

## Case Reports

# Moyamoya Disease Presenting with Chorea

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Three children with moyamoya disease are reported whose initial and predominant manifestations were choreic movements. Two of the patients presented with unsteady gait and the other with clumsiness. Choreic movements were recurrent and were often triggered by excitement, emotional tension, or crying. They occurred unilaterally or bilaterally and often alternated between the right and left. Moyamoya disease must be considered in the differential diagnosis of acquired chorea in children.

Watanabe K, Negoro T, Maehara M, Takahashi I, Nomura K, Miura K. Moyamoya disease presenting with chorea. *Pediatr Neurol* 1990;6:40-2.

## Introduction

Moyamoya disease is characterized by the angiographic appearance of an abnormal vascular network at the base of the brain and bilateral stenosis of the intracranial portions of the internal carotid arteries and the trunks of the anterior and middle cerebral arteries [1]. The disease usually presents in children as recurrent episodes of transient cerebral ischemia which manifests as acute motor and sensory deficit, speech disturbance, headache, and seizures. Involuntary movements occurred in about 3% of a series of patients [2]; however, in another study, about 6% of ischemic patients – mostly children – exhibited these movements [3]. We report 3 patients with moyamoya disease whose initial and predominant manifestations were choreic movements; 2 patients had been treated for Sydenham chorea.

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## Case Reports

*Patient 1.* This 4-year-old girl became unsteady at 3 years, 8 months of age when she ran at an athletic competition. Two months later, she again became increasingly unsteady and was admitted to a local hospital with a diagnosis of acute cerebellar ataxia. In several days, unsteadiness improved and she was discharged. Computed tomography (CT) was unremarkable. She later became clumsy and often would spill foods. At 4 years, 3 months of age, she again became unsteady after falling from a slide and striking her head; she cried intensely. She had left leg weakness which improved after a short rest. Her speech was slow and unclear. She also became restless and emotionally unstable, prompting admission to our hospital.

On admission, she was alert, intelligent, and well oriented. She had difficulty walking. Cranial nerves were normal. Speech was slow and dysarthric. Finger-to-nose test revealed no dysmetria, although there were involuntary movements during the maneuver. Sensation was normal. She had mild left arm weakness and mild, generalized hypotonia. Deep tendon reflexes were normal or slightly decreased; there were no pathologic reflexes. There were constant, irregular, choreic movements which involved the face, neck, shoulder, and all limbs and were aggravated by emotional tension or excitement.

Laboratory examinations were not remarkable, including ASO and ASK. CT disclosed multiple low-density areas in the heads of both caudate nuclei and bilateral frontal white matter and punctate high-density areas in bilateral putamen. Magnetic resonance imaging (MRI) revealed high-intensity lesions around the anterior and posterior horns and in the heads of (more marked on the left) of the caudate nuclei on T<sub>2</sub>-weighted sequences (Fig 1) and punctate low-intensity lesions in both basal ganglia on T<sub>1</sub>-weighted images. Three-vessel cerebral angiography revealed marked stenosis of the C1 portion of the carotid

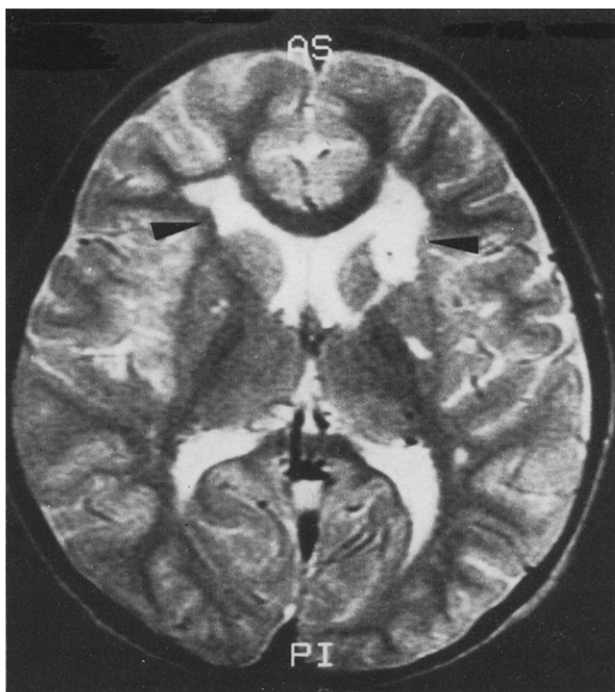


Figure 1. MRI of Patient 1. T<sub>2</sub>-weighted image (TR: 2,000, TE: 80) reveals high-intensity lesions mainly around the anterior horns and in the head of the left caudate nucleus (arrowheads).

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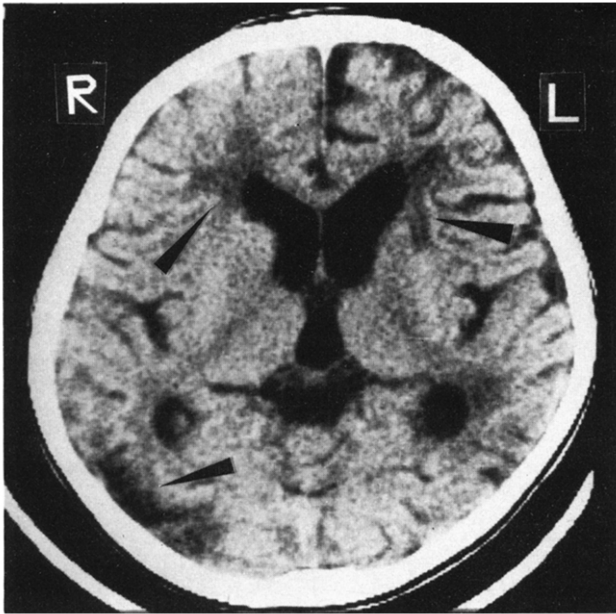


Figure 2. CT of Patient 2 demonstrating cerebral atrophy, especially in the left hemisphere with multiple low-density lesions around both frontal horns, the head of the left caudate nucleus, and in the right parieto-occipital region (arrowheads).

arteries with an abnormal network of vessels in the basal ganglia which is consistent with moyamoya disease.

She underwent encephalo-duro-arterio-synangiosis on both sides and became free of neurologic signs immediately after surgery. There have been no recurrences for 14 months.

**Patient 2.** This 7-year-old boy was admitted to our hospital with chorea and mental deterioration. At 3 years, 10 months of age, he had the first episode of neurologic disorder which lasted about 1 week and was characterized by gait difficulty, dysarthria, and choreic movements involving mainly the left upper and lower limbs; he was admitted to a local hospital. Enhanced and unenhanced CT scans, right transbrachial cerebral angiography, and electroencephalography all were normal. He had several recurrences, lasting from several days to weeks, and was treated for chorea minor. In spite of treatment with various drugs for chorea minor, he had continuous but fluctuating degrees of predominantly right-sided choreic movements and mental deterioration; IQ dropped from 107 at age 5 years to 77 1 year later. He then was referred to our institution.

On admission, he was alert and well oriented. He had an unsteady gait but no signs of cerebellar dysfunction. Cranial nerves were normal. Speech was somewhat unclear. Sensation was normal. Strength and deep tendon reflexes were normal; there were no pathologic reflexes. Muscle tone was slightly decreased. There were almost constant distal choreic movements of all limbs, most marked in the right hand.

General laboratory examinations were not remarkable. Cranial CT demonstrated generalized brain atrophy more prominent in the left hemisphere and multifocal low-density areas, including the left caudate nucleus, deep frontal regions, and the left parieto-occipital regions (Fig 2). MRI demonstrated high-signal intensity on the T<sub>2</sub>-weighted sequence in the same regions as on CT. Three-vessel cerebral angiography displayed findings typical of moyamoya disease, consisting of marked stenosis of C1 portions of the carotid arteries and an abnormal network of vessels in the basal regions.

He underwent a left superior temporal artery-middle cerebral artery (STA-MCA) anastomosis; 5 months later, an operation on the right one was performed. Choreic movements have almost disappeared completely but intelligence has not improved. There has been no relapse of neurologic dysfunction for 26 months.

**Patient 3.** This 14-year-old girl was referred to us with longstanding choreic movements unresponsive to various treatments for Sydenham chorea. At the age of 8 years, she could not easily put on her socks because of involuntary movements. She was admitted to a local hospital and was diagnosed as having Sydenham chorea which seemingly improved with treatment. CT was reportedly normal. At age 11 years, involuntary movements exacerbated and she had difficulty running. Choreic movements persisted and school grades worsened.

On admission, she was alert and well oriented. Cranial nerves were normal. Speech became dysarthric when she spoke rapidly. Gait was slightly unsteady. She was clumsy but had no signs of cerebellar dysfunction. Muscle power, tone, deep tendon reflexes, and sensation were normal. There were distal choreic movements of the arms and legs.

General laboratory examinations were unremarkable. CT revealed multiple low-density areas in deep frontal and parieto-occipital regions. MRI demonstrated many typical fluid voids in both basal ganglia on a T<sub>1</sub>-weighted image (Fig 3) in addition to high-signal intensity lesions on a T<sub>2</sub>-weighted image corresponding to lesions on CT. Three-vessel cerebral angiography confirmed the diagnosis.

She underwent a right STA-MCA anastomosis and 5 months later a similar procedure was performed on the left which resulted in the disappearance of choreic movements. She has been free of neurologic dysfunction for 10 months.

## Discussion

Among the various causes of chorea in childhood, Sydenham chorea had been the most common of the acquired choreas, but now has become exceedingly rare because rheumatic fever has virtually disappeared; therefore, moyamoya disease must always be considered in the differential diagnosis of acquired chorea in children. The incidence of involuntary movements associated with childhood moyamoya disease is higher than in the adult type. Choreic movements in this disease usually wax and



Figure 3. MRI of Patient 3. T<sub>1</sub>-weighted image (TR: 500, TE: 20) demonstrates numerous fluid voids corresponding to moyamoya vessels in the basal ganglia (arrowheads).

wane and are often triggered by excitement, emotional strain, or crying. They may be unilateral or bilateral and often appear alternately in the left and right sides.

In adults, chorea has been reported to be caused by a striatal lesion, mainly infarction [4,5]. It is believed to result from release of the globus pallidus from inhibition by the caudate nucleus, putamen, or their efferent pathways in the presence of functioning thalamus, motor cortex, and pyramidal tracts [6]. These striatal lesions may be caused by transient ischemia. In our patients, choreic movements were intermittent, reversible, and improved after surgery. Moreover, in 2 of 3 patients under study, CT was reported to be initially normal; Patient 3 did not have striatal infarction. Therefore, reversible, transient ischemia may have caused the striatal hypofunction which resulted in disinhibition of the globus pallidus and produced chorea.

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