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Case Report

Prolonged and regionally progressive symptomatic cerebral hyperperfusion syndrome after superficial temporal artery-middle cerebral artery anastomosis in a patient with moyamoya disease

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Abstract

Background: The incidence of symptomatic hyperperfusion syndrome after superficial temporal artery-middle cerebral artery (STA-MCA) anastomosis for patients with moyamoya disease (MMD) approaches 30%. In most cases, hyperperfusion occurs in a localized area and disappears within 1–2 weeks.

Case Description: A 59-year-old female diagnosed with asymptomatic MMD for 4 months became rapidly symptomatic with transient ischemic attacks (TIAs). After left STA-MCA anastomosis surgery, she developed symptomatic hyperperfusion, initially (1–2 weeks after surgery) manifesting with severe headache and lesions located in the left basal ganglia. She then developed (2–5 weeks after surgery) aphasia and right hemiparesis caused by new hyperperfusion lesions located in the left frontal area. At discharge (7 weeks after surgery), she recovered fully without any remaining neurologic deficit and no ischemic lesions.

Conclusion: This report details a rare case of a patient with MMD who presented with regionally progressive hyperperfusion lesions after STA-MCA anastomosis and symptoms that persisted for 5 weeks following surgery. Results from this case suggest that regional differences exist in the functional recovery of cerebrovascular reactivity (CVR) in a patient with rapidly progressive MMD.

Key Words: Cerebral hyperperfusion, moyamoya disease, STA-MCA anastomosis

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INTRODUCTION

Moyamoya disease (MMD) is characterized by the presence of moyamoya vessels, which are a collateral network of fine vascular pathways that form due to progressive stenosis or occlusion of the bilateral terminal internal carotid arteries.[21] Many patients with MMD suffer a ischemic attack, such as cerebral infarction or transient ischemic attack (TIA). Unfortunately, no specific medical treatment for MMD has been identified.[10,22]

Surgical revascularization for MMD prevents cerebral ischemic attacks by improving cerebral blood flow (CBF). In general, superficial temporal artery-middle cerebral artery (STA-MCA) anastomosis with or without indirect pial synangiosis is considered the standard surgical revascularization treatment for MMD.[6,7,16,18] Although direct anastomosis surgery (STA-MCA anastomosis) for MMD improves outcomes, during the acute stage, this direct bypass surgery carries the risk of temporary neurologic deterioration due to focal cerebral hyperperfusion at the site of the anastomosis.[1-5,13]

Recently, some authors have reported that symptomatic hyperperfusion after STA-MCA anastomosis for patients with MMD occurs only in the anastomosed region. These authors have also shown that the hyperperfusion state, which can be detected using single-photon emission computed tomography (SPECT), usually disappears within 1 week after bypass surgery, although in most cases, symptoms persisted for 1–2 weeks.[4,7,11,16,18] This report details a patient with MMD who, after STA-MCA anastomosis, presented with hyperperfusion and prolonged symptoms of 5 weeks duration.

CASE REPORT

A 59-year-old female presented with complaints of dull headache. Magnetic resonance (MR) imaging showed no ischemic lesion, but MR angiography showed bilateral stenosis at the terminal portion of internal carotid artery (ICA), proximal anterior cerebral artery (ACA), and middle cerebral artery (MCA). Angiography showed severe stenosis involving the bilateral distal ICA and dense moyamoya vessels. A diagnosis of MMD was made according to criteria from the Research Committee on Spontaneous Occlusion of the Circle of Willis of the Ministry of Health Labor and Welfare, Japan. *N*-isopropyl-*p*-[^{123}I]-iodoamphetamine single-photon emission computed tomography (^{123}I-IMP-SPECT) did not demonstrate dramatically decreased CBF in the bilateral ACA and MCA territories. ^{123}I-IMP-SPECT with acetazolamide showed no impairment in vascular reactivity. At the time, the working diagnosis was asymptomatic MMD without a decrease in CBF.

Four months after initial presentation, the patient complained of frequent episodes of transient right hemiparesis. MR imaging showed rapid progression of the previously identified stenosis and small white matter ischemic lesions in the bilateral frontal areas. ^{123}I-IMP-SPECT showed decreased CBF in the bilateral ACA and MCA territories and ^{123}I-IMP-SPECT with acetazolamide showed markedly impaired vascular reactivity bilaterally [Figure 1]. Due to the presence of symptoms originating in the left hemisphere and a more severe decrease in left-hemispheric CBF, left STA-MCA bypass with encephalo-duro-myo-synangiosis was performed. The recipient artery at the M4 segment was a small-diameter (0.7 mm) vessel and the diameter of the STA was larger than the recipient artery (1.3 mm). Patency of the STA-MCA anastomosis was confirmed using Doppler and intraoperative indocyanine-green (ICG) angiography.

The patient had no neurologic deficits immediately after surgery, but she complained of a severe headache on postoperative day (POD) 1. On POD 2, the headache worsened and the patient began vomiting. On POD 4, ^{123}I-IMP-SPECT showed a significant increase in CBF in the left basal ganglia [Figure 2]. Postoperative MRI showed a patent STA-MCA bypass and no evidence of new ischemic changes. The patient was diagnosed with symptomatic cerebral hyperperfusion and intensive blood pressure control (under 125 mmHg of systolic blood pressure) with diltiazem was initiated, which gradually relieved her severe headache and vomiting.

On POD 7, although the systolic blood pressure was well controlled ranging from 120 to 110mmHg during POD 4–6, the patient returned with recurrent severe headache accompanied by aphasia, dysarthria, right hemiparesis, and numbness in the right upper extremity. Imaging showed that the hyperperfusion lesion had shifted from the left basal ganglia to the regions of the cortex surrounding the anastomosis and included left temporal, parietal, and lateral frontal lesions. The free radical scavenger, edaravone (Mitsubishi Tanabe Pharma Co., Tokyo, Japan), was initiated at this time. Two weeks after surgery, the hyperperfusion area had localized to the left lateral frontal area; however, her symptoms persisted for several weeks. Three weeks after surgery, hyperperfusion on ^{123}I-IMP-SPECT had improved, demonstrating only a small hyperperfusion spot. However, the right hand numbness and right hemiparesis persisted until 5 weeks after surgery. At the time of discharge (7 weeks after surgery), the patient had no neurologic sequelae. Throughout her entire postoperative course, MRI showed no new ischemic or hemorrhagic lesions.

DISCUSSION

Recent reports have shown that the incidence of symptomatic cerebral hyperperfusion after STA-MCA anastomosis for MMD is as high as 27.5–38.2%.[1-5,13,15] Clinically, it is important to definitively diagnose hyperperfusion syndrome, because the clinical presentation is similar to an ischemic attack, but medical treatments for hyperperfusion syndrome and ischemic attacks are in direct opposition.

In this case, the first symptom to appear was severe headache without apparent focal neurologic deficits, which was associated with hyperperfusion only detectable in the basal ganglia ipsilateral to the operation. Although, Fujimura *et al*.[3] described headache in these cases related to an intracranial hemorrhagic lesion, no intracranial hemorrhagic lesions were identified on imaging. Cerebral hyperperfusion syndrome after carotid endarterectomy (CEA) is characterized by unilateral

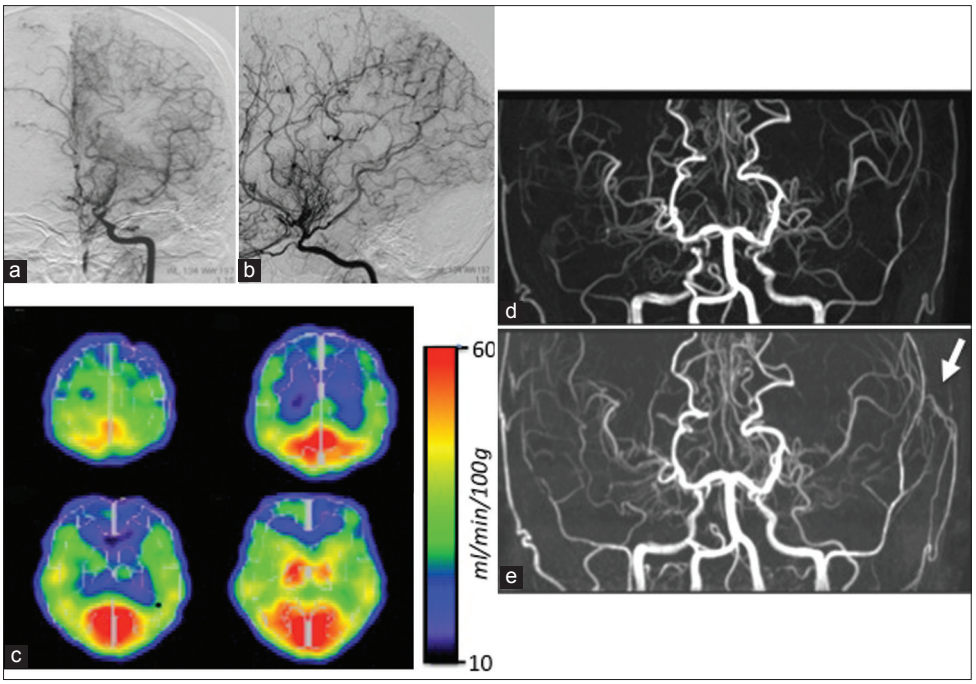

Figure 1: Preoperative neuro-radiologic examinations and postoperative MR-angiography. (a-d) Left internal carotid artery angiograms; (a) anterior-posterior view, (b) lateral view, (c) 123I-IMP SPECT at rest, and (d) MR-angiography. (e) MR angiography on postoperative Day 1 revealed the patent STA-MCA anastomosis (arrow)

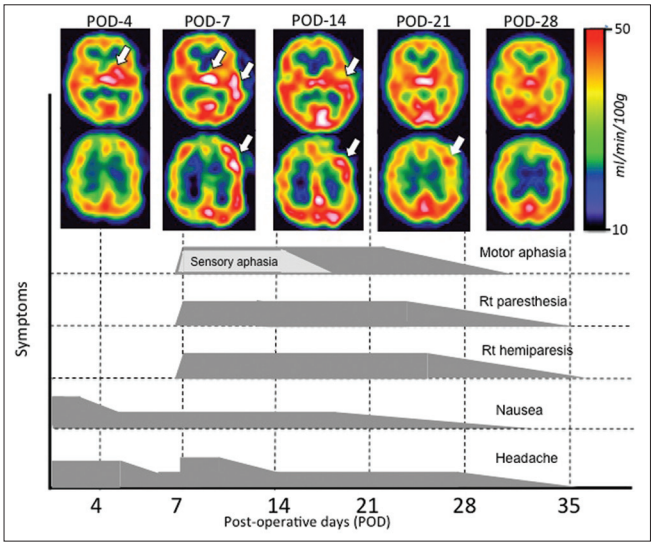

Figure 2: The postoperative clinical course and the changes in CBF detected with 123I-IMP SPECT. POD: postoperative days. Arrows indicate the regions of hyperperfusion. The scale of the x-axis in each symptom revealed two grades; maximum (100% height), and incomplete improvement containing over 1 day (50% height)

headache or other neurologic symptoms.[17,19,20,23] Previous studies have found hyperperfusion after CEA diffusely in all hemispheres, including the basal ganglia, but after STA-MCA anastomosis for MMD, hyperperfusion has frequently been reported in localized cortical areas.[1-5,13,15] Thus, severe headache in this case may be related to basal ganglia hyperperfusion.

Reported risk factors for hyperperfusion include poor cerebrovascular reactivity (CVR), hemorrhagic-onset, adult-onset, and a small recipient MCA (diameter <1 mm).[1,3,4] In this case, three risk factors were present, adult-onset, poor CVR, and a small recipient MCA, resulting in a high-risk of symptomatic hyperperfusion syndrome.

In most patients, the hyperperfusion state, as detected by SPECT, disappeared within 1 week of bypass surgery, although symptoms persisted for 1–2 weeks.[1-5,13,15] One available case report included a symptomatic hyperperfusion syndrome with a long duration of symptoms that persisted for 30 days following surgery.[1] The current case represents the longest reported duration of symptoms, continuing 5 weeks after STA-MCA anastomosis. It is likely that this prolonged symptomatic period was caused by sequentially changing hyperperfusion lesions; occurring first in the basal ganglia followed by a second lesion located in the front parietal region that developed in the second week after surgery and persisted for 3–4 weeks. It is likely that the two sequential lesions were related in that during the early phase after STA-MCA anastomosis, the relatively large amount of blood flow allowed through the 1.3-mm diameter STA into the small M4 branch of the MCA (0.7 mm diameter) rapidly returned to the proximal MCA, which has a lower resistance than that of the distal MCA. The resultant increase in the relative blood flow to the basal ganglia caused a symptomatic cerebral hyperperfusion syndrome. After 7 days, auto-regulation of the M1 and M2 arteries gradually recovered (through recovery of CVR) allowing adequate arterial constriction and causing a change in the direction of blood flow from the large STA to the distal

side of the anastomosis site, which resulted in a front-parietal symptomatic cerebral hyperperfusion syndrome with aphasia, hemiparesthesia, and hemiparesis.

This case represents the first report of a regionally progressive hyperperfusion syndrome, and the exact mechanisms underlying why the specific time courses of each lesion differ remains unknown. This case showed rapid progression of a temporary ischemic attack over the course of several months, with rapidly progressive M1 stenosis, despite a CBF study 4 months prior that showed no obvious decrease in CBF [Figures 1a and 1d]. CVR may change dramatically over several months and it is possible that regional differences exist in the functional recovery of CVR.

Despite the favorable long-term outcome, some authors suggest that focal cerebral hyperperfusion may cause not only transient focal neurological deficit but also intracranial hemorrhage with the potential for development of a permanent deficit.[1,3,8,9,11,14,15] Thus, the establishment of effective treatment for hyperperfusion is needed. This patient was treated with the free radical scavenger, edaravone, and blood pressure control and the patient fully recovered without neurologic deficit. Fujimura *et al.*[1] suggested that production of reactive oxygen species (ROS) after vascular reconstruction may play an important role. In fact, edaravone has been reported to prevent hyperperfusion injury after CEA[12] and after STA-MCA anastomosis in patients with MMD,[1] making ROS an important therapeutic target.

The mechanism and pathophysiology behind hyperperfusion remains unclear and future studies are needed to investigate strategies for prevention of symptomatic cerebral hyperperfusion syndrome.

CONCLUSION

This report details a rare case of a patient with MMD who presented with regionally progressive hyperperfusion lesions after STA-MCA anastomosis and symptoms that persisted for 5 weeks following surgery. Results from this case suggest that regional differences exist in the functional recovery of CVR in a patient with rapidly progressive MMD.

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