

# Reversible striatal hypermetabolism in chorea associated with moyamoya disease: a report of two cases

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## Abstract

**Background** The pathophysiological mechanism of chorea as a presentation of pediatric moyamoya disease remains unknown, although ischemia is suspected as a likely cause. The authors describe two cases of pediatric moyamoya disease, both of which presented with hemichorea in the stable phase after successful bypass surgery.

**Clinical Presentation** Cerebral blood flow was almost normal in one case and decreased in the basal ganglia and watershed area in the other case due to infarcts occurring before surgery. In both cases, <sup>18</sup>F-fluorodeoxyglucose positron emission tomography revealed elevated glucose metabolism in the corresponding side of the striatum, which reverted to normal after recovery from chorea. Magnetic resonance angiography revealed a dilated and extended lenticulostriate artery at the exact site of the hypermetabolic lesion.

**Keywords** Moyamoya disease · Chorea · Direct bypass

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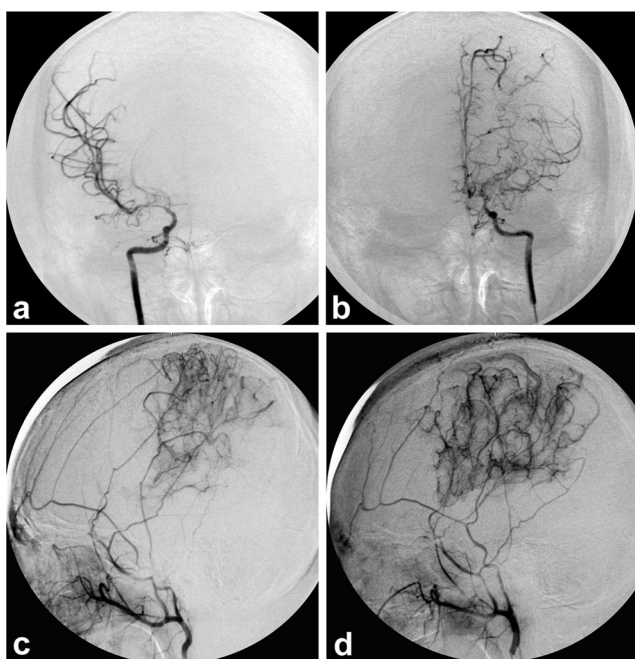
## Introduction

Moyamoya disease is characterized as chronic progressive stenosis of the terminal portion of the bilateral internal carotid arteries and development of abnormal collaterals [22]. The various manifestations of the disease include transient ischemic attack, ischemic and hemorrhagic stroke, epileptic seizure, and headache. Movement disorders such as chorea are an uncommon presentation of pediatric moyamoya disease [1, 2, 11, 15–18, 21, 23, 25]. Because such chorea typically improves after bypass surgery, some researchers have postulated ischemia in certain parts of the brain, including the basal ganglia, as a cause of chorea in moyamoya disease [11, 14, 15, 21, 23]. Neither the pathophysiological mechanism nor the origin of such chorea has been clarified, however. We describe two cases with moyamoya disease presenting with hemichorea in the stable phase after successful direct bypass in which marked elevation of glucose metabolism in the corresponding side of the striatum was observed with <sup>18</sup>F-fluorodeoxyglucose positron emission tomography (<sup>18</sup>F-FDG PET).

## Case presentation

### Case 1

A 12-year-old girl experienced transient numbness and weakness on the left side of the body during running and was admitted to our hospital. Angiography revealed severe stenosis in the terminal portion of the bilateral internal carotid artery with extensive development of abnormal collateral vessel networks (Fig. 1) symptomatic of moyamoya disease. She underwent superficial temporal artery-to-middle cerebral artery anastomosis of the right hemisphere;



**Fig. 1** Case 1 (left hemichorea). Anterior-posterior view of right (a) and left (b) internal carotid artery angiography before surgery revealing occlusion of the terminal portion of the internal carotid artery with development of moyamoya vessels. Lateral view of right (c) and left (d) external carotid artery angiography after surgery revealing good patency of bypasses

this was repeated on the left side 1 month later. The surgery eliminated the transient ischemic attacks. Angiography 3 months after surgery revealed good patency of the bypasses (Fig. 1). She remained symptom-free for 5 years after surgery.

At the age of 17 years, she suddenly experienced continuous involuntary movement of the left arm. Her manifestation was characterized as brief, involuntary, and irregular movements of her left forearm, shoulder, fingers, and face that

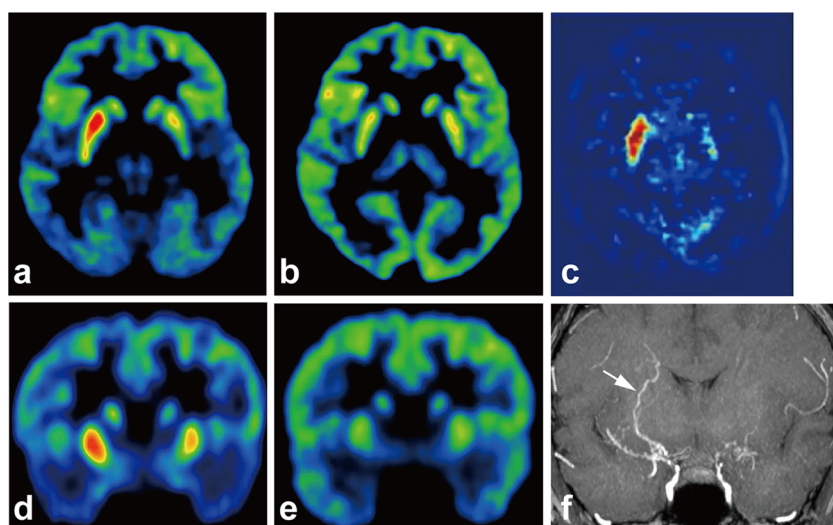
continued on their own without conscious effort (video), leading to a diagnosis of hemichorea. Laboratory examinations were normal, including the level of free thyroxine, the thyroid-stimulating hormone. Single-photon emission computed tomography (SPECT) revealed normal cerebral blood flow. Magnetic resonance imaging (MRI) revealed a small asymptomatic ischemic lesion in the right frontal white matter.  $^{18}\text{F}$ -FDG PET revealed markedly elevated glucose metabolism in the right striatum (Fig. 2). Coronal section of MR angiography demonstrated a dilated and extended lenticulostriate artery passing through the right striatum and connecting to the medullary artery in the periventricular area (Fig. 2). She was treated with oral intake of haloperidol, with the chorea gradually improving thereafter. Elimination of the chorea was confirmed when she visited our clinic 2 months after onset.  $^{18}\text{F}$ -FDG PET performed at that time revealed normalized glucose metabolism in the right striatum (Fig. 2).

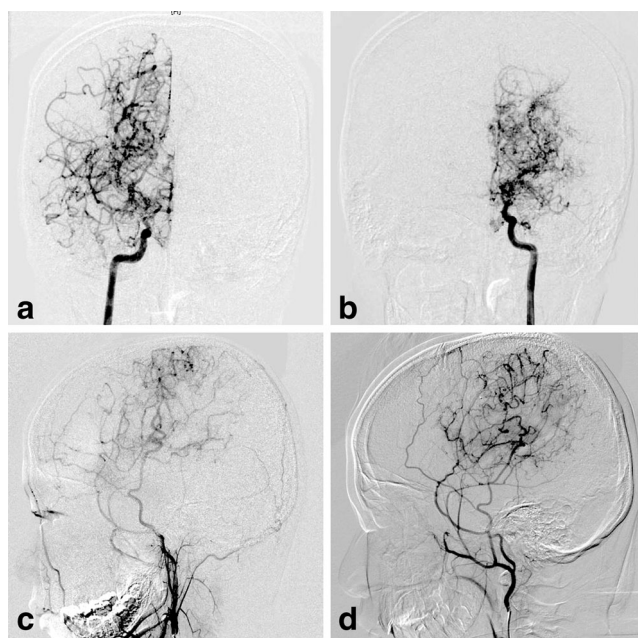
## Case 2

A 10-year-old girl experienced transient motor weakness of the left leg while playing and was admitted to our hospital. Her father was diagnosed as neurofibromatosis type 1. She had many café-au-lait spots on her skin, a finding symptomatic of neurofibromatosis. Angiography revealed severe stenosis in the terminal portion of the bilateral internal carotid artery with extensive development of abnormal collateral vessel networks (Fig. 3) symptomatic of moyamoya syndrome. She underwent superficial temporal artery-to-middle cerebral artery anastomosis of the left hemisphere; this was repeated on the right side 1 month later. Surgery eliminated the transient ischemic attacks.

Three months after surgery, she experienced involuntary movement of her right extremities. The involuntary

**Fig. 2** Case 1 (left hemichorea). Axial image of  $^{18}\text{F}$ -FDG PET during the acute phase (a) and during recovery from chorea (b). Subtraction image generated by registered acute and recovery phase  $^{18}\text{F}$ -FDG PET images (c) revealing marked hypermetabolism in the right striatum in the acute phase. Coronal image of  $^{18}\text{F}$ -FDG PET during the acute phase of (d) and during recovery from chorea (e). Coronal section of MR angiography (f) revealing an extensively dilated and extended lenticulostriate artery in the right striatum (arrow)





**Fig. 3** Case 2 (right hemichorea). Anterior-posterior view of right (a) and left (b) internal carotid artery angiography before surgery revealing occlusion of the terminal portion of the internal carotid artery with extensive development of moyamoya vessels. Lateral view of right (c) and left (d) external carotid artery angiography after surgery revealing good patency of bypasses

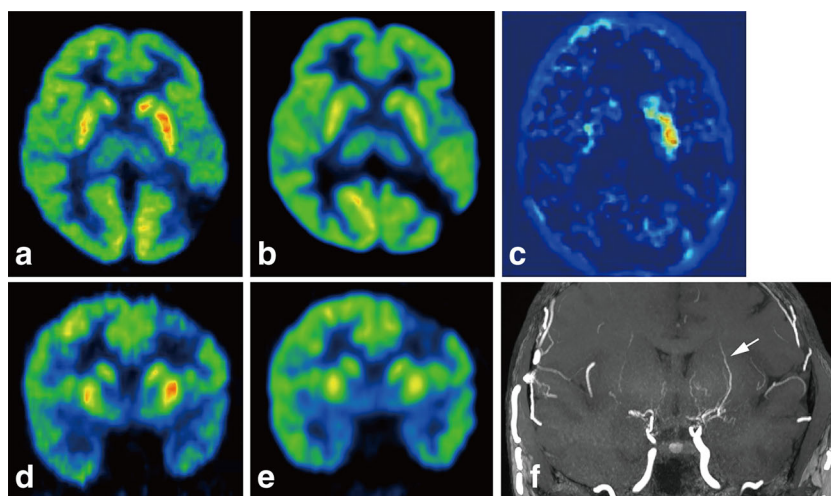
movements, especially prominent in her fingers, were diagnosed as hemichorea. While angiography revealed good patency of bypasses (Fig. 3), SPECT revealed hypoperfusion in the left basal ganglia and watershed area attributable to the infarcts occurring before surgery. She underwent  $^{18}\text{F}$ -FDG PET, revealing elevation of glucose metabolism in the left striatum (Fig. 4). Coronal section of MR angiography revealed a dilated and extended lenticulostriate artery in the left

striatum (Fig. 4). The chorea gradually improved after administration of haloperidol.  $^{18}\text{F}$ -FDG PET performed after recovery revealed normalized glucose metabolism in the right striatum (Fig. 4).

### Discussion

In the present cases, the striatal metabolism was elevated and subsequently normalized, corresponding to the activity of chorea and suggesting a probable association between the two phenomena. Striatal hypermetabolism has been reported in various types of chorea, including chorea caused by group A streptococcal infection (Sydenham’s chorea) [5, 10, 20], hyperthyroidism [3, 9], and hyperglycemia [12, 19, 24]. As of this writing, however, no reports have surfaced in relation to moyamoya disease. Although the prospect of a causal relationship between chorea and striatal hypermetabolism remains a subject of discussion [8, 10], striatal hypermetabolism could represent activation of a direct pathway—an inhibitory signal pathway to the internal segment of the globus pallidus and substantia nigra, both of which negatively regulates thalamocortical projection [4]—resulting in activation of the motor circuit. Several possible mechanisms have been suggested to explain striatal hypermetabolism observed in chorea: immunological reaction in Sydenham’s chorea and hyperthyroidism [3, 8, 9, 13, 20], and the failure of vascular autoregulation in hyperglycemia [12, 19, 24].

In moyamoya disease, ischemia or decreased cerebral blood flow is a commonly hypothesized cause of chorea [2,



**Fig. 4** Case 2 (right hemichorea). Axial image of  $^{18}\text{F}$ -FDG PET during the acute phase (a) and during recovery from chorea (b). Subtraction image generated by registered acute and recovery phase  $^{18}\text{F}$ -FDG PET images (c) revealing marked hypermetabolism in the left stratum in the

acute phase. Coronal image of  $^{18}\text{F}$ -FDG PET during the acute phase (a) and during recovery from chorea (b). Coronal section of MR angiography (f) revealing an extensively dilated and extended lenticulostriate artery in the left striatum (arrow)

11, 15–18, 21, 23, 25]. The basal ganglia [2, 11, 15, 17, 23] and the frontal cortical/subcortical region [14, 21] are candidates for the origin of chorea in moyamoya disease. The location where MRI revealed ischemic lesions in our cases is compatible with those reported previously, and such ischemic lesions occurring before surgery could cause delayed-onset chorea. On the other hand, it might be a unique finding of our cases that chorea occurred in the stable phase despite good augmentation of blood flow from successful bypass. Furthermore, the striatal hypermetabolism observed in our cases is unlikely to be related to cerebral blood flow, as it occurred regardless of the presence of the perfusion defect in SPECT. A different pathological mechanism of chorea might be involved in our cases.

Both of our cases had a specific vascular feature—a dilated and extended lenticulostriate artery—at the exact site of the hypermetabolic lesion. We have defined this type of collateral as periventricular anastomosis, which is specific to moyamoya disease [6, 7]. Ahn et al. reported that all their cases of moyamoya disease-induced chorea had similar angiographic features on the side corresponding to the chorea [1]. They speculated that these vessels might contribute to the development of chorea through their physical disruption of normal striatal signaling. In our cases, these vessels might also involve activation of the striatum through some mechanism, such as impairment of vascular autoregulation, resulting in a hypermetabolic state in the striatum. Further studies of both striatal metabolism and vascular morphology might elucidate the pathophysiological mechanism of chorea in moyamoya disease.

#### Compliance with ethical standards

**Conflict of interest** The authors declare that they have no conflict of interest.

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