



Case Report

Truncal dystonia with isolated middle cerebral artery ischemia: A case report of revascularization therapy for dystonia

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ABSTRACT

Background: Dystonia is a rare movement disorder with some cases being difficult to treat. Although dystonia can occur as a symptom of moyamoya disease, few studies have reported truncal dystonia occurring with middle cerebral artery (MCA) stenosis. Here, we report a case of truncal dystonia with MCA occlusion.

Case Description: The patient was a 48-year-old female clerical worker who lived alone. An abnormal cervical posture initially appeared 7 years before (right flexion). Symptoms improved with medication and botulinum toxin injection. Five years before this report, her symptoms worsened, so the dose of oral medication was increased and botulinum treatment was performed, but the symptoms did not improve. The patient showed decreased cerebral blood flow (CBF) in the cortical areas but not in the basal ganglia. We performed superficial temporal artery-MCA bypass surgery because we believed that the dystonia was due to right MCA stenosis. The patient's symptoms improved immediately after surgery, except for her mild cervical backbend. Seven months after the surgery, the patient's involuntary movements showed further improvement, and symptoms have not worsened even after 2 years.

Conclusion: Revascularization therapy improved CBF and truncal dystonia and could be a viable treatment option for dystonia with ischemia in the MCA region. Extensive cerebral ischemia can result in cortical inhibition loss or over-adapted cerebral plasticity and cause dystonia. Revascularization therapy may be useful for patients with dystonia and decreased CBF in the MCA region.

Keywords: Cerebral blood flow, Dystonia, Middle cerebral artery stenosis, Network pathophysiology model, Revascularization

INTRODUCTION

Dystonia is a clinical syndrome characterized by sustained muscle contractions that cause twisting, repetitive movements, or abnormal postures.^[6] Dystonia can cause functional impairments that may affect an individual's daily activities and ability to work.^[14] Cerebrovascular diseases represent up to 22% of secondary movement disorders, and involuntary movements develop in 1–4% of cases

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after stroke. Some of these disorders occur immediately after acute stroke. Others can develop later, which are represented as delayed-onset progressive movement disorders.^[4,15]

Involuntary movements can be diagnosed based on clinical symptoms. However, there are various causes, and it may be difficult to select a treatment option. Involuntary movements, such as dystonia and ballism, occur as symptoms of moyamoya disease.^[8,9,12,20,23] There are few reports of involuntary movement occurring in association with middle cerebral artery (MCA) stenosis.^[2,19] We report a case of truncal dystonia with decreased cerebral blood flow (CBF) in the MCA region, in which symptoms improved after revascularization therapy.

CASE DESCRIPTION

The patient was a 48-year-old female clerical worker who lived alone. An abnormal cervical posture initially appeared 7 years before this study (right flexion). The treatment included the prescription of trihexyphenidyl hydrochloride (6 mg), clonazepam (1.5 mg), and baclofen (15 mg), which was effective and resulted in good outcomes. Botulinum treatment was also prescribed and effective, and the symptoms almost disappeared. Five years before this study, the abnormal cervical posture worsened, and symptoms of cervical retroflexion and trunk anteflexion developed. Doses were increased (trihexyphenidyl hydrochloride, 12 mg; clonazepam, 3 mg; and baclofen, 30 mg); however, there was no improvement. Botulinum treatment was also ineffective. During the same period, the right MCA stenosis was observed on head magnetic resonance angiography (MRA). The patient was referred to our hospital and her medical history revealed a persistent depressive disorder. The patient had been in a psychiatric clinic and treated since the age of 22 years. Her current doctor has been treating her since she was 30 years old. Her mother, who had already died, had a delusional disorder. The patient has lived with her mother since childhood. When she was 17 years old, she wanted to go to college because her academic performances were good; however, she dropped out of high school at the request of her mother. Her energy was often exhausted partly because of frequent relocations due to neighborhood troubles caused by her mother, who died when she was 42 years old. She had little interpersonal interaction and often failed when she tried to interact. She frequently made damaging remarks and was often allowed to make detrimental statements about it. She often stayed at home because of her interpersonal relationships and fear of going out. Based on the above-mentioned course and no psychiatric symptoms that could be concluded as delusions, her psychiatric symptoms were judged to be mainly depressed, and she was treated internally as a persistent depressive disorder. She was prescribed propriciazine (15 mg), ramelteon (8 mg), etizolam (2 mg), brotizolam (0.25 mg), mirtazapine (15 mg),

nortriptyline hydrochloride (30 mg), levomepromazine hydrochloride (15 mg), and eszopiclone (2 mg) when she visited our hospital. She consumed beer daily (700 mL/day) but had not smoked for 10 years.

Involuntary neurological movements, including cervical back bending and truncal forward bending, were observed while walking [Supplementary Video 1]. No other neurological focal signs were noted. Her symptoms impaired her ability to work and perform daily activities. However, her symptoms improved while walking backward, in line with task specificity, in which involuntary movements of the trunk occurred only when walking forward. The patient's blood examination findings, including antinuclear antibody, anti-SS-A antibody, anti-SS-B antibody, anti-glutamic acid decarboxylase antibody, hemoglobin A1c, and ceruloplasmin, were normal. Involuntary movements associated with autoimmune (e.g., stiff person syndrome) or metabolic diseases (e.g., Wilson disease) were not observed. Findings on spinal radiographs were normal and genetic testing was negative for known hereditary dystonia-related genes.

When the patient visited our hospital, MRA revealed the right MCA occlusion [Figures 1 and 2]. In our hospital,

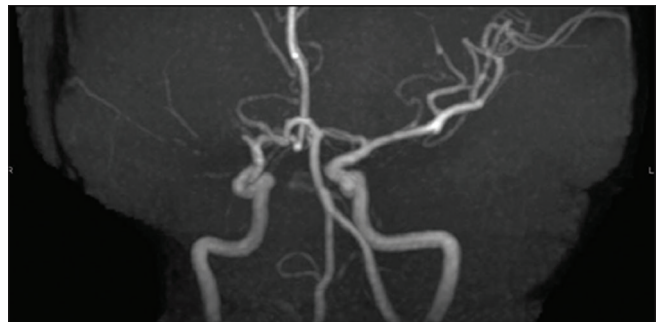


Figure 1: Upper part: magnetic resonance angiography (MRA). Before surgery: MRA showing the occlusion of the right middle cerebral artery (MCA).

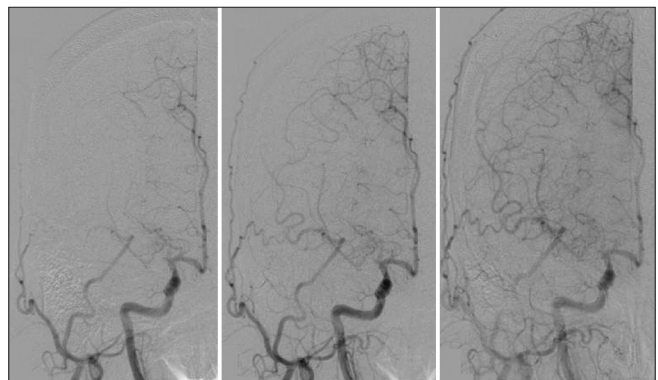


Figure 2: Angiography showing almost complete occlusion of the right horizontal segment of the middle cerebral artery and development of collateral circulation through pial anastomosis from the anterior cerebral artery.

perfusion computed tomography (CT) is used to assess CBF in six regions of interest in the cerebral cortex. In this case, we obtained the ratio of the right CBF to the left CBF in four regions, excluding the frontal and occipital regions [Figure 3a]. In addition, we examined the laterality of blood flow in the basal ganglia [Figure 3b]. Perfusion CT revealed that the CBF on the right side was 64% and 94% of that on the left side in the cortical areas and basal ganglia, respectively [Figure 4]. Digital subtraction angiography revealed almost complete occlusion of the right horizontal segment of the MCA as well as collateral circulation through the pial anastomosis from the anterior cerebral artery [Figure 2].

We performed superficial temporal artery (STA)-MCA bypass surgery because we believed that the dystonia was

due to right MCA stenosis [Figure 5]. The patient's symptoms improved immediately after the surgery, except for her mild cervical backbend [Supplementary Video 2]. Perfusion CT immediately after the surgery showed that the CBF on the right side was 95% and 97% of that on the left side in the cortical areas and basal ganglia, respectively. Furthermore, perfusion CT performed after 6 months showed that the CBF on the right side was 92% and 108% of that on the left side in the cortical areas and basal ganglia, respectively. The right MCA was assessed using MRA 6 months after the operation [Figure 6]. Seven months after the surgery, the patient's involuntary movements showed further improvement [Supplementary Video 3], and symptoms have not worsened 2 years later. She had been treated for persistent depressive disorder for a long time; however, changes in psychiatric symptoms were observed 2 years after the operation. Verbosity, the ease with which topics change and the rise in energy when speaking have begun to show a clear rise in mood, although it is not difficult to stop. Therefore, nortriptyline hydrochloride (30 mg) is currently discontinued, and the patient is being followed up.

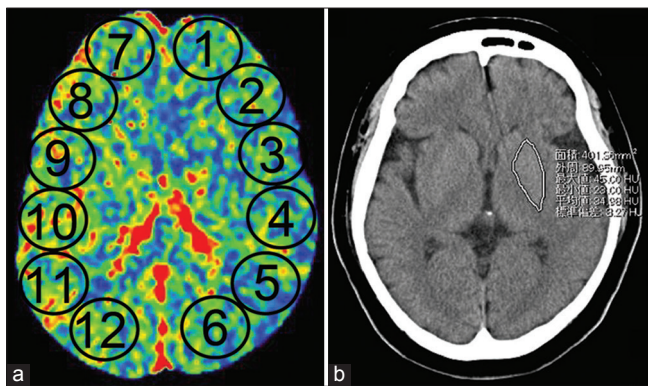


Figure 3: (a) Region of interest for cerebral blood flow measurement using perfusion computed tomography. Comparison of the average cerebral blood flow in ②, ③, ④, and ⑤ (left MCA territory) with that in ⑧, ⑨, ⑩, and ⑪ (right MCA territory). (b) Region of interest for blood flow measurement in the basal ganglia through perfusion computed tomography. Tracing of the basal ganglia and comparison of the blood flow on the left and right sides.

DISCUSSION

We encountered a case of truncal dystonia associated with decreased MCA blood flow. Few reports of revascularization for involuntary movements associated with decreased MCA blood flow exist. Chung *et al.* reported a case of a 26-year-old woman with involuntary movements of the right upper and lower limbs with 48% stenosis in the MCA. In this case, the MCA was completely occluded 1 year later, and the symptoms disappeared after bypass surgery.^[2] In contrast, Shibata *et al.* reported a case of ballismus after STA-MCA anastomosis. They suggested that the ability to autoregulate

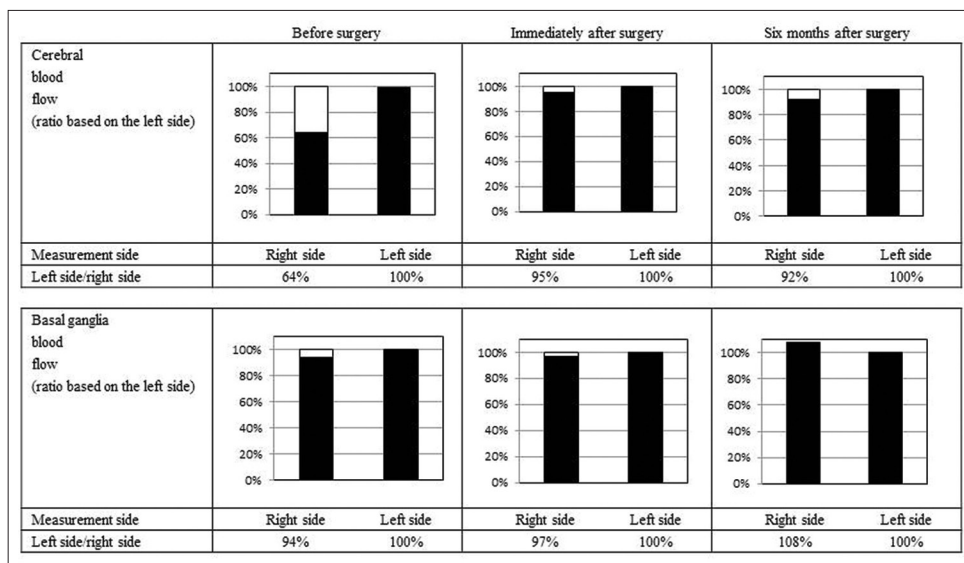


Figure 4: Perfusion computed tomography findings.

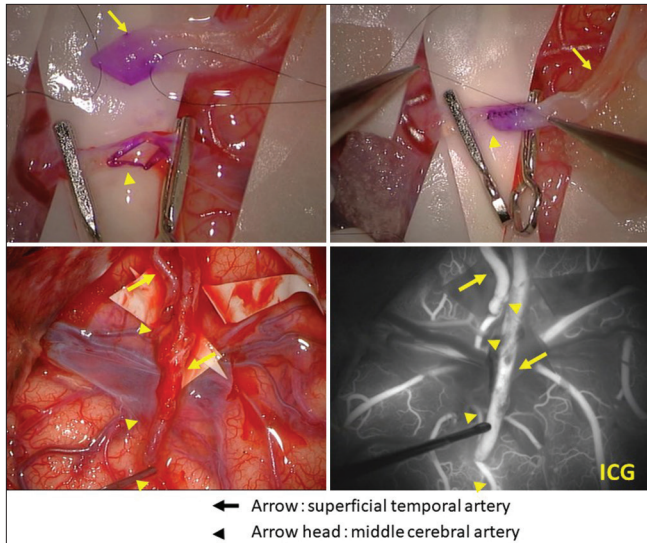


Figure 5: Intraoperative photographs. ICG: Indocyanine green.

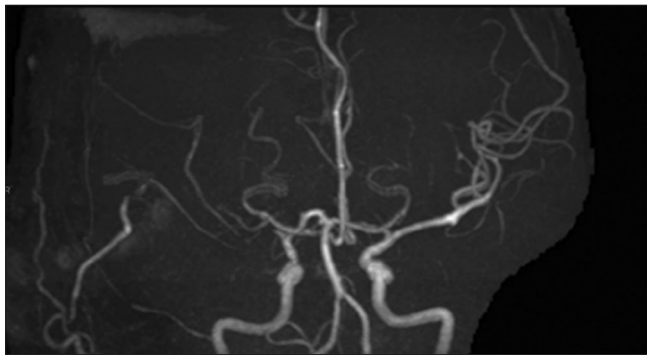


Figure 6: Six months after bypass surgery: the MCA with the intact bypass. Lower part: perfusion computed tomography.

CBF may have been disrupted after anastomosis, resulting in metabolic abnormalities in the striatum and subthalamic nucleus.^[19] We should carefully consider how to treat stenosis of the MCA in clinical cases of truncal dystonia.

Dystonia may be characterized by two main pathophysiological abnormalities: reduced excitability of inhibitory systems at many levels of the sensorimotor systems and increased plasticity of neural connections in sensorimotor circuits, resulting in overall cortical hyperexcitability.^[11,17] A widespread decrease in CBF may cause cerebral over-adapted plasticity or loss of cortical inhibition, resulting in involuntary movements.

Similar conditions have been reported for dystonia, hemiballismus, and limb shaking associated with moyamoya disease.^[13] In addition, some reports have pointed out that involuntary movements improved after vascular reconstruction for moyamoya disease.^[8,9,12,20,23] From these reports, a consensus has been established that surgery is indicated for limb shaking and hemiballismus associated with cerebral ischemia.^[3]

The indications for bypass surgery for MCA stenosis during acute cerebral ischemia are controversial. The JET study in Japan has reported that symptomatic cases with a CBF of 80% or less are indicated for surgery.^[10] However, there are no clear criteria for chronic MCA stenosis diagnosis. In our hospital, CBF is assessed using perfusion CT. Since assessment of CBF through perfusion CT also shows blood flow in blood vessels, its accuracy is limited. In this case, CBF on the affected side was reduced to 64% of that on the healthy side.

We performed revascularization and obtained good clinical outcomes; however, the pathophysiology of dystonia secondary to MCA ischemia is unknown. The pathophysiology of dystonia is being investigated by neurophysiological methods because there are few pathological reports, and neuronal loss is rarely reported.^[7,24] The benefits of thalamotomy,^[1] pallidotomy,^[16] and basal ganglia deep brain stimulation^[22] support the hypothesis that dystonia is caused by basal ganglia dysfunction. Functional magnetic resonance imaging and positron emission tomography studies have also supported the dysfunction of the basal ganglia in dystonia.^[5] However, lesions of the basal ganglia alone cannot fully explain the pathophysiology of dystonia because dystonia may also be caused by lesions other than those of the basal ganglia.^[18,21] Thus, a network pathophysiological model has been proposed. The network disorder involves the motor cortex, where motor commands are generated, as well as the basal ganglia and cerebellum, which allow for smoother and more coordinated movements.^[18] Involuntary movements due to MCA stenosis may be caused by a network of disorders (plastic disorders) that cause functional deterioration due to widespread blood flow reduction in the brain. This deterioration may not be associated with functional deterioration of the basal ganglia due to decreased blood flow in the MCA alone.

Here, we reported a case of truncal dystonia with MCA occlusion that improved with revascularization. On the other hand, this case has a long-term persistent depressive disorder, and the relationship between involuntary movements and persistent depressive disorder cannot be completely ruled out, which is a limitation. Revascularization therapy may be a treatment option for dystonia with ischemia in the MCA region, but further studies may be needed to determine the correct treatment protocol.

CONCLUSION

Revascularization therapy may be a treatment option for dystonia with ischemia in the MCA region.

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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SUPPLEMENTARY VIDEOS



Supplementary Video 1: Before treatment, involuntary movements of cervical back bending and truncal forward bending are seen while walking.



Supplementary Video 2: Immediately after bypass surgery, involuntary movements of truncal forward bending show improvement.



Supplementary Video 3: Seven months after bypass surgery, involuntary movements of cervical back bending and truncal forward bending show improvement.