Case Report

Efficacy of Direct Revascularization Surgery for Hemorrhagic Moyamoya Syndrome As a Late Complication of Cranial Irradiation for Childhood Craniopharyngioma

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Moyamoya syndrome (MMS) is an uncommon late complication after cranial irradiation. Its hemorrhagic presentation from the associated pseudo-aneurysm is extremely rare, and the optimal management strategy is undetermined. We herein report a 36-year-old man who developed intraventricular hemorrhage from a pseudo-aneurysm at the extended left anterior choroidal artery as an abnormal collateral of MMS 30 years after surgical removal and cranial irradiation for childhood craniopharyngioma. Catheter angiography confirmed the diagnosis of MMS, and multiple pseudo-aneurysms were evident at the ipsilateral abnormal choroidal collateral, one of which was considered to be a source of bleeding. The patient underwent left superficial temporal artery (STA)-middle cerebral artery (MCA) anastomosis with indirect pial synangiosis based on the observation that the development of choroidal collateral may be associated with a high rebleeding risk in hemorrhagic moyamoya disease. The patient was discharged without neurological deficit, and postoperative magnetic resonance angiography confirmed the STA-MCA bypass to be patent. Catheter angiography 1 year after revascularization surgery revealed the complete disappearance of the pseudoaneurysms with the apparently patent STA-MCA bypass. The patient did not exhibit any cerebrovascular events during the follow-up period of 16 months. In conclusion, hemorrhagic MMS with choroidal collateral as a dangerous anastomosis was effectively managed by STA-MCA anastomosis. Although long-term follow-up is necessary to evaluate our strategy, the favorable disappearance of pseudoaneurysms after revascularization surgery in the present case strongly suggests that STA-MCA anastomosis has a potential role for preventing rebleeding in MMS after cranial irradiation.

Key Words: Moyamoya syndrome—cranial irradiation—craniopharyngioma—extracranial-intracranial bypass

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Introduction

Moyamoya disease (MMD) is a chronic, occlusive cerebrovascular disease characterized by bilateral steno-occlusive changes at the terminal portion of the internal carotid artery and an abnormal vascular network at the base of the brain.[1] Moyamoya syndrome (MMS) is defined as MMD in association with an underlying disease,[2,3] and is also known as an uncommon late complication after cranial irradiation.[4-9] The hemorrhagic presentation of MMS is uncommon, and its association with a pseudoaneurysm at the abnormal collateral is extremely rare.[4,7,9] Although a recent multicenter randomized controlled trial, the Japan Adult Moyamoya (JAM) trial, strongly suggested that superficial temporal artery (STA)-middle cerebral artery (MCA) anastomosis can prevent rebleeding in patients with hemorrhagic-onset MMD,[10,11] its exact efficacy for hemorrhagic MMS with a variety of associated disorders remains undetermined.[12]

We herein present a 36-year-old patient with MMS as a late complication of cranial irradiation, manifesting as intraventricular hemorrhage from a pseudoaneurysm at the extended anterior choroidal artery 30 years after the management of childhood craniopharyngioma. The patient was successfully managed by STA-MCA anastomosis, resulting in complete disappearance of the pseudoaneurysms.

Case Presentation

A 36-year-old man developed intraventricular hemorrhage from a pseudoaneurysm at the extended left anterior choroidal artery as an abnormal collateral of MMS 30 years after surgical removal and cranial irradiation for childhood craniopharyngioma. He was admitted to our hospital 1 day after the sudden onset of headache. His neurological examination was normal, but computed tomography revealed intraventricular hemorrhage predominantly on the left side (Fig 1A). The patient had undergone microsurgical removal of the craniopharyngioma by bifrontal craniotomy and subsequent fractionated cranial irradiation with a total radiation dose of 50 Gy. There had been no tumor recurrence for 30 years. T2-star weighted magnetic resonance (MR) imaging delineated multiple low-intensity lesions, one of which was exposed to the left lateral ventricle (arrowhead in Fig 1B). Catheter angiography demonstrated steno-occlusive changes at the terminal portion of the bilateral internal carotid arteries associated with abnormal vascular networks at the base of the brain, leading to the definitive diagnosis of MMS (Fig 1 C, D). Multiple pseudoaneurysms were also evident at the extended anterior choroidal artery by left carotid angiography (arrowhead in Fig 1D), one of which was considered to be a source of bleeding.[13] Considering the anatomical location of the pseudoaneurysms and the site of the low-intensity spot on T2-star weighted MR imaging, the posterior side of the aneurysm was suspected as the source of hemorrhage.

One month after the onset, the patient underwent left STA-MCA anastomosis with indirect pial synangiosis.[14] The stump of the STA was microsurgically anastomosed to the M4 segment of the left MCA, which supplied the parietal lobe for a temporary occlusion time of 21 minutes (Fig 2A). The patency of the bypass was confirmed by Doppler ultrasound sonography and indocyanine green video-angiography (Fig 2B), and encephalo-duro-myo-synangiosis were subsequently performed. The postoperative course was uneventful and the patient was discharged without neurological deficit. No additional parenchymal lesions were noted on postoperative MR imaging (Fig 2C, D), and MR angiography confirmed the patent STA-MCA bypass (arrow in Fig 2E). The follow-up catheter angiography 1 year after revascularization surgery revealed the complete disappearance of the pseudoaneurysms (Fig 2F, G) with the apparently patent STA-MCA bypass. The patient did not exhibit any cerebrovascular events during the follow-up period of 16 months.

Discussion

MMS, also called quasi-MMD or akin MMD, is defined as MMD in association with an underlying disease.[2,3] Underlying diseases of MMS include atherosclerosis, autoimmune disease, meningitis, von Recklinghausen disease, Down's syndrome, traumatic brain injury, brain tumors, and cranial irradiation.[2,3] Regarding MMS associated with cranial irradiation, it is also known as "radiation-induced moyamoya vasculopathy" as a late complication of radiation therapy for primary brain tumors during childhood.[4-9] According to previous reports, the incidence of MMS is as high as 3.5%-17.1% after radiation therapy for primary brain tumors.[4,5] The most common clinical presentation of MMS after cranial irradiation is cerebral infarction, and its hemorrhagic manifestation from the associated pseudoaneurysm is extremely rare.[4,7] Ullrich et al reported that the risk of infarction was much higher in patients with MMS after cranial irradiation compared with that in definitive MMD patients,[4] and none of the 12 patients in their series had hemorrhagic onset. Wang et al performed a literature review and found only 2 cases of hemorrhage presentation among the 47 cases of MMS associated with cranial irradiation, and the interval between radiation therapy and the development of moyamoya vasculopathy was 3.3 years.[7] In our case, intraventricular hemorrhage with multiple pseudoaneurysms developed 30 years after radiation therapy for childhood craniopharyngioma, which is considered to be an extremely rare condition.

In general, the management strategy for MMS should be consistent with that for idiopathic MMD.[2,3,12] Therefore, STA-MCA bypass with or without pial synangiosis is widely used for ischemic-onset MMS,[2,3] but the exact

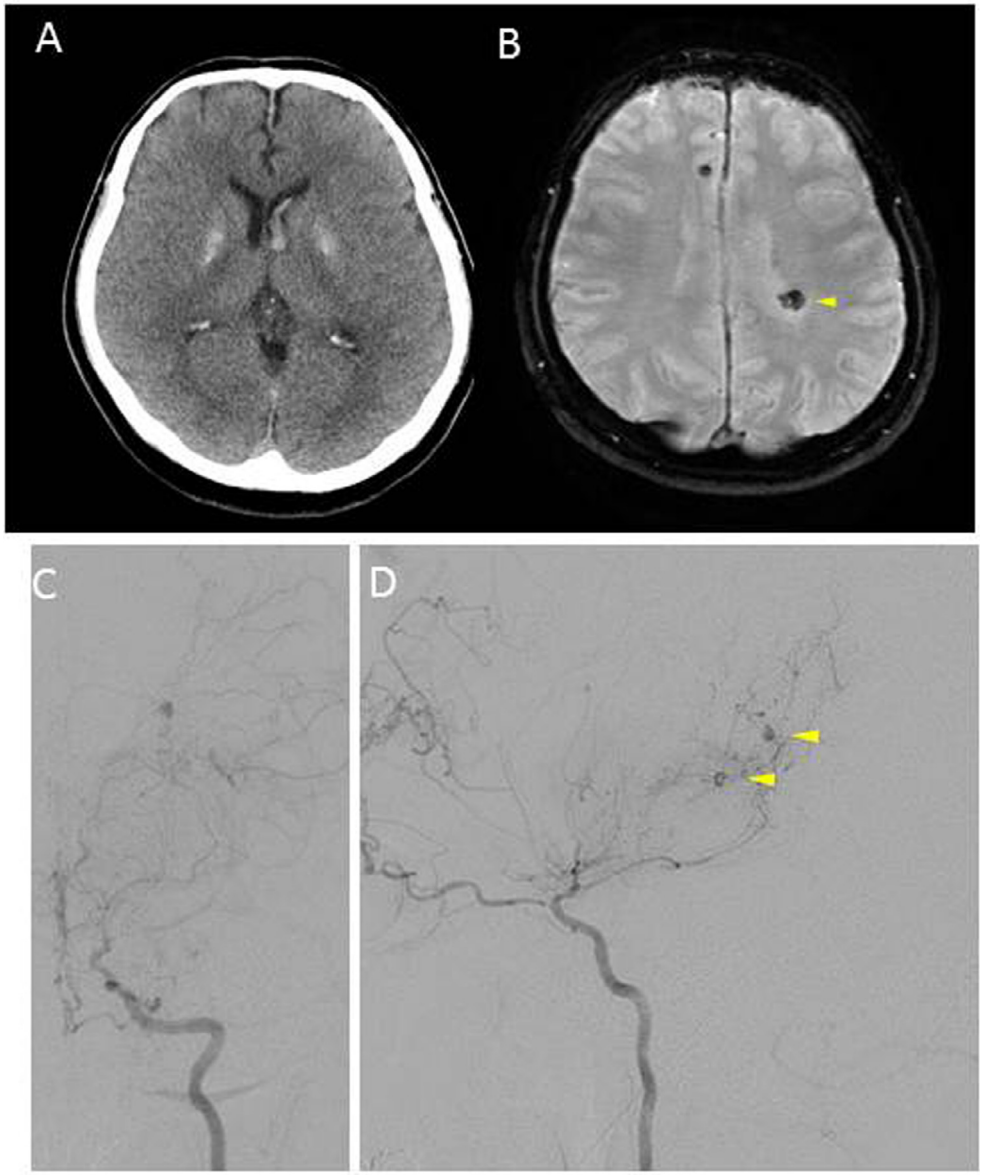

Figure 1. *(A) Initial computed tomography scan showing intraventricular hemorrhage predominantly on the left side. (B) T2-star weighted magnetic resonance (MR) imaging demonstrating small low-intensity spots, one of which at the lateral wall of the left ventricle was suspected to be a source of bleeding. (C, D) Left internal carotid angiogram showing marked stenosis at the left terminal internal carotid artery associated with moyamoya vessels. Pseudoaneurysms in the lateral plexus branch arising from the extended anterior choroidal artery (arrowheads). (C) Anterior-posterior view, (D) lateral view.*

efficacy of STA-MCA bypass to prevent rebleeding in hemorrhagic MMS is still unknown.[12] Regarding idiopathic MMD with hemorrhagic presentation, the JAM trial, a multicenter randomized controlled trial, examined the efficacy of STA-MCA bypass to prevent rebleeding in adult patients with hemorrhagic MMD.[10] The JAM trial also suggested that MMD patients with posterior hemorrhage had a significantly high annual rebleeding risk of 17.1% per year, and STA-MCA bypass markedly reduced the risk of rebleeding in this patient population.[11] Furthermore, the supplemental analysis of the JAM trial revealed that extension and dilatation of the anterior choroidal artery, so-called choroidal collateral development, is significantly associated with posterior hemorrhage, and was found to be an independent risk factor for rebleeding attacks.[15,16] Based on these most recent reports on hemorrhagic MMD,[17] we considered STA-MCA bypass to be able to effectively reduce the risk of rebleeding in our case based on the development of the choroidal collateral with a pseudoaneurysm as a bleeding source.

Several management strategies of the pseudoaneurysm on the collateral vessels in MMD patients were reported previously. Efficacy of the endovascular direct obliteration of the pseudoaneurysm on the collateral vessels were reported in case series by Kim et al,[18] and by other authors.[19,20] Kikuta et al alternatively reported a 65-year-old

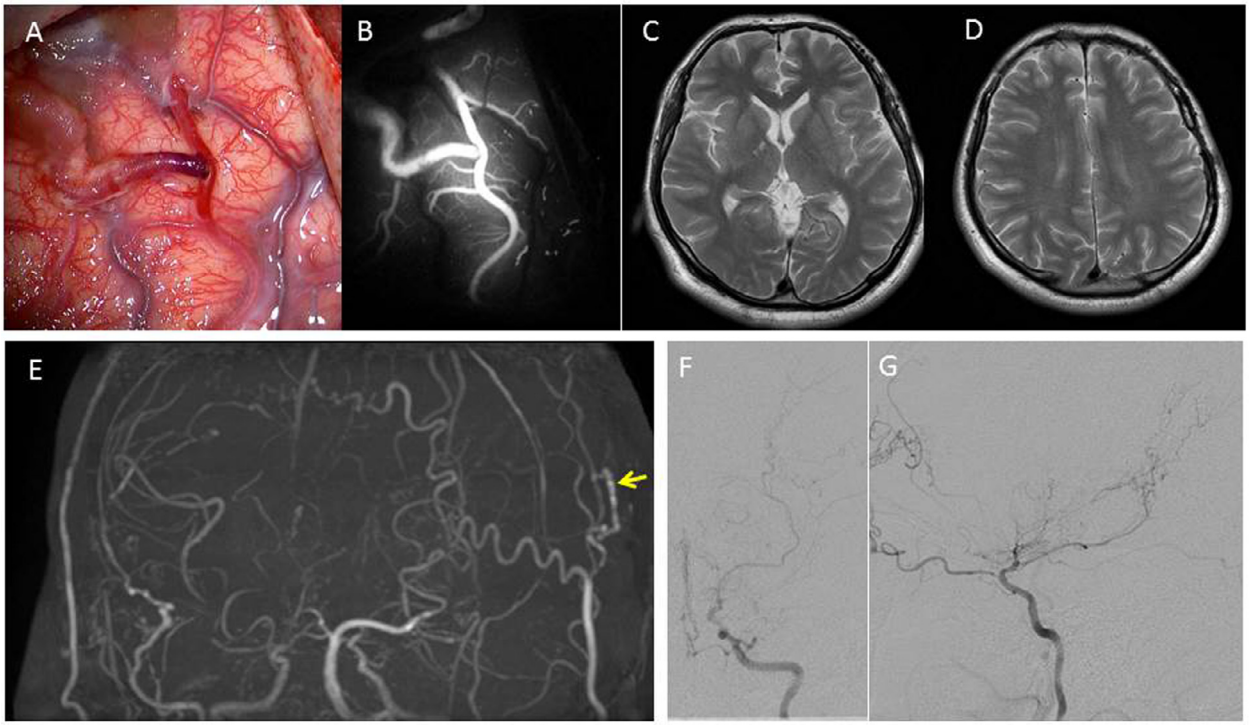

Figure 2. *(A) Intraoperative microscopic view after left STA-MCA bypass. (B) Indocyanine green video-angiography demonstrated the apparently patent bypass with favorable distribution of bypass flow. (C, D) T2-weighted MR imaging on postoperative day 2 showing no parenchymal lesion after the surgery. (E) Postoperative MR angiography showing the apparently patent STA-MCA bypass (arrow). (F, G) Left internal carotid artery angiograms obtained 1 year after surgery showing the complete disappearance of the pseudoaneurysms. (F) anterior-posterior view, (G) lateral view. Abbreviation: STA-MCA, superficial temporal artery-middle cerebral artery. (Color version of figure is available online.)*

MMD patient undergoing direct microsurgical approach to the pseudoaneurysm with STA-MCA bypass.[21] Spontaneous disappearance of pseudoaneurysm under conservative management was also reported previously.[22] Contrarily to these strategies, we have selected STA-MCA bypass without direct obliteration of the ruptured pseudoaneurysm, in light of the multiplicity of the pseudoaneurysms as well as the contribution of the abnormal collateral with pseudoaneurysms to the vascular supply into the intact cortex in our patient. Our strategy was supported by the previous report by Kuroda et al demonstrating that STA-MCA bypass may prevent pseudoaneurysms at the abnormal collateral in MMD patients.[23] Similar effects of STA-MCA bypass were also reported in other studies, which further supported our strategy.[24,25] The present case was hemorrhagic-onset MMS after radiation therapy, and the source of hemorrhage was suspected to be the pseudoaneurysms on choroidal anastomosis. Direct revascularization surgery was performed and disappearance of the aneurysms was confirmed after the 1-year follow-up period. The patient had no cerebrovascular events. Although the follow-up period in our case was short, 16 months, complete disappearance of the pseudoaneurysms 1 year after direct revascularization surgery strongly suggests that STA-MCA bypass effectively reduced the risk of rebleeding in this case. Thus, we recommend STA-MCA bypass as an optimal management strategy for hemorrhagic-onset MMS after cranial irradiation associated with the choroidal collaterals as a source of bleeding.

Conclusions

Hemorrhagic MMS with the choroidal collateral as an unstable anastomosis may be effectively managed by STA-MCA anastomosis. Although long-term follow-up is necessary to evaluate our strategy, the favorable disappearance of the pseudoaneurysms after revascularization surgery in the present case strongly suggests that STA-MCA anastomosis can prevent rebleeding in MMS after cranial irradiation.

Disclosures

The authors have no conflicting remarks.

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