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Case Report

Limb-shaking syndrome derived from the contralateral hemisphere following unilateral revascularisation for moyamoya disease

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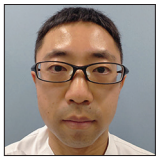

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ABSTRACT

Background: Moyamoya disease is a rare chronic steno-occlusive cerebrovascular disease. It may have variable clinical symptoms associated with cerebral stroke, including motor paralysis, sensory disturbances, seizures, or headaches. However, patients with moyamoya disease rarely present with involuntary movement disorders, including limb-shaking syndrome, with no previous reports of limb-shaking syndrome occurring after revascularization procedures for this disease. Although watershed shifts can elicit transient neurological deterioration after revascularisation, symptoms originating from the contralateral hemisphere following the revascularization procedure are rare. Here, we report the case of moyamoya disease wherein the patient developed limb-shaking syndrome derived from the contralateral hemisphere after unilateral revascularisation.

Case Description: A 16-year-old girl presented with transient left upper and lower limb numbness and headache. Based on digital subtraction angiography, she was diagnosed with symptomatic moyamoya disease. Single-photon emission computed tomography (SPECT) showed decreased cerebral blood flow (CBF) on the right side, and she underwent direct and indirect bypasses on this side. Involuntary movements appeared in her right upper limb immediately postoperatively. SPECT showed decreased CBF to the bilateral frontal lobes. Subsequently, the patient was diagnosed with limb-shaking syndrome. After performing left-hemispheric revascularisation, the patient's symptoms resolved, and SPECT imaging confirmed improvements in CBF to the bilateral frontal lobes.

Conclusion: Revascularization for moyamoya disease can lead to watershed shifts, which can induce limb-shaking syndrome derived from abnormalities in the contralateral hemisphere of the revascularized side. For patients with new-onset limb-shaking syndrome after moyamoya revascularisation procedures, additional revascularization may be warranted for treatment of low perfusion areas.

Keywords: Limb-shaking syndrome, Moyamoya disease, Postoperative involuntary movement, Watershed shift

INTRODUCTION

Moyamoya disease is characterized by progressive steno-occlusive changes at the terminal portions of the internal carotid artery (ICA) and the development of moyamoya vessels. Patients with moyamoya disease usually present with clinical symptoms associated with ischaemic attacks and intracranial hemorrhages, including motor paralysis, sensory disturbances, seizures, or headaches.[6,10,16] Involuntary movement disorders, such as limb-shaking syndrome, are more

rarely observed in patients with moyamoya disease.[10] These symptoms may improve after revascularization.[6]

Although the most effective treatment for moyamoya disease is revascularization surgery, postoperative complications such as ischemic/hemorrhagic stroke or cerebral hyperperfusion syndrome have been reported.[2,7] It is known that patients occasionally experience transient neurological deterioration after revascularization procedures for moyamoya disease;[10] however, there have been no reports of limb-shaking syndrome appearing after these surgeries. In addition, the symptoms of this syndrome have never been reported to originate from the contralateral hemisphere of the revascularized side.

Herein, we present the case of limb-shaking syndrome derived from the contralateral hemisphere after a unilateral revascularisation procedure for moyamoya disease.

CASE REPORT

A 16-year-old girl presented with transient left upper and lower limb numbness and headache while participating in chorus and sports. She was referred to our department for surgical treatment. She had a past medical history of immunoglobulin A nephropathy and was being treated with steroids. Magnetic resonance (MR) images showed no ischaemic changes in her brain, but MR angiography showed intracranial vascular stenosis [Figure 1a]. Digital subtraction angiography showed bilateral stenoses from the terminal portion of the ICA to the proximal parts of the middle cerebral and anterior cerebral arteries [Figure 1b]. Extended perforating branches were also noted. Single-photon emission computed tomography (SPECT) imaging showed decreased cerebral blood flow (CBF) on the right side [Figure 2a], which worsened after an acetazolamide challenge [Figure 2b]. Based on these findings, she was diagnosed with Suzuki Stage 2 moyamoya disease bilaterally[13] and underwent revascularisation on the symptomatic right side.

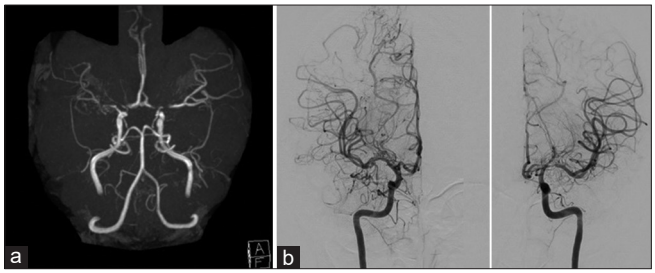


Figure 1: Patient angiography findings at initial presentation. (a) Intracranial vascular stenosis was identified on magnetic resonance angiography. (b) Digital subtraction angiography suggested bilateral stenoses from the terminal portion of the internal carotid artery to the proximal parts of the middle and anterior cerebral arteries, with extended lenticulostriate arteries.

During surgery, both direct and indirect bypasses were successfully performed; however, involuntary movements appeared immediately after the operation, with her right upper limb episodically shaking at a frequency of 3 to 5 Hz [Video 1]. These episodes lasted for a few minutes up to a few hours, with no associated disturbances in her level of consciousness and no incontinence. The involuntary movements were limited to the right upper limb and did not occur during sleep. On the next day, MR angiography showed good patency of bypass vessels, and MR images showed neither cerebral infarction nor hemorrhage [Figure 3a and b]. Electroencephalography (EEG) showed no abnormal brainwaves suggestive of epilepsy. SPECT performed on postoperative day 3 revealed decreased CBF in the bilateral frontal lobes [Figure 4].

The frequency of her involuntary movements gradually decreased, and SPECT imaging showed slight improvement of the hypoperfusion in her bilateral frontal lobes; however, this symptom and her abnormal imaging findings persisted, though to a lesser degree. Therefore, a left-sided revascularisation procedure was performed on postoperative day 47. The following day, her symptoms disappeared and did

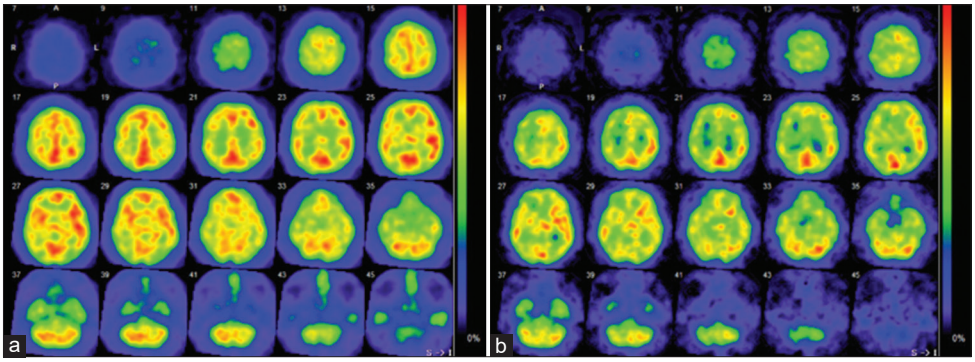


Figure 2: Preoperative single-photon emission computed tomography (SPECT) imaging. (a) N-isopropyl-p-[123I] iodoamphetamine SPECT showed decreased cerebral blood flow (CBF) in the right hemisphere at rest, preoperatively. (b) Paradoxical reduction of CBF after acetazolamide administration was identified in the right hemisphere before surgery.

not recur since then. Furthermore, blood flow in the bilateral frontal lobes was improved according to SPECT imaging performed 7 days after the second surgery [Figure 4].

DISCUSSION

Involuntary movements are rarely associated with moyamoya disease. Pandey *et al.*[10] reported that the frequency of involuntary movements in moyamoya disease was 4 out of 446 (0.89%) in the total study population and 4 out of 118 (3.4%) in children. Limb-shaking syndrome associated with moyamoya disease was reported to be present preoperatively in only a few previous cases.[4,8,10] Contrastingly, in our case, the patient's involuntary movements developed after a revascularization procedure. These symptoms were limited to the upper right limb and had an irregular rhythm, unlike a tremor, ballismus, or chorea. She did not display any disturbances in consciousness and could voluntarily move the affected arm. In addition, her symptoms did not occur during sleep. Based on these clinical features, EEG findings, and MR images, we diagnosed limb-shaking syndrome rather than focal motor seizures in the patient.

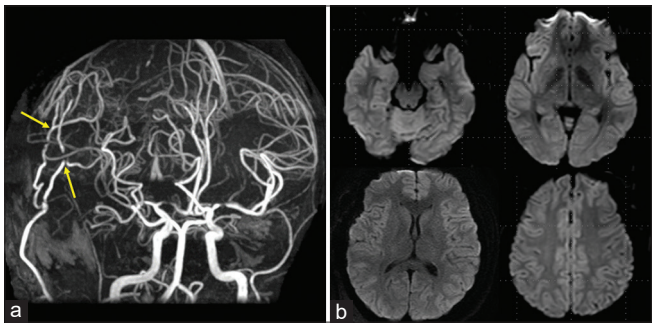


Figure 3: Postoperative magnetic resonance (MR) studies. (a) MR angiography on postoperative day 1 showed patency of the right superficial temporal artery to the middle cerebral artery anastomosis (arrows). (b) MR images showed no ischaemic lesions.

Limb-shaking syndrome is an involuntary movement disorder first reported by Fisher in 1962.[1] Symptoms include hyperkinesia and flailing and jerking movements, often lasting for several minutes.[1] These movements are usually limited to one limb and do not generally spread to other body parts. The duration of symptoms tends to be short, and the movements are commonly coarse (3–5 Hz).[6,11,14] Moreover, no abnormalities are identified on EEG, and antiepileptic drugs are ineffective.[12]

Rather than an epileptic disorder, limb-shaking syndrome is considered an ischaemic disorder caused by ICA occlusion or stenosis.[4-6,8,9,11,12,14] In general, involuntary movements tend to occur when portions of the basal ganglia, including the hypothalamic nucleus or the pallidus, are directly impaired. However, limb-shaking syndrome is believed to result from ischemia in the frontal lobe cortex rather than from damage to the basal ganglia.[10,15] In this case, although preoperative

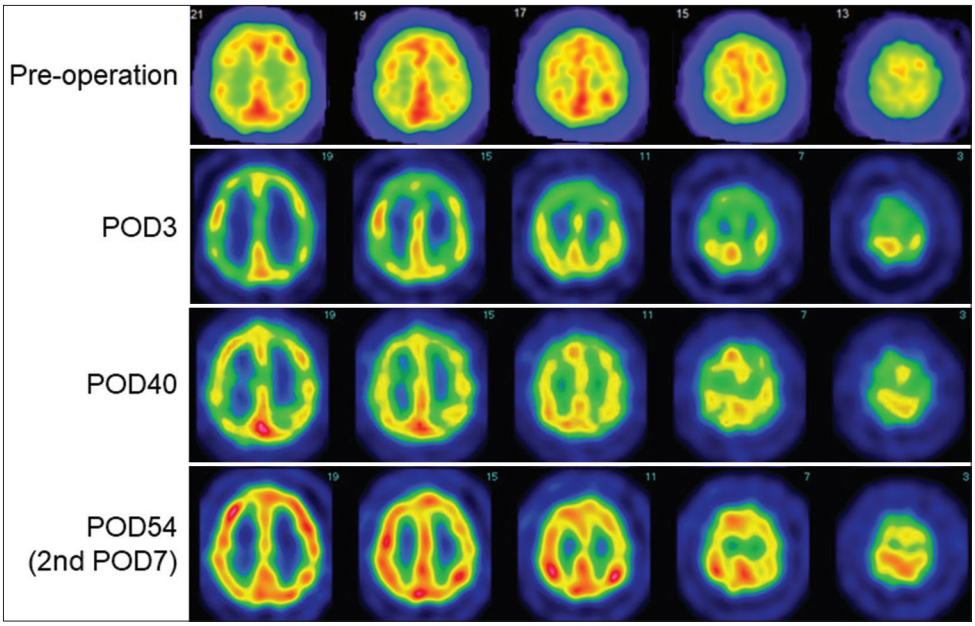
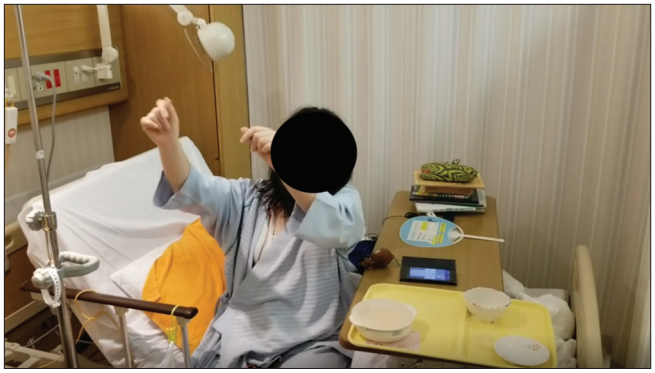


Figure 4: Postoperative single-photon emission computed tomography (SPECT). Technetium-99m ethyl cysteinate dimer (99mTc-ECD) SPECT on postoperative day (POD) 3 did not show hyperperfusion in the right hemisphere but did show decreased cerebral blood flow (CBF) in the bilateral frontal lobe areas. Forty days after surgery, hypoperfusion of bilateral frontal lobes on SPECT was slightly improved but remained to a lesser extent. Seven days after the second revascularisation procedure, 123I-IMP SPECT showed improvement of whole-brain CBF, including in both frontal lobes.



Video 1: Involuntary movement of her right upper limb shaking at a frequency of 3 to 5 Hz.

SPECT showed reduced blood flow to the right hemisphere, SPECT performed 3 days after the initial operation showed decreased blood flow to both frontal lobes.

The watershed-shift phenomenon has recently been shown to cause transient neurological deterioration after revascularization procedures for moyamoya disease. This phenomenon was defined by Hayashi *et al.*[3] as the induction of new cerebral infarcts away from the anastomotic site after revascularization surgery. Strokes associated with watershed shifts occur in 1.2% to 5.7% of all patients who undergo bypass treatment for this disease.[9] Tu *et al.*[17] reported two cases that developed remote infarctions due to watershed shifts after revascularization for moyamoya disease. SPECT studies of these patients showed a marked increase of CBF at the area of anastomosis, with a paradoxical decrease in CBF in the surrounding areas. When revascularization procedures change CBF, low perfusion areas may be shifted, and the resultant blood flow changes may worsen symptoms. In our case, the paradoxical decrease in postsurgical CBF was evident not only in the right frontal lobe where the direct bypass was performed but also in the contralateral frontal lobe.

Our case suggests that watershed shifts occurring after unilateral revascularization procedures in patients with moyamoya disease may lead to perfusion changes in the contralateral hemisphere and may result in limb-shaking syndrome. In these cases, an additional bypass surgery may be needed as soon as possible.

CONCLUSION

Revascularization for moyamoya disease may cause limb-shaking syndrome by inducing CBF reductions in the contralateral frontal lobe due to watershed shifts. For patients with moyamoya disease who develop new involuntary movements after surgery, additional revascularization procedures may be warranted in areas where CBF reduction is confirmed using SPECT.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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