

Dramatic Disappearance of Moyamoya Disease-Induced Chorea After Indirect Bypass Surgery

—Case Report—

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Abstract

A 31-year-old woman with moyamoya disease presented with choreiform movements persisting for 4.5 years. Magnetic resonance imaging showed a fine vascular plexus in the base of the brain but no parenchymal brain damage. Cerebral angiography revealed intracranial vascular abnormalities compatible with moyamoya disease. Single photon emission computed tomography with N-isopropyl-p-¹²³I-iodoamphetamine showed definite reduction of the regional cerebral blood flow (rCBF) in the bilateral striata and frontotemporoparietal cortex. rCBF study with acetazolamide administration indicated marked decrease of rCBF reserve in those regions. She underwent indirect bypass surgery (encephalo-duro-arterio-myo-synangiosis) under a diagnosis of moyamoya disease. The choreic involuntary movements disappeared shortly after surgery. Postoperative angiography showed neovascularization in the extracranial to intracranial direction, associated with dramatic increase in rCBF in the involved regions.

Key words: moyamoya disease, chorea, regional cerebral blood flow, indirect bypass surgery

Introduction

Moyamoya disease is defined as an idiopathic cerebral vasculopathy characterized by steno-occlusive changes in the terminal portion of the bilateral internal carotid arteries and the formation of aberrant arterial networks in the vicinity of this lesion.

Moyamoya disease occurs predominantly in children and young adults of Asian ethnicity, who usually present with headache, ischemic or hemorrhagic stroke, and seizures.³⁾ Involuntary movement is relatively rare as a clinical symptom of moyamoya disease. Analysis of 1500 moyamoya disease cases in Japan showed that 2.6% of the initial symptoms, and

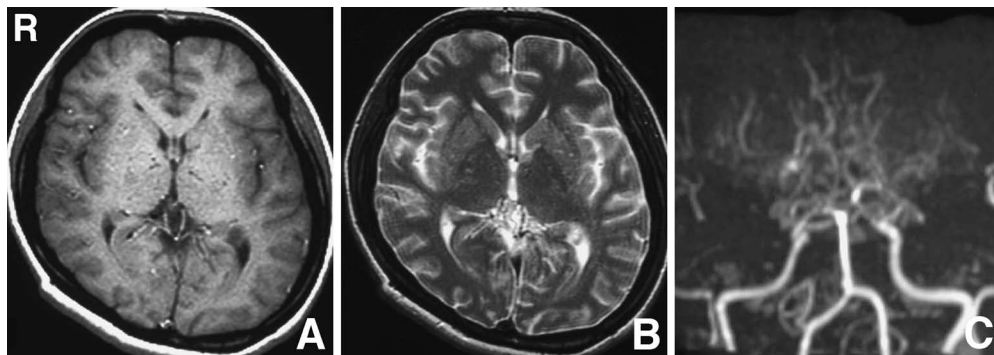


Fig. 1 Preoperative axial T₁-weighted (A) and T₂-weighted (B) magnetic resonance (MR) images, and MR angiogram (C), showing the development of fine vascular plexus in the base of the brain, but no infarction or hemorrhage associated with moyamoya disease.

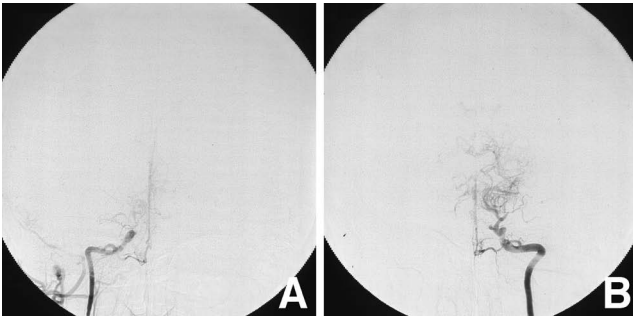


Fig. 2 Preoperative right (A) and left (B) carotid angiograms showing bilateral carotid arterial occlusions and abnormal basal vascular network typical of moyamoya disease.

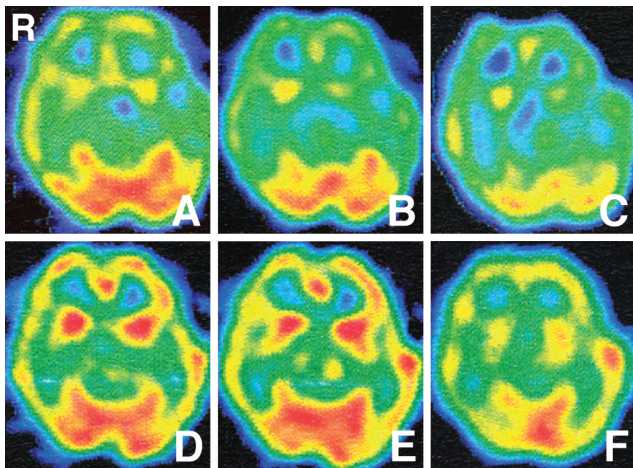


Fig. 3 Preoperative N-isopropyl-p-¹²³I-iodoamphetamine single photon emission computed tomography (¹²³I-IMP SPECT) scans prior to (A) and after (B) acetazolamide administration, and subtraction image (C), demonstrating reduction in the regional cerebral blood flow and decreased vascular reserve in bilateral striata and the frontotemporoparietal cortex. Postoperative ¹²³I-IMP SPECT scans prior to (D) and after (E) acetazolamide administration, and subtraction image (F), demonstrating remarkable improvement of cerebral perfusion in the left hemisphere and bilateral striata.

3.3% of the neurological symptoms on admission comprised involuntary movements.³⁾ Only 23 cases have been reported with choreiform movements, occurring in 6 adults and 17 children.^{1,2,4-6,8-12,14-19)} Two adults and 14 children underwent either direct or indirect bypass surgery, all resulting in favorable outcomes.

We describe an adult patient with moyamoya dis-

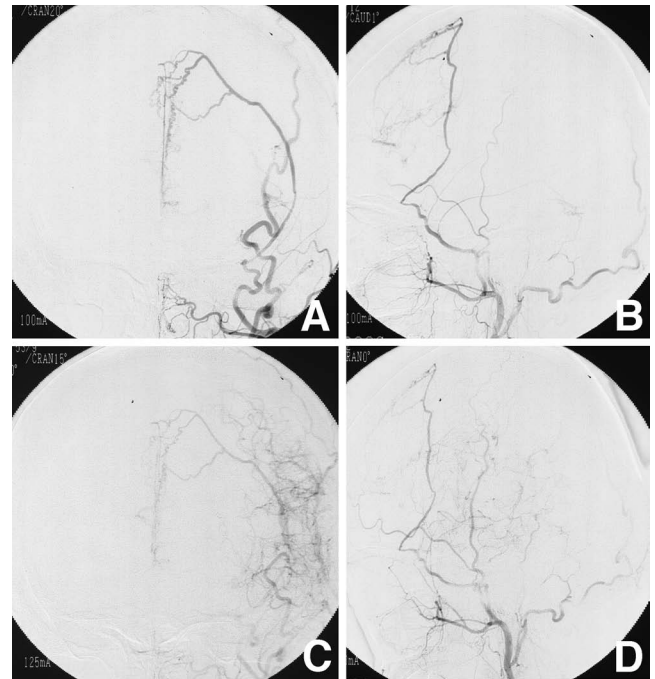


Fig. 4 Preoperative left external carotid angiograms, anteroposterior (A) and lateral (B) views, and left external carotid angiograms, anteroposterior (C) and lateral (D) views, 2 months after surgery demonstrating the formation of neovascularization from the extracranial anastomosis.

ease initially presenting with persistent choreic movement, who was successfully treated with cerebral revascularization.

Case Report

A 31-year-old right-handed woman presented with choreic involuntary movements beginning in her right thumb and middle finger in 1998, which then expanded to the right side of her face several months later. Administration of haloperidol 0.75 mg/day by a local physician resulted in temporary subsidence of the choreic involuntary movements. However, 4 years later, choreic movements recurred in her right upper and lower extremities, expanding to the left upper extremity after several months. The choreic movements were resistant to medication, and she was referred to us.

Neurological examination found she was intact except for the choreic involuntary movements and slight hypotonicity in her extremities. The involuntary movements were bilateral, but more frequent on the right. Laboratory examination found no abnormalities in blood count, urinalysis, liver and renal

function, electrolytes, blood glucose, and thyroid function. Prothrombin time and activated thromboplastin time were normal. Antinuclear antibody and rheumatoid factor levels were all within normal limits. Magnetic resonance (MR) imaging showed development of fine vascular plexus in the base of the brain, but no infarct or hemorrhage (Fig. 1). Cerebral angiography showed occlusion of the bilateral internal carotid arteries just after the branching of the ophthalmic artery on the right, and the posterior communicating artery on the left. The bilateral anterior and middle cerebral arteries were not well visualized and numerous fine collateral vessels were seen (Fig. 2). Single photon emission computed tomography scans with N-isopropyl-p-¹²³I-iodoamphetamine (¹²³I-IMP SPECT) before and after administration of acetazolamide showed reduced regional cerebral blood flow (rCBF) in the bilateral striata and frontotemporoparietal cortex, more prominent on the left (Fig. 3A, B). These findings implied a lack of vascular reserve in these regions, suggesting misery perfusion (Fig. 3C).

We initially planned superficial temporal artery-

middle cerebral artery (STA-MCA) anastomosis plus encephalo-duro-arterio-myo-synangiosis (EDAMS). However, the recipient MCA was too thin (less than 0.4 mm in diameter) for direct revascularization. Therefore, we performed EDAMS on the left. Postoperative angiography taken 2 months after surgery showed abundant neovascularization from the extracranial arteries, compared with the preoperative finding (Fig. 4). Postoperative ¹²³I-IMP SPECT demonstrated remarkable improvement of cerebral perfusion in the left hemisphere and bilateral striata (Fig. 3D-F). The choreic involuntary movements improved satisfactorily without medication within 2 months after surgery. She has remained symptom-free for 4 years without additional contralateral surgery.

Discussion

Choreiform movement has been reported in association with striatal lesions caused by either cerebral stroke or degenerative diseases.^{7,13} Therefore, moyamoya disease-induced chorea is likely to be

Table 1 Cases of moyamoya disease-induced chorea examined by cerebral blood flow (CBF) studies

Author (Year)	Age (yrs), Sex	Abnormal signal on MR imaging	CBF studies (SPECT/PET)	Effective therapy (Postoperative rCBF)
Tsuchiyama et al. (1992) ¹⁷⁾	33, F	caudate nucleus	hypoperfusion in the caudate nucleus and the parietal lobe (SPECT)	medical therapy (*)
Hosoya (1997) ⁵⁾	9, F	frontal, parietal, and occipital areas	hypoperfusion in these regions as well as in the basal ganglia (SPECT)	STA-MCA anastomosis (increased rCBF in the whole hemisphere including the basal ganglia)
Unno et al. (2000) ¹⁸⁾	16, F	deep in the frontal white matter	hypoperfusion and decreased vascular reserve in the frontal and parietal lobes and the basal ganglia (SPECT)	chorea during pregnancy, completely subsided after abortion (*)
Han et al. (2000) ²⁾	29, F	frontal white matter	decreased vascular reserve in the frontal and temporal lobes and the basal ganglia (SPECT)	EDAS (*)
Lyoo et al. (2000) ⁹⁾	22, F	frontotemporoparietal white matter	hypoperfusion in the frontotemporoparietal lobe (SPECT)	spontaneous resolution (*)
Miura et al. (2002) ¹⁰⁾	54, M	no infarcts	misery perfusion in the temporoparietal lobe and the striatum (PET)	haloperidol (*)
Hong et al. (2002) ⁴⁾	20, F	centrum semiovale	perfusion defect in the basal ganglia and decreased vascular reserve in the frontal lobe (SPECT)	STA-MCA anastomosis (normal perfusion in the basal ganglia, no improvement in the left frontal lobe)
Im et al. (2004) ⁶⁾	7, F	frontal white matter	perfusion defect in the hemisphere (SPECT)	EDAS (*)
	13, F	frontoparietal white matter	perfusion defect and decreased vascular reserve in the posterior border zone (SPECT)	EDAS and EGS (*)
Kim et al. (2006) ⁸⁾	8, F	no infarcts	decreased vascular reserve in the frontoparietal lobe (SPECT)	EDAMS (*)
Present case	31, F	no infarcts	hypoperfusion and decreased vascular reserve in the frontotemporoparietal lobe and the striatum (SPECT)	EDAMS (markedly increased rCBF in these regions)

EDAMS: encephalo-duro-arterio-myo-synangiosis, EDAS: encephalo-duro-arterio-synangiosis, EGS: encephalo-galeo-synangiosis, MR: magnetic resonance, PET: positron emission tomography, rCBF: regional CBF, SPECT: single photon emission computed tomography, STA-MCA: superficial temporal artery-middle cerebral artery, (*): not described.

caused by ischemia of the striatopallidum, or compression by the abnormal vascular network.^{4,6,8-10,14} CBF studies have been performed in 11 moyamoya disease patients with chorea including the present case (Table 1).^{2,4-6,8-10,17,18} Even if MR imaging showed no damage of the basal ganglia, basal blood flow reduction or decreased vascular reserve in basal ganglia was noted in almost all cases. Surgical vascular reconstruction resulted in disappearance of involuntary movement such as chorea in all 17 patients. Postoperative CBF study (SPECT) demonstrated marked improvement of rCBF in the basal ganglia in all 3 patients investigated.^{4,5} One moyamoya disease patient with involuntary movements showed postoperative improvement of perfusion in the basal ganglia on SPECT, whereas the rCBF response to acetazolamide in the frontal lobe was not improved.⁴ These results suggested that the hypoperfusion in the basal ganglia, rather than the cerebral cortex, was responsible for the generation of hemichorea.

The present case showed dramatic improvement of bilateral choreic movements after EDAMS on the left, which corresponded with remarkable improvement of perfusion in bilateral striata on postoperative ¹²³I-IMP SPECT. These findings strongly suggest that hemodynamic compromise in the striatum is responsible for the manifestation of choreic movement in moyamoya disease patients.

Patients with choreic involuntary movements caused by moyamoya disease with poor vascular reserve in the relevant region may be candidates for surgical intervention.

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