Biphasic Development of Focal Cerebral Hyperperfusion After Revascularization Surgery for Adult Moyamoya Disease Associated With Autosomal Dominant Polycystic Kidney Disease

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Background: Cerebral hyperperfusion (CHP) syndrome is a potential complication of superficial temporal artery-middle cerebral artery (STA-MCA) anastomosis for moyamoya disease (MMD), but its biphasic and delayed development is extremely rare. *Case report:* A 47-year-old woman with autosomal dominant kidney disease (ADPKD) presented with transient ischemic attacks due to MMD, and underwent left STA-MCA anastomosis. N-isopropyl-p-[^{123}I] iodoamphetamine single-photon emission computed tomography (^{123}IMP-SPECT) 1 day after surgery revealed asymptomatic CHP at the site of anastomosis. Strict blood pressure control and minocycline hydrochloride relieved CHP at postoperative day 7. However, 2 days later, the patient complained of sensory aphasia, and ^{123}IMP-SPECT demonstrated significant focal CHP at the site of anastomosis accompanying high-intensity signal on magnetic resonance (MR) imaging of fluid attenuated inversion recovery (FLAIR) in her left temporal lobe near the site of anastomosis. We continued strict blood pressure control and additionally administered free radical scavenger (Edaravone) and antiepileptic agents, which gradually improved sensory aphasia. MR imaging and ^{123}IMP-SPECT also confirmed the amelioration of the FLAIR-high lesion and focal CHP in her left temporal lobe. Two months later, the patient underwent right STA-MCA anastomosis without complications. *Conclusions:* Although the underlying mechanism is unknown, biphasic development of focal CHP after revascularization surgery in an MMD patient with ADPKD is unique. Due to the potential vulnerability of the systemic vessels in ADPKD, it is conceivable that intrinsic vascular wall fragility in MMD could be enhanced by ADPKD and have partly led to this rare complication.

Key Words: Moyamoya disease—cerebral hyperperfusion syndrome—autosomal dominant polycystic kidney disease—Revascularization surgery—surgical complication

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Introduction

Moyamoya disease (MMD) is a chronic and occlusive cerebrovascular disease characterized by bilateral stenoocclusive changes at the terminal portion of the internal carotid artery and an abnormal vascular network at the base of the brain.[1] Superficial temporal artery-middle cerebral artery (STA-MCA) anastomosis is widely performed for standard management of adult MMD.[2-4] Cerebral hyperperfusion (CHP) syndrome is one of the potential complications of STA-MCA anastomosis for adult MMD,[5-7] but it generally has a favorable prognosis under modern postoperative management protocols with strict blood pressure control and administration of

minocycline hydrochloride.[8,9] However delayed and/or biphasic development of CHP is extremely rare. We herein report a patient with adult MMD who developed prolonged CHP with a biphasic pattern after revascularization surgery, despite intensive postoperative management, associated with autosomal dominant polycystic kidney disease (ADPKD).

Case Presentation

A 47-year-old woman with MMD developed transient paralysis of her extremities after laughing. Six years before the occurrence of the transient ischemic attacks (TIAs), she was found to have multiple cysts in the liver and kidney during a medical check-up, and was diagnosed with ADPKD. Initial magnetic resonance (MR) imaging for the screening of intracranial aneurysms did not reveal any saccular aneurysm formation, but stenosis of the terminal portion of her bilateral internal carotid arteries was noted. MMD was suspected and she began yearly follow-up by MR imaging/angiography. Her mother also had a history of ADPKD and MMD. After the development of transient ischemic attacks, the patient underwent catheter angiography, which 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 (Fig 1A), leading to the definitive diagnosis of MMD. At this point, the patient was free from hypertension and renal dysfunction. On genetic analysis of RING-finger protein-213, a susceptibility gene for MMD, a heterozygous variant in c.14576 G>A of the RING-finger-213 gene was found. N-isopropyl-p-[^{123}I] iodoamphetamine single-photon emission computed tomography (^{123}I-IMP SPECT) revealed a significant decrease in cerebral blood flow (CBF) in the territory of left middle cerebral artery (Fig 1B). She then underwent left STA-MCA anastomosis with encephalo-duro-myo-synangiosis. The stump of the parietal branch of the STA was anastomosed to the M4 segment of the left MCA, which supplied the temporal lobe (Fig 2A). The diameter of the recipient artery was 1.0 mm, and the temporary occlusion time was 22 minutes. Intraoperative

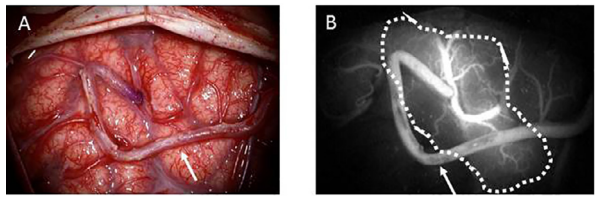

Figure 2. *(A) Intraoperative microscopic view demonstrating the dilated cortical pial arteries near the site of anastomosis. The arrow indicates the superficial temporal artery (STA). (B) Intraoperative indocyanine green video angiography after the anastomosis showing focal early filling (dotted line) from the STA.*

indocyanine green video angiography after the anastomosis demonstrated localized early filling through the STA with early venous filling of the cortical vein around the site of anastomosis (Fig 2B). No neurological deterioration was exhibited immediately after surgery, but a focal increase in CBF at the site of the STA-MCA anastomosis was observed on ^{123}I-IMP SPECT one day after surgery (arrowhead in Fig 3A) and fluid attenuated inversion recovery indicated increased ivy signs in her left temporal lobe (Fig 3B). The apparently patent left STA-MCA bypass with high signal intensity was observed on MR angiography (arrow in Fig 3C). Following our standard management of blood pressure lowering and minocycline hydrochloride administration,[8] the patient did not develop neurological deterioration due to CHP for one week after surgery. ^{123}I-IMP SPECT on postoperative day 7 confirmed the amelioration of CHP (Fig 3D). Two days later, however, the patient complained of sensory aphasia, and MR imaging demonstrated high-intensity signal in the white matter of her left temporal lobe on fluid attenuated inversion recovery (Fig 3F), accompanying increased signal intensity on diffusion-weighted imaging in the surface of left temporal lobe (Fig 3G) with an increased apparent diffusion coefficient (Fig 3H). MR angiography revealed the increased intensity of left STA-MCA bypass (Fig 3I), which is in accordance with ^{123}IMP-SPECT finding.[10,11] A focally intense increase in CBF at the site of anastomosis was noted on ^{123}I-IMP SPECT, indicating the biphasic occurrence of CHP (asterisk in Fig 3E), since the patient met the following diagnostic criteria for CHP; the

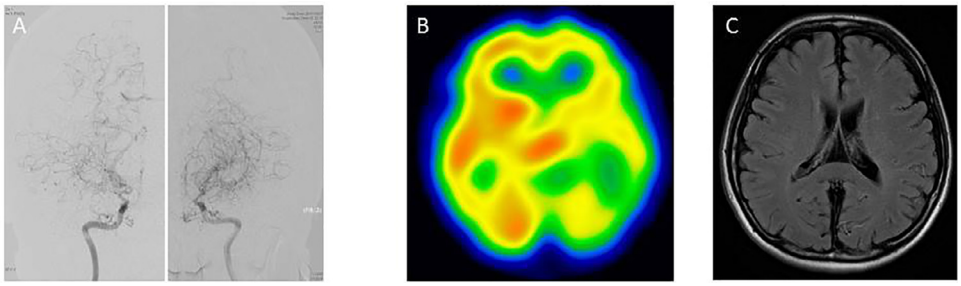

Figure 1. ***(A)*** *Preoperative catheter angiography showing stenosis of the terminal portion of bilateral internal carotid arteries and abnormal vascular network at the base of the brain. (B) N-isopropyl-p-[^{123}I] iodoamphetamine single photon emission computed tomography (^{123}I-IMP SPECT) showing decreased cerebral blood flow in the territory of the left middle cerebral artery (MCA). (C) Fluid-attenuated inversion recovery (FLAIR) magnetic resonance (MR) imaging showing ivy signs in the left hemisphere.*