis, silent microbleeds, surgical revascularization

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BACKGROUND

Moyamoya disease is a unique cerebrovascular disorder characterized by progressive occlusion in the terminal portion of internal carotid artery (ICA) and its main branches, anterior and middle cerebral arteries (ACA and MCA, respectively). The perforating arteries are markedly dilated to supply collateral blood flow to the ischemic

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brain, called as moyamoya vessels. Pediatric patients usually develop transient ischemic attack (TIA) or ischemic stroke, whereas adult patients also develop hemorrhagic stroke.^[9,13] Main cause of hemorrhagic stroke is known as a longstanding hemodynamic stress on the dilated, fragile moyamoya vessels. Very recently, a randomized clinical trial demonstrated that direct or combined surgical revascularization can significantly reduce the risk of recurrent hemorrhagic stroke in adult patients with moyamoya disease.^[12,14] However, the beneficial effects of surgical revascularization have not been established in asymptomatic patients with moyamoya disease.^[7,8]

On the other hand, recent development of magnetic resonance imaging (MRI) has shown that silent microbleeds can be observed in a certain subgroup of adult patients with moyamoya disease. Silent microbleeds are identified more frequently in patients with moyamoya disease than in healthy individuals. They are found in the basal ganglia, thalamus, and periventricular region where intracranial bleeding often occurs in moyamoya disease.^[1,3-6,11] They are found in ischemic-type patients, hemorrhagic-type patients, and even asymptomatic patients.^[11] Kikuta *et al.* (2007) resected the microbleed lesion during surgery and proved that the lesion consisted of several fragile arteries and the surrounding microbleed.^[5] More importantly, the patients with single or multiple microbleeds are at significantly higher risk for subsequent hemorrhagic stroke.^[6,11]

Here, the authors present a 61-year-old female who underwent surgical revascularization for asymptomatic moyamoya disease because serial MRI demonstrated an increase in the number of silent cerebral microbleeds. For the last 7 years, she has been free from both *de novo* microbleeds and any cerebrovascular events.

CASE DESCRIPTION

A 61-year-old female complained of chronic, nonspecific headache and was referred to our hospital. She had no past history and no family history of moyamoya disease. Neurological and laboratory examinations revealed no definite abnormalities. Although T1- and T2-weighted MRI demonstrated no lesion, T2-weighted MRI revealed a microbleed in the genu of corpus callosum [Figure 1a]. MRA showed the occlusion of left ICA and severe stenosis of the terminal portion of right ICA and its branches. On cerebral angiography, the left ICA was completely occluded at the cervical portion (Suzuki's angiographical stage 6). The terminal portion of the right ICA and its main branches were severely stenotic, associated with basal moyamoya vessels (stage 3; Figure 1b and c). ¹⁵O-gas positron emission tomography (PET) detected no hemodynamic insufficiency. Based on these findings, she was conservatively followed up at outpatient clinic.

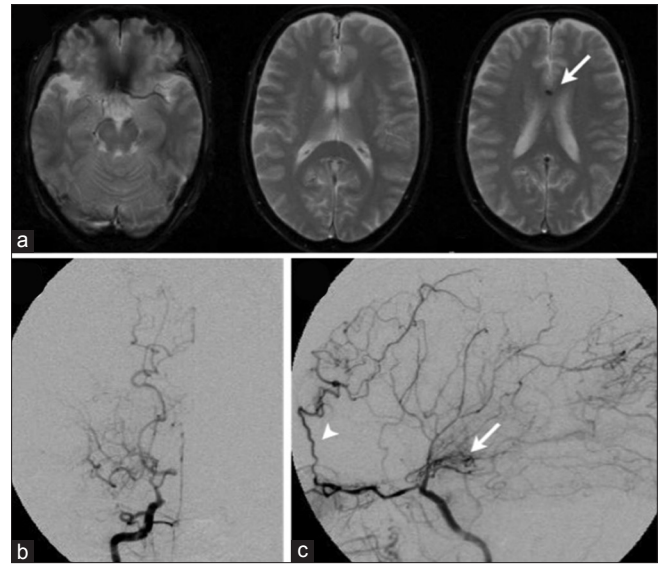


Figure 1: T2-weighted MRI at initial presentation (a) showed silent microbleed in the genu of corpus callosum (arrow). AP (b) and lateral views (c) of preoperative right internal carotid angiography showed severe stenosis in the terminal portion of the right internal carotid artery (ICA) associated with the development of basal moyamoya vessels (arrow). Note the development of collateral pathway to the anterior cerebral artery through the ophthalmic artery

Follow-up MRI was obtained 6 months later. Although she developed no cerebrovascular episodes, T2-weighted MRI revealed two *de novo* microbleeds in the right deep temporal lobe adjacent to the temporal horn and the right frontal lobe [Figure 2]. She was considered at very high risk for subsequent hemorrhagic stroke and underwent superficial temporal artery to middle cerebral artery (STA-MCA) anastomosis and encephalo-duro-myo-arterio-pericranial synangiosis (EDMAPS) on the right side.^[10] Informed consent concerning surgical treatment was obtained from the patient herself and her family. Surgery was performed in supine position under general anesthesia. As described before, the frontal and parietal branches of the STA were dissected from the scalp under a surgical microscope. The temporal muscle and frontal pericranium were dissected as the vascularized flaps for the indirect bypass. To perform large frontotemporal craniotomy, five burr holes were made. The burr hole at the center of craniotomy site was made rostral to the pterion to preserve the anterior branch of the middle meningeal artery (MMA) because it is known to pierce the bony tunnel of the middle meningeal groove just beneath the junction of the sphenoparietal, sphenosquamosal, and squamosal sutures. A heart-shaped craniotomy was performed, preserving the lesser wing of the sphenoid bone. Then, the lesser wing was carefully resected, preserving the anterior branch of the MMA, using a high-speed drill. The dura was incised and rolled back, preserving the main branches of the MMA. The frontal branch of the STA was anastomosed

to the prefrontal artery, and the parietal branch of the STA was anastomosed to the middle temporal artery in an end-to-side fashion with 10-0 nylon threads. The patency of the anterior branch of the MMA was confirmed using indocyanine green (ICG) videoangiography during surgery. The anastomosed STA grafts were attached onto the brain surface as long as possible, which can induce indirect angiogenesis between them. The dural flaps were turned into the epiarachnoid space. Then, the brain surface was covered, using the temporal muscle and frontal pericranium. As a result, the right temporal lobe with silent microbleeds was surgically revascularized with both direct and indirect bypass procedures. Finally, cranioplasty was performed and the wound was closed.^[10] Postoperative course was uneventful. Follow-up cerebral angiography was performed 10 months after the surgery. Right common carotid angiogram demonstrated well-developed surgical collaterals through both direct and indirect bypass and significant diminishment of moyamoya vessels [Figure 3a and b]. She has been free from any cerebrovascular events, and serial MR examinations have shown no *de novo* microbleeds for the last 7 years after surgery [Figure 4].

DISCUSSION

The present case was considered at very high risk for hemorrhagic stroke in near future and underwent surgical revascularization without any cerebrovascular episodes. There are several reasons for this exceptional decision. First, initial T2-weighted MRI identified a silent microbleed in the corpus callosum. The annual risk of hemorrhagic stroke is significantly higher in adult patients with silent microbleeds at initial presentation than those without (6.6% and 0%, respectively).^[11] Second, follow-up T2-weighted MRI detected two *de novo* microbleeds apart from original one at 6 months after initial presentation. Kikuta *et al.* (2007) reported that the patients with multiple microbleeds are at a significantly higher risk for hemorrhagic stroke than either those with single microbleed or those without ($P = 0.0380$).^[6] Third, the microbleeds themselves are a sign of bleeding from the fragile perforating arteries in moyamoya disease, suggesting the presence of long-standing hemodynamic stress in moyamoya vessels.^[5] Finally, the annual risk of ischemic or hemorrhagic stroke is 3.2%, being never low even in asymptomatic moyamoya disease.^[8] In this case, therefore, surgical revascularization was indicated to reduce the risk of hemorrhagic stroke by relieving long-standing hemodynamic stress in the dilated moyamoya vessels. Indeed, there were no cerebrovascular events in this case, including hemorrhagic stroke for the last 7 years after surgery. The moyamoya vessels have diminished on postoperative cerebral angiography, and serial MR examinations also detect any *de novo* microbleeds after surgery.

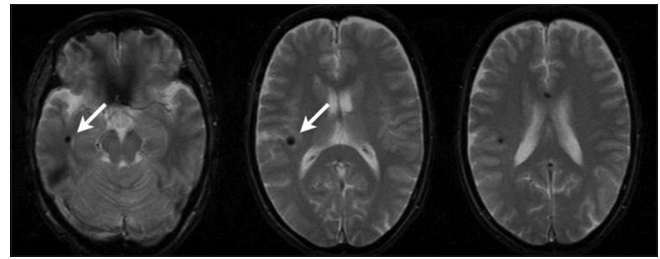


Figure 2: T2-weighted MRI taken 6 months later demonstrated two *de novo* microbleeds in the right deep temporal lobe adjacent to the temporal horn and the right frontal lobe (arrows)

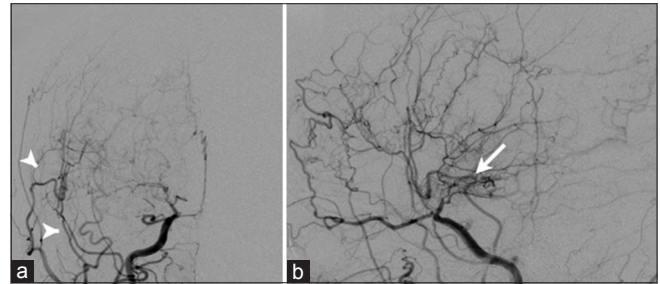


Figure 3: AP (a) and lateral views (b) of postoperative right common carotid angiography showed well developed collaterals through the external carotid system. Note a marked diminishment of basal moyamoya vessels after surgery

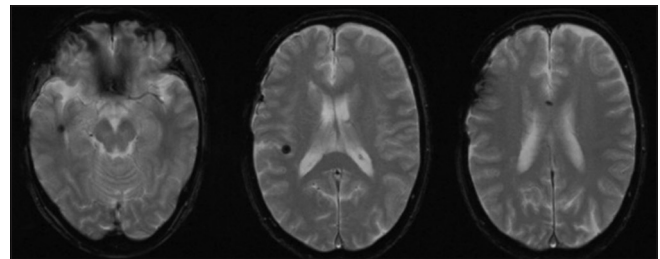


Figure 4: T2-weighted MRI taken 7 years after right superficial temporal artery to middle cerebral artery anastomosis and indirect bypass showed no newly developed microbleeds

Surgical revascularization is widely known to be useful in reducing the risk of both ischemic and hemorrhagic stroke.^[9,12] However, beneficial effects of surgical revascularization on asymptomatic moyamoya disease are not established yet. A nationwide observational study in Japan enrolled 40 asymptomatic patients with moyamoya disease and followed them for a mean of 43.7 months. Of the 34 non-surgically treated patients, 4 experienced cerebrovascular events, including ischemic stroke in one patient and hemorrhagic stroke in three patients. Therefore, the annual risk of any stroke was 3.2%. On the other hand, 6 other patients underwent direct or combined surgical revascularization and experienced no cerebrovascular events during follow-up. However, this observational study could not reach any conclusion about the benefits of surgical revascularization on asymptomatic moyamoya disease because of small sample size and a nonrandomized study design. Subsequently,

Kawai *et al.* (2010) reported two asymptomatic patients whose disease stage rapidly progressed during follow-up. Their cerebral blood flow study revealed a deterioration of cerebral hemodynamics in the involved hemisphere, although they were still asymptomatic. They successfully underwent combined surgical revascularization to reduce the risk of subsequent ischemic stroke.^[2] Therefore, further studies would be warranted to establish the treatment strategy for asymptomatic moyamoya disease.

CONCLUSION

The authors report an adult case with asymptomatic moyamoya disease that successfully underwent combined surgical revascularization because follow-up T2-weighted MRI identified *de novo* microbleeds. She has reported no cerebrovascular events in the last 7 years after surgery. The benefits of surgical revascularization on asymptomatic moyamoya disease should carefully be evaluated in the near future.

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Conflicts of interest

There are no conflicts of interest.

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