Moyamoya Syndrome Associated With Gamma Knife Surgery for Cerebral Arteriovenous Malformation

—Case Report—

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Abstract

A 30-year-old female developed moyamoya syndrome after gamma knife surgery (GKS) for cerebral arteriovenous malformation (AVM), and was treated with bypass surgery. She suffered from fluttering scotoma, right transient hemianopsia, and headache for 1 year. Cerebral angiography revealed a Spetzler-Martin grade III AVM located in the left occipital lobe. After staged embolization, GKS was performed with a minimum dose of 20 Gy to the periphery of the nidus at the 50% isodose level of the maximum target dose. Gradual nidus regression was achieved, and the clinical symptoms disappeared completely. However, at 30 months after GKS, the patient suffered transient ischemic attack. Cerebral angiography showed left middle cerebral artery occlusion with moyamoya vessels. The patient underwent direct and indirect bypass surgery. After surgery, the patient was free from ischemic symptoms. Chronic inflammation and long-term changes in expression of cytokines and growth factors after GKS may have triggered this case.

Key words: moyamoya syndrome, gamma knife surgery, arteriovenous malformation, revascularization

Introduction

Gamma knife surgery (GKS) is an important therapeutic modality for cerebral arteriovenous malformation (AVM).^{7,9)} Progressive intracranial artery occlusion due to radiation-induced vasculopathy is a serious side effect of conventional radiation therapy for intracranial disease,^{2,8,12)} but moyamoya syndrome is very rare after GKS.¹⁷⁾ Here we report a case of moyamoya syndrome associated with GKS performed for AVM, which was treated by bypass surgery.

Case Presentation

A 30-year-old female suffered from fluttering scotoma, right transient hemianopsia, and headache for 1 year. She was admitted to our institution. Cerebral angiography revealed a Spetzler-Martin grade III AVM located in the left occipital lobe and splenium of the corpus callosum (Fig. 1A, B). Staged embolization of the left posterior cerebral artery and anterior cerebral artery was performed. The nidus was reduced by 30% (Fig. 1C, D). Angiography before

and after the embolization detected no focal stenosis of the intracranial vessels. After embolization, GKS was performed with a minimum dose of 20 Gy to the periphery of the AVM nidus at the 50% isodose level of the maximum target dose (Fig. 2A). The nidus volume was estimated as 16.6 ml. The right internal carotid artery (ICA) terminal, right middle cerebral artery (MCA) M1 segment, left ICA terminal, and left MCA M1 segment received 1.4, 1.0, 2.1, and 1.9 Gy, respectively. Gradual nidus regression was observed on serial follow-up brain magnetic resonance (MR) imaging and cerebral angiography. Her clinical symptoms disappeared completely. However, at 30 months after GKS, the patient began to suffer from transient right hemiparesis.

Technetium-99m-ethylcysteinate dimer (^{99m}Tc-ECD) single photon emission computed tomography (SPECT) at rest demonstrated low cerebral blood flow (CBF) in the left cerebral hemisphere (Fig. 2C). Cerebral angiography showed occlusion of the left MCA M1 segment with moyamoya-like vessels. No anastomosis extended from the external carotid arteries to the intracranial arteries (Fig. 2B, D). The symptoms were considered to be transient ischemic attack. Direct and indirect bypass surgery consisting of superficial temporal artery (STA)-MCA anasto-

Fig. 1 A, B: Left carotid angiograms, anteroposterior (A) and lateral views (B), revealing a Spetzler-Martin grade III arteriovenous malformation located in the left occipital lobe and splenium of the corpus callosum fed by the left anterior, middle, and posterior cerebral arteries. C, D: Left carotid angiograms, anteroposterior (C) and lateral views (D), after the second transarterial embolization showing some nidus reduction.

Fig. 2 A: T₁-weighted magnetic resonance image indicating the scheduled dose plan. B, D: Left carotid angiograms, anteroposterior (B) and lateral views (D), at 21 months after gamma knife surgery showing proximal middle cerebral artery occlusion with moyamoya-like vessels forming the internal carotid artery bifurcation. C: Technetium-99m-ethylcysteinate dimer single photon emission computed tomography scans at rest demonstrating low cerebral blood flow in the left cerebral hemisphere.

Fig. 3 A: Left carotid angiogram at 12 months after the surgery confirming adequate bypass flow in the frontal and temporal branches of the superficial temporal artery to the frontal and temporal lobes (black and white arrowheads), respectively. B: Technetium-99m-ethylcysteinate dimer single photon emission computed tomography scans at rest after surgery demonstrating improved cerebral blood flow in the left cerebral hemisphere. C, D: Serial left carotid angiograms at 31 (C) and 43 months (D) after gamma knife surgery (GKS) revealing progression of the right M1 stenosis (arrowheads). E: Magnetic resonance angiogram at 46 months after GKS showing right severe M1 stenosis (arrow).

mosis and encephalo-myo-synangiosis were performed on the left.

After surgery, the patient became free from ischemic symptoms. Adequate bypass flow was confirmed by follow-up angiography and ^{99m}Tc-ECD SPECT at rest (Fig. 3A, B). Serial angiography revealed progression of the right M1 stenosis (Fig. 3C, D). At 47 months after GKS, MR angiography showed severe right M1 stenosis (Fig. 3E). The patient remained neurologically stable and was free from clinical symptoms. Further observation is needed.

Discussion

This is an extremely rare case of moyamoya syndrome associated with GKS for AVM treated with bypass surgery. AVM associated with moyamoya disease is rare. Most cases had both neurovascular anomalies at the first presentation.^{1,3,6,10,11,13,16} High CBF related to the AVM may cause hemodynamic stress and affect the endothelium of the circle of Willis.^{10,11} Some cases of moyamoya disease show de novo development of AVM in the follow-up periods.^{4,15} In these cases, angiogenesis factors such as vascular endothelial growth factor (VEGF) might be activated by developing pial collateral formation, and subsequently induced AVM formation.¹⁶ Only two cases have shown de novo development of moyamoya vessels within the follow-up period of several years after treatment for AVMs (including our case).¹⁶

Our present patient and the previous patient in Taiwan developed bilateral moyamoya vessels after GKS. The high directional characteristic of GKS helps to avoid irradiation to the area surrounding the lesion. Our patient received 1.0–2.1 Gy at each ICA terminal and MCA M1 segment. Most cases of radiation-induced moyamoya syndrome received 22–120 Gy (mean 55.2 Gy).^{2,8)} These patients were treated with conventional radiation therapy for parasellar brain tumor. The irradiation field included the ICA terminal and the MCA M1 segment. Radiation-induced moyamoya syndrome is likely to occur within the irradiation field after high dose radiation therapy. Therefore, direct radiation injury may not be the main cause of moyamoya syndrome after GKS for AVM. Recently, radiation therapy was suggested to induce chronic oxidative stress and inflammation,¹⁸⁾ and irradiated tissue may undergo long-term changes in the expression of cytokines and growth factors.⁵⁾ The guidelines of research on intractable diseases of the Ministry of Health Labour and Welfare, Japan, suggest that inflammation leads to moyamoya syndrome.¹⁴⁾ Therefore, chronic inflammation due to GKS may have caused, in part, the formation of moyamoya vessels in this case. The long-term follow up of this case and accumulation of similar cases are required to resolve the mechanism of pathogenesis.

The treatment strategy of this case is controversial. Bypass surgery was performed because the patient suffered from transient ischemic attack after GKS and decreased CBF. However, this procedure carried some risk of AVM progression and rupture due to the nidus flow increase from the bypass flow. AVM progression has been reported after STA-MCA anastomosis for moyamoya disease.⁶⁾ Our case must be followed up in the long term to detect this rare complication.

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