

Hemidystonia and Hemichoreoathetosis as an Initial Manifestation of Moyamoya Disease

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Objective: To describe hemidystonia and hemichoreoathetosis in an adult patient with moyamoya disease without a previous history of cerebrovascular accident.

Design: Case report.

Setting: Tertiary care center.

Patient: A 22-year-old woman suddenly developed dystonic spasms in her left hand and left foot after a severe emotional stress. The dyskinesia gradually subsided over the next 4 months. Five months after the onset, she suddenly developed choreoathetoid movement in her right hand and right foot.

Main Outcome and Results: The patient had both somatic and cortical sensory deficits in the right hand and right foot. Magnetic resonance imaging of the brain

showed an infarction at the right putamen and lesions involving the right frontal lobe and the left frontotemporo-parietal lobe. Magnetic resonance cerebral angiography showed severe stenoses of both internal carotid arteries at the supraclinoid portion and numerous collateral vessels, compatible with moyamoya disease. Single photon emission tomography of the brain showed hypoperfused areas at the right frontal and left frontotemporo-parietal lobes. The choreoathetosis of the right limbs improved markedly, along with improvement of sensory deficits.

Conclusions: To our knowledge, this is the first report of an adult patient presenting with hemidystonia and hemichoreoathetosis as the initial manifestations of moyamoya disease.

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AS AN INITIAL manifestation, children with moyamoya disease frequently develop seizures or focal motor signs as a result of cerebral ischemia. On the other hand, in adult patients, frequent initial symptoms are headache, sudden loss of consciousness, or focal motor signs. Three percent of 1500 Japanese patients with moyamoya disease developed dyskinesia as an initial manifestation.¹ However, their detailed clinical histories and characteristics of the involuntary movements were not described.

In the English-language literature, 3 cases involving children who initially developed chorea in association with moyamoya disease have been reported.^{2,3} We describe a 22-year-old woman with moyamoya disease. She acutely developed dystonia involving one side of the body and

choreoathetosis involving the other side of the body at an interval of 5 months.

REPORT OF A CASE

A 22-year-old right-handed female office worker suddenly developed a headache and a tingling sensation in her left fingers and toes 3 days after her job was threatened by her employer. Several days later, she noticed the sudden onset of involuntary spasms in her left hand and left foot. Over a period of 4 months, the dyskinesia gradually resolved. Five months after the onset, she felt sudden pain and tingling followed by involuntary spasm, and dyskinesia occurred in her right hand and right foot. The involuntary movement gradually worsened and interfered with her daily activities. When she was evaluated 4 months after the onset of the second episode, she had mild impairment in calcu-

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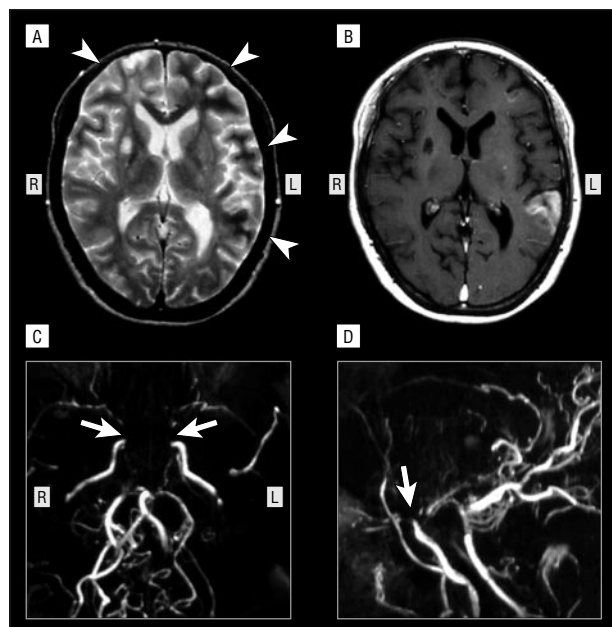


Figure 1. A, A T2-weighted axial magnetic resonance image (MRI) of the brain shows a high signal intensity lesion at the right putamen. There are multiple low signal intensity lesions at the right frontal and left frontotemporoparietal subcortical white matter (arrowheads). B, A gadolinium-enhanced T1-weighted axial MRI of the brain shows abnormal enhancement in the left parietal lobe and a low signal intensity lesion in the right putamen. C and D, A magnetic resonance cerebral angiogram shows severe stenosis of both internal carotid arteries at the supraclinoid portion (arrows) and numerous collateral vessels originating mainly from the posterior circulation.

lation and agraphesthesia in her right hand. Her strength was normal. Sitting on a chair, she had intermittent slow choreoathetoid movements in her right fingers and right foot. When both her arms were extended, dystonic posturing (hyperextension) became evident in her left fourth and fifth fingers, and choreoathetoid movements of her left arm were aggravated. She had decreased pain sensation in her right cheek and right forearm distal to the elbow, and position sensation was impaired in both great toes and the right thumb. Vibration sensation was impaired distally in both hands and feet. A 2-point discrimination test showed impairment in the right upper and lower limbs. The patient's tandem gait was normal, but the findings of other cerebellar function tests were difficult to interpret owing to her involuntary movements. Her deep tendon reflexes were normal except for a hyperactive right biceps jerk. The right plantar reflex was extensor. The results of routine laboratory and blood tests looking for organic causes of the chorea were all normal (eg, thyroid function tests, serum rheumatoid factor, LE cell test, antinuclear antibody, anti-DNA, anti-Ro, anti-La, anticardiolipin antibody, and lupus anticoagulant tests).

T2-weighted (repetition time, 2200 milliseconds; echo time, 80 milliseconds; and acquisition, 1 time) magnetic resonance images (MRIs) of the brain showed a high signal intensity lesion at the right putamen and low signal intensity lesions in the right frontal and the left frontotemporoparietal subcortical white matter (**Figure 1**, A). Gadolinium-enhanced T1-weighted (repetition time, 600 milliseconds; echo time, 14 milliseconds; and ac-

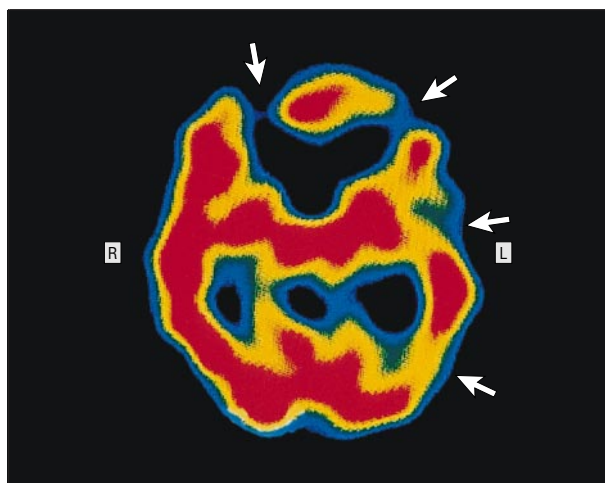


Figure 2. A single photon emission computed tomogram of the brain shows multiple hypoperfused cortical areas (arrows) corresponding to the lesions seen on the magnetic resonance images of the brain.

quisition, 2 times) MRIs of the brain showed an enhancing lesion in the left parietal lobe and a low signal intensity lesion at the right putamen (**Figure 1**, B), but no lesion in the left basal ganglia. Magnetic resonance angiography revealed severe stenosis of both supraclinoid carotid arteries. There were numerous collateral vessels originating mainly from the posterior circulation (**Figure 1**, C and D). Brain single photon emission tomography showed hypoperfused areas at the right frontal and left frontotemporoparietal lobes, but there was no evidence of perfusion defects of the basal ganglia or thalami (**Figure 2**). Six days after the patient's admission, choreic movements of her right arm disappeared completely, and she showed mild abnormal posture in her right hand while stretching out the hand in front of her. At that time, her somatic and cortical sensory deficits had improved markedly. She refused the neurosurgical management and was unavailable for follow-up.

COMMENT

In the English-language literature, 5 patients who developed bilateral chorea attributed to moyamoya disease have been described.²⁻⁴ Two of the 5 patients had prior acute neurological deficits, and the other 3 developed chorea as an initial manifestation of moyamoya disease before the age of 10 years. To our knowledge, our patient is the first adult with moyamoya disease who presented with hemidystonia and hemichoreoathetosis as the initial manifestations.

Four of the 5 patients described with choreoathetosis had lesions of the thalamus or basal ganglia, and the other one had multiple lesions involving the frontoparieto-occipital white matter. Interestingly, the brain MRIs of our patient showed frontal subcortical and putamenal lesions contralateral to the limbs that were affected by dystonia and multiple white matter lesions contralateral to the limbs that were affected by choreoathetosis. Single photon emission computed tomography of the brain showed hypoperfused areas corresponding to the lesions seen on the brain MRI scans.

The dystonia of the left hand was most likely caused by damage to the right putamen. In contrast, there was no objective evidence suggesting damage to the left basal ganglia or thalamus contralateral to the limbs that were affected by choreoathetosis. The choreic movements of the right arm and right foot improved, as did the somatic and cortical sensory deficits. This clinical course might suggest that our patient had pseudo-choreoathetosis.

Our patient's organic dyskinesia demonstrated unusual clinical features (eg, sudden onset, affecting one side first and the other side later, and additional cortical signs). She was under extreme emotional stress just before the onset of dyskinesia, and our first diagnosis was functional dyskinesia. Although the emotional stress could be coincidental, stress might play some role in precipitating symptoms due to moyamoya disease.

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