Hyperperfusion Syndrome after Extracranial-Intracranial Bypass in a Patient with Moyamoya Disease

—Case Report—

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Abstract

A 47-year-old female developed hyperperfusion syndrome after superficial temporal artery-middle cerebral artery (STA-MCA) bypass for moyamoya disease. She presented with right hemiparesis and motor aphasia due to left cerebral infarction. She underwent left STA-MCA bypass. One day after surgery, she manifested neurological deterioration. Magnetic resonance (MR) imaging 4 days after the operation indicated regional brain edema in the territory supplied by the bypass, and single photon emission computed tomography 17 days after the operation demonstrated hyperperfusion in that area. Symptoms improved within 1 week after surgery, and MR imaging showed disappearance of edema and return to the preoperative appearance. Such events are rare, but hyperperfusion syndrome may occur after STA-MCA bypass for moyamoya disease.

Key words: extracranial-intracranial bypass, moyamoya disease, hyperperfusion, magnetic resonance imaging

Introduction

Moyamoya disease is a bilateral occlusive disorder located at the terminal of the internal carotid artery (ICA),¹⁰⁾ but the angiographic stage of progression may differ between sides.⁷⁾ Pediatric patients present with ischemic cerebral symptoms, whereas adults often develop cerebral hemorrhage.⁵⁾ Recently, adults manifesting transient ischemic attack or ischemic stroke have been identified by magnetic resonance (MR) imaging and MR angiography.⁴⁾ Superficial temporal artery-middle cerebral artery (STA-MCA) bypass is the best treatment for such patients.⁵⁾ Hyperperfusion syndrome after extracranial-intracranial (EC/IC) bypass,^{2,3,8)} or carotid endarterectomy have been reported,^{1,9)} but hyperperfusion syndrome following EC/IC bypass in moyamoya disease has never been observed. We report a patient with moyamoya disease who underwent EC/IC bypass complicated by hyperperfusion syndrome observed by MR imaging and single pho-

ton emission computed tomography (SPECT).

Case Report

A 47-year-old female suffered onset of right hemiparesis and motor aphasia on March 3, 1996. She was receiving a calcium antagonist for hypertension. Initial computed tomography (CT) showed no abnormality. Left carotid angiography showed severe stenosis involving the distal ICA, basal moyamoya vessels (Fig. 1A, B), and poor collateral circulation to the left MCA territory via the posterior cerebral artery. Right carotid angiography showed moderate stenosis of the MCA just distal to the ICA bifurcation (Fig. 1D, E). Initial T₂-weighted MR imaging 12 hours after onset demonstrated increased signal intensity in the left insula and supramarginal gyrus (Fig. 2 upper). The diagnosis was cerebral infarction due to moyamoya disease. Antiplatelet medication was administered. SPECT with technetium-99m-hexamethylpropyleneamine oxime (^{99m}Tc-HMPAO) showed decreased cerebral blood flow (CBF) in both the area of infarction and the temporoparietal region (Fig. 3). ^{99m}Tc-HMPAO SPECT with

Fig. 1 A, B: Left carotid angiograms showing severe stenosis at the end of internal carotid artery and moyamoya vessels. C: Left carotid angiogram 1 year after onset showing progression of the terminal internal carotid artery stenosis (arrow). D, E: Right carotid angiograms showing moderate stenosis of the middle cerebral artery just distal to the internal carotid artery bifurcation.

acetazolamide showed vascular reactivity was impaired in the same region. We considered that STA-MCA bypass with encephalo-myo-synangiosis was indicated if her major neurological state improved. After transfer for rehabilitation to a local hospital, her mild right hemiparesis and motor aphasia gradually improved, permitting independent living. Therefore, we decided to perform left-sided prophylactic revascularization surgery.

She was readmitted to our hospital on February 18, 1997. Progression of moyamoya disease and changes in collateral circulation to the left MCA territory were evaluated by follow-up angiography on February 21, 1997. Cerebral angiography showed progression of the terminal left ICA stenosis (Fig. 1C). Preoperative MR imaging demonstrated the old left cerebral infarction in the insula and supramarginal gyrus (Fig. 2 lower). Her blood pressure was controlled to 120–140 mmHg with a calcium antagonist

administration. Left STA-MCA bypass with encephalo-myo-synangiosis was performed on February 28. The flow volume of the STA was 40 ml/min, and patencies of the STA and the recipient vessel, the angular artery, were very good. Postoperatively she showed no immediate neurological change. CT after the operation detected no new lesion. However, her motor aphasia deteriorated beginning the next morning, although left STA pulsation was very good and follow-up CT found no change. Her blood pressure was controlled to normotension, between 120 and 140 mmHg, by continuous intravenous injection of calcium antagonist. MR imaging 4 days after operation indicated slightly decreased intensity on T₁-weighted image and hyperintensity on T₂-weighted image in the left frontoparietal region (Fig. 4 left). At this point, we considered that the cause of her deterioration was hyperperfusion of the bypass territory or ischemia due to interaction between the flow

Fig. 2 upper: T₂-weighted magnetic resonance (MR) images 12 hours after onset showing hyperintensity in the left insula and supramarginal gyrus. lower: T₂-weighted MR images before the operation demonstrating the old cerebral infarction in the left insula and supramarginal gyrus.

from the STA and proximal MCA at the frontoparietal region. We continued to control her blood pressure at normotension (120–140 mmHg) and administered steroids and glycerol. Seven days after surgery, her motor aphasia had gradually improved, and MR imaging 17 days after surgery showed decreases in the abnormal intensity in the left frontoparietal region (Fig. 4 center). ^{99m}Tc-HMPAO SPECT on the same day showed a regional increase in CBF in this area (Fig. 5). At this time her neurological state had improved to the preoperative state.

She was discharged on April 5, 1997 with mild motor aphasia and right hemiparesis, in nearly the same clinical condition as her preoperative status. She returned to independent daily life. MR imaging 54 days after the operation indicated that the abnormal intensity had disappeared except for the old infarct (Fig. 4 right). Follow-up angiography 3 months after the STA-MCA bypass surgery showed good patency of the bypass graft (Fig. 6).

Discussion

Five cases of hyperperfusion syndrome after STA-MCA bypass have occurred in patients with atherosclerosis and poor collateral circulation.²⁾ These patients were neurologically well on awaking from anesthesia. CT after STA-MCA bypass did not show cerebral infarction, and their neurological deterioration improved within 2 weeks. Postoperative SPECT demonstrated hyperperfusion syndrome after EC/IC bypass in patients with ICA occlusion.⁸⁾ Follow-up SPECT 3 months after surgery revealed normalization of regional CBF in the territory of the STA-MCA bypass. SPECT also detected luxury perfusion syndrome after STA-MCA bypass.³⁾ Our patient awakened from anesthesia in good condition and CT showed no new lesion. Follow-up CT after neurological deterioration also showed no new lesion. At this time her blood pressure was between 120 and 140 mmHg. Her blood pressure was maintained at normotension, and her neurological state stabilized although MR imaging 4 days after the operation indicated regional edema in the area supplied by the bypass artery. We speculate that the cause of her neurological deterioration was hyperperfusion based on the following reasons. The flow volume of the STA was higher than the average flow volume of STA in patients with moyamoya disease (mean flow volume in 12 recent patients undergoing bypass surgery was 23.3 ± 13.2 ml). SPECT 17 days after the operation still showed hyperperfusion where postoperative MR imaging on the same day indicated regional edema and follow-up MR imaging showed no new cerebral infarction. Her neurological deterioration was improved by 14 days after the operation.

Hyperperfusion syndrome may also occur after carotid endarterectomy in patients with severe ICA stenosis and poor collateral flow.^{1,9)} Acetazolamide SPECT demonstrated poor reactivity in these patients,⁸⁾ so vasoparalysis might induce hyperperfusion in regions with impaired autoregulation.^{2,3,6,8,9)} In our patient, acetazolamide SPECT before the operation showed poor reactivity in the territory of the bypass. Vessels are considered to be in vasoparalysis in moyamoya disease. Therefore, the hyperperfusion in our patient might have occurred because the blood flow from the STA was abundant compared with the average flow volume of the STA. However, SPECT 17 days after operation did not demonstrate hyperperfusion syndrome. Possibly SPECT should have been performed on the day of neurological deterioration.

The symptoms of hyperperfusion syndrome include headache, ocular pain, convulsions, and impaired consciousness. If hyperperfusion is compli-

Fig. 3 Preoperative single photon emission computed tomography scans demonstrating decreased cerebral blood flow in the infarction area and the temporoparietal region.

Fig. 5 Single photon emission computed tomography scans 17 days after the operation showing increased cerebral blood flow in the frontoparietal lobe (arrows).

Fig. 4 *left:* Postoperative T_1 - and T_2 -weighted magnetic resonance (MR) images 4 days after the operation showing cerebral edema in the left frontoparietal lobe. *center:* MR images 17 days after the operation showing decreased abnormal intensity in the same area. *right:* MR images 54 days after the operation demonstrating disappearance of brain edema in the frontoparietal lobe.

Fig. 6 Left carotid angiograms 3 months after the operation showing good patency of the bypass graft.

cated by intracranial hemorrhage, mortality exceeds 50%.⁹⁾ Our patient improved gradually up to about 14 days after operation.²⁾ Symptoms had improved to the preoperative condition 7 days after surgery. Repeat MR imaging after the operation demonstrated abnormal intensity in the temporoparietal region, with the MR imaging appearance improving over the following week. Hyperperfusion may appear as reversible low-density lesion on CT, considered to be due to regional ischemia or edema.⁶⁾ We suspect that the abnormal intensity on MR imaging after the STA-MCA bypass represents brain edema due to hyperperfusion, because angiography after the STA-MCA bypass showed good patency of the STA and postoperative SPECT revealed a hot spot corresponding to the abnormal area on MR imaging.

Treatment of hyperperfusion syndrome involves controlling the blood pressure to normotension or slight hypotension.^{8,9)} Our patient's systolic blood pressure was maintained between 120 and 140 mmHg using a calcium antagonist. If the patient develops convulsion or the level of consciousness decreases, barbiturate anesthesia should be given, although such symptoms predict a poor outcome.^{1,9)} If the patient shows neurological deterioration without a new lesion on CT, the cause of deterioration should be immediately evaluated by MR imaging and SPECT. Treatment of patients with hyperperfusion is facilitated by immediate diagnosis before symptoms become severe.

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