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## A case of moyamoya disease presenting with chorea

Received: 4 August 2004  
Published online: 30 April 2005  
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**Abstract** *Case report:* We describe a 15-year-old girl with moyamoya disease whose initial manifestation was chorea-like involuntary movements. T2-weighted magnetic resonance imaging showed high signal intensity lesions in the left frontal lobe, right parieto-occipital lobes, and frontal subcortical white matter. Single-photon emission computed tomography (SPECT) showed diffuse hypoperfusion of the whole brain. Bilateral direct and indirect cerebrovascular bypass surgeries were performed. Chorea disappeared 2 days after the surgery. Follow-up SPECT demonstrated

increased cerebral perfusion in the bilateral frontal, temporal, and parietal regions. *Conclusions:* Chorea accompanied with moyamoya disease can be properly managed by revascularization surgery. Moyamoya disease should be remembered as being one of the differential diagnoses of chorea, which is treatable by surgery.

**Keywords** Moyamoya disease · Involuntary movements · Subcortical white matter · Surgical treatment · Chorea · Ischemia · Cerebral perfusion

### Introduction

Moyamoya disease is known to produce variable neurological symptoms. It tends to manifest as a transient ischemic attack or seizure in children, whereas the hemorrhagic stroke is more common in adult patients. Among other miscellaneous symptoms, the involuntary movements of chorea have been known to occur in 3–6% of patients with moyamoya disease [1]. However, this combination has rarely been reported and the mechanism of these involuntary movements accompanied by moyamoya disease and the effect of revascularization surgery have been investigated very little.

In this case report, we describe a 15-year-old girl with moyamoya disease whose initial manifestation was chorea-like involuntary movements and in whom revascularization surgery was quite effective. The mechanism of chorea due to moyamoya disease and the effect of the revascularization surgery on chorea are discussed.

### Case report

A 15-year-old right-handed girl was observed throwing pens or chopsticks unwarily without any inducement. Several days later, she noticed the involuntary movements of her right arm extending and bending back and forth occasionally. Ten days later, the same symptoms had extended to all four limbs.

Physical examination on admission revealed occasional involuntary movements in all four limbs. Neurological examination was unremarkable except for left inferior homonymous quadrantanopia. The high mental function examination WISC-R ranked 58, which was lower than the normal level. Her father had onset of transient ischemic attacks (TIA) that was attributed to moyamoya disease when he was 35 years old. Her elder sister had undergone an operation for a scalp arteriovenous malformation (AVM).

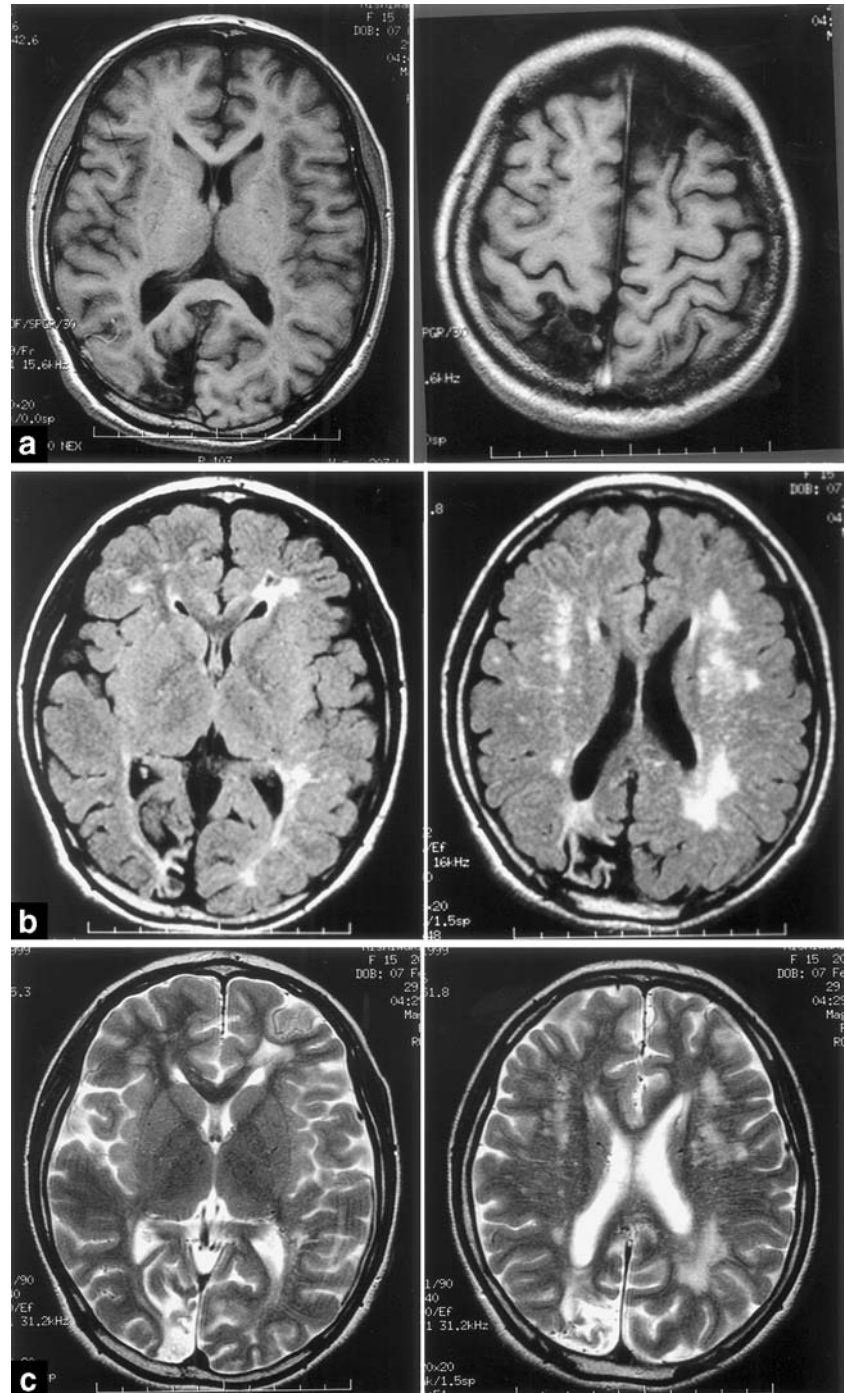
Magnetic resonance imaging (MRI) of the brain showed high intensity lesions on a T2-weighted image in the left

frontal lobe, right parieto-occipital lobes, and frontal sub-cortical white matter. No significant intensity change was observed on the T1-weighted images, the T2-weighted images, and on the water suppressed proton weighted images of the bilateral basal ganglia and thalamus (Fig. 1). Diffusion-weighted imaging was not performed. These MR images suggested that the lesion was mainly the cerebral infarction of the cortex and white matter, but not the basal

ganglia. The possibility of some degenerative disease could not be fully ignored, but the following angiographical findings supported the suggestion that the ischemic lesion was due to moyamoya disease.

On the digital subtraction angiography (DSA) of the bilateral carotid and vertebral arteries, the bilateral anterior cerebral artery (ACA), and the middle cerebral artery (MCA) could not be identified. Numerous small collater-

**Fig. 1** **a** T1-weighted magnetic resonance images reveal cortical infarction. **b** Water suppressed proton images (FLAIR) reveal no abnormal intensity in the basal ganglia and an abnormally high signal lesion in the sub-cortical white matter. **c** T2-weighted magnetic resonance images reveal similar findings to those seen on the FLAIR images

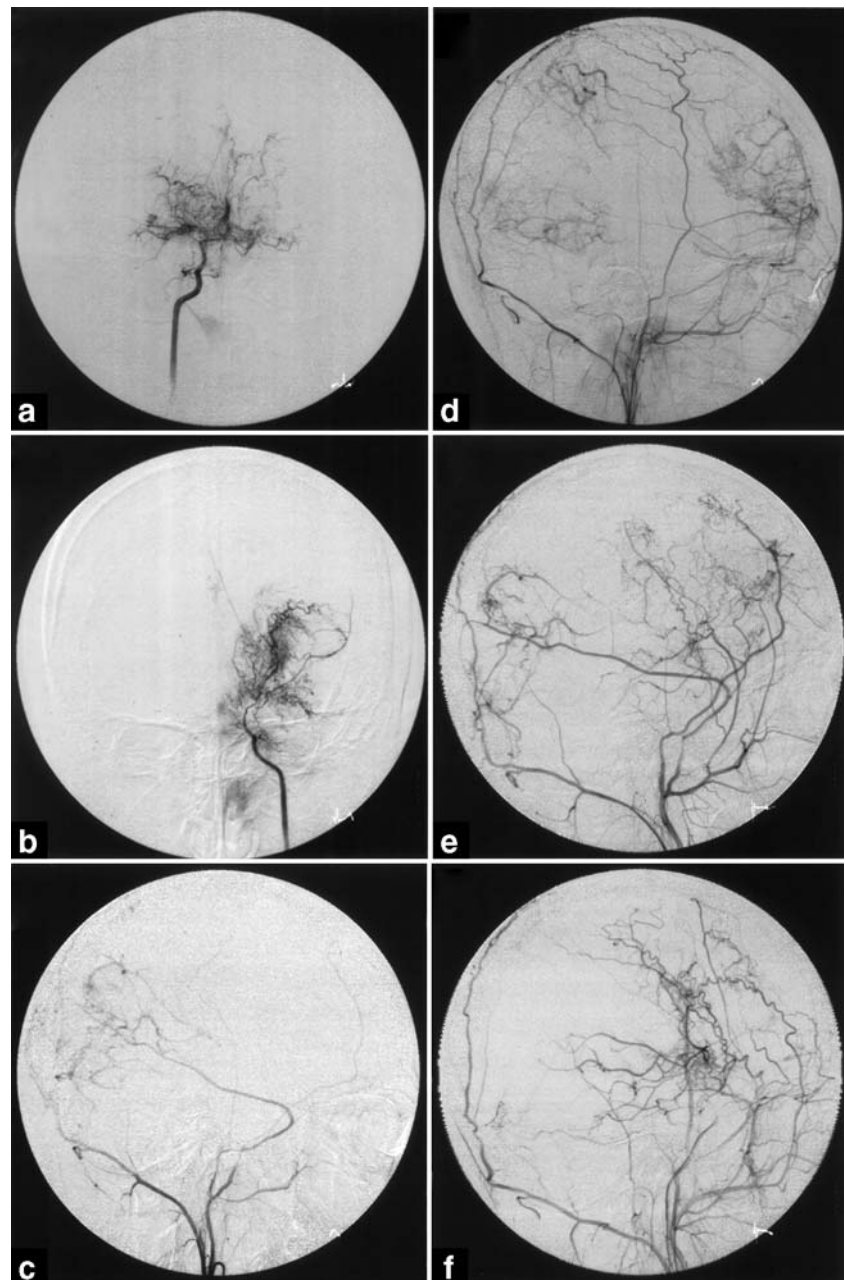


al vessel networks were found at the base of the brain (Fig. 2a, b), collateral flow from the occipital artery and the internal maxillary artery to the internal carotid regions (Fig. 2c, d). *N*-Isopropyl-*p*-(I-123) iodoamphetamine ( $^{123}\text{I}$ -IMP) single-photon emission computed tomography (SPECT) showed a focal perfusion defect in the left frontal and right occipital regions. The quantitative study showed diffuse hypoperfusion within the whole brain (Fig. 3a). The mean cerebral blood flow at the level of the body of the ventricle was 33.2 ml/100 g/min on the right side and 32.2 ml/100 g/min on the left side.

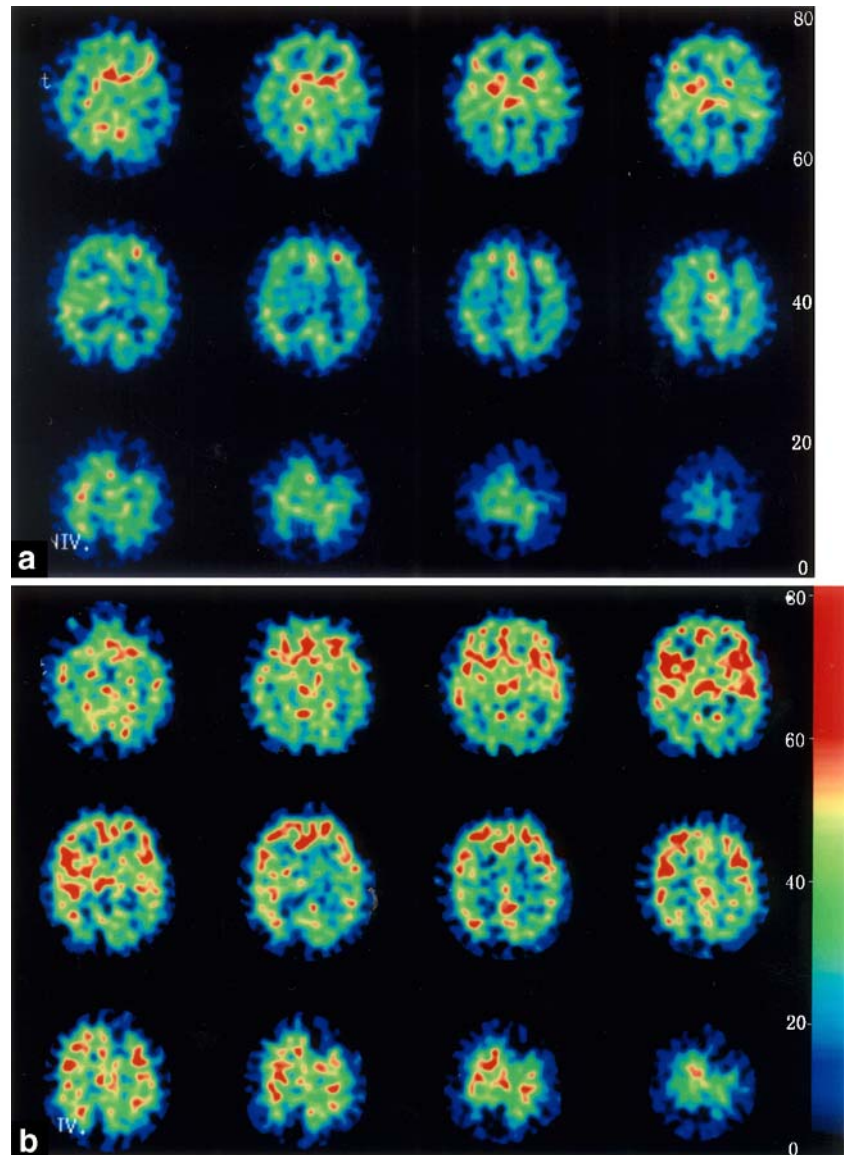
Two weeks after admission, a right superficial temporal artery–middle cerebral artery (STA–MCA) bypass and encephalo-duro-arterio-myosynangiosis (EDAMS) were performed. The same surgery was performed on the other side 3 weeks later. The chorea disappeared 2 days after the first operation. One week after the second operation, 100 mg aspirin was taken per day as medical therapy.

The follow-up DSA 6 months after the operation revealed MCA and anterior cerebral artery regions were filled via the STA and the deep temporal artery (Fig. 2e, f). The SPECT demonstrated that cerebral perfusion had

**Fig. 2** a, b Preoperative angiography of the internal carotid arteries showed stenosis of both internal carotid arteries at the supraclinoid portion and numerous collateral vessel networks at the base of the brain. c, d Preoperative angiography of the bilateral external carotid arteries showed collateral flow from the occipital artery and internal maxillary artery to the internal carotid regions. e, f Postoperative angiography of the bilateral external carotid arteries showed that collateral flow increased markedly



**Fig. 3** Quantitative cerebral blood flow images (SPECT). **a** Diffuse cerebral hypoperfusion in the whole brain before surgery. **b** Cerebral perfusion increased markedly within the whole brain after surgery



increased in the bilateral frontal, temporal and parietal regions (Fig. 3b). The mean cerebral blood flow increased to 43.5 ml/100 g/min on right side and 43.2 ml/100 g/min on left side, which were significant increases compared with the preoperative values. The high WISC-R score measuring mental function improved from 58 to 65. The involuntary movements have not recurred for 2 years although MRI revealed no significant change.

## Discussion

Chorea is an uncommon abnormal movement that can be seen in a wide range of disorders. These diseases are classified as follows: inherited disorders, drug-induced diseases, systemic diseases, rheumatic diseases, vascular-related dis-

eases, tumor-related diseases, and other rare causes [2]. The probable pathophysiologic mechanism of chorea seems to result from under-activity in the indirect pathway from the basal ganglia-thalamocortical motor circuits [3]. Although there are obvious degenerative lesions in the caudate and putamen in Huntington's disease, the inherited disorders that are characterized by chorea, ischemic dysfunctions of the basal ganglia, cortex, subcortical white matter, thalamus, and subthalamic nucleus, have been suggested as causes of many subtypes of involuntary movements [4, 5].

The common clinical presentations of pediatric moyamoya disease are transient ischemic attacks that are typically induced by hyperventilation, epileptic seizures, and migraine-like headaches. Involuntary movement disorder including chorea is not always a common clinical presentation in moyamoya disease. However, recently, there

have been several reports describing cases of moyamoya disease presenting with movement disorder [6–13] and it has been known to occur in 3–6% of patients. In most reported cases of chorea with moyamoya disease, the ischemic changes in the basal ganglia-thalamocortical circuits are considered to be responsible for involuntary movements including chorea, although some papers revealed that cortical ischemic lesions are responsible for chorea [14–19]. It is well known that cerebral ischemia is generally an important cause of some movement disorders. In our case, the MRI of the brain showed high signal intensity lesions on T2-weighted images in the left frontal lobe, right parieto-occipital lobes, and frontal subcortical white matter, but showed no significant ischemic change in the bilateral basal ganglia. This suggests that any interruption of the basal ganglia-thalamocortical circuits might cause the involuntary movements.

The surgical therapy for moyamoya disease can be categorized as direct revascularization, indirect revasculariza-

tion, or a combination of both. It is reported that combined surgery (STA–MCA bypass with EDAMS) for pediatric moyamoya disease was effective in reducing the risk of postoperative ischemic attacks compared with indirect surgery [20]. In the present case, we performed an STA–MCA bypass combined with EDAMS bilaterally. The involuntary movements disappeared 2 days after the first operation. The follow-up DSA showed that MCA and ACA regions were filled through the external carotid system. The SPECT revealed remarkable perfusion improvement in the subcortical white matter. These results suggest that proper surgical treatment can increase cerebral perfusion, improve clinical symptoms, and prevent the development of the disease.

**Acknowledgements** This work was supported in part by a grant from the Research Committee on Spontaneous Occlusion of the Circle of Willis sponsored by the Ministry of Health and Welfare of Japan.

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