

Moyamoya Disease Presenting with Hemichoreoathetosis and Hemidystonia

Moyamoya disease (MMD) is an idiopathic occlusive intracranial vasculopathy affecting mainly children and young adults, and characterized by the involvement of the circle of Willis and development of an extensive collateral circulation.¹ It tends to manifest as transient ischemic attacks or seizures in children as a result of cerebral ischemia; however, hemorrhagic strokes are more common in adult patients. Movement disorders as an initial manifestation of this condition are rare, with an estimated frequency of 3 to 6%.^{2,3} Here, we report on a 54-year-old patient with MMD who developed hemichoreoathetosis and hemidystonia as an initial manifestation.

A 54-year-old woman presented with choreoathetoid movement in her left hand for 6 months. She had also noted numbness of her left forearm for one year. The patient's general physical examination was unremarkable. Neurological examination revealed hemidystonia and hemichoreoathetosis in her left arm and leg. There was no limb weakness or somatic or cortical sensory loss. The patient's tandem gait was normal and there were no other cerebellar signs. Her deep tendon reflexes were normal and the plantar reflexes were flexor.

Magnetic resonance imaging (MRI) of the brain showed multiple small round low intensity areas extending from the suprasellar cisterns to the bilateral basal ganglia (Fig. 1A). Magnetic resonance cerebral angiography demonstrated severe stenosis of both internal carotid arteries at supraclinoid portion with numerous collateral vessels (Fig. 1B), compatible with MMD. A ^{99m}Tc-HMPAO brain Single Photon Emission Computed Tomography (SPECT) showed decreased perfusion in the right occipital and high frontal regions (Fig. 1C). A ^{99m}Tc-TRODAT-1 brain SPECT showed decreased uptake in the bilateral striatum (Fig. 1D). The involuntary movements improved gradually under treatment with anticholinergics.

MMD has a bimodal age distribution with peaks in the first and fourth decades of life. The clinical presentations of pediatric MMD are mainly ischemic in nature, including transient ischemic attacks, which are often precipitated by hyperventilation, such as blowing or crying, migraine-like headaches, epileptic seizures, mental deterioration, and cerebral infarction. However, the initial manifestations of MMD in adults are more often headache, sudden loss of consciousness, or focal motor signs due to intracerebral hemorrhage. Involuntary movement disorders in an adult with MMD as our case are very rare. Only a few cases of MMD presenting with movement disorders have been reported and these have been found to occur in 3 to 6% of patients, mostly in children. Involuntary movements include chorea, hemidystonia, hemichoreoathetosis, and paroxysmal dyskinesia.²⁻⁷ These movement disorders were attributed to ischemic changes in the basal ganglia-thalamocortical cir-

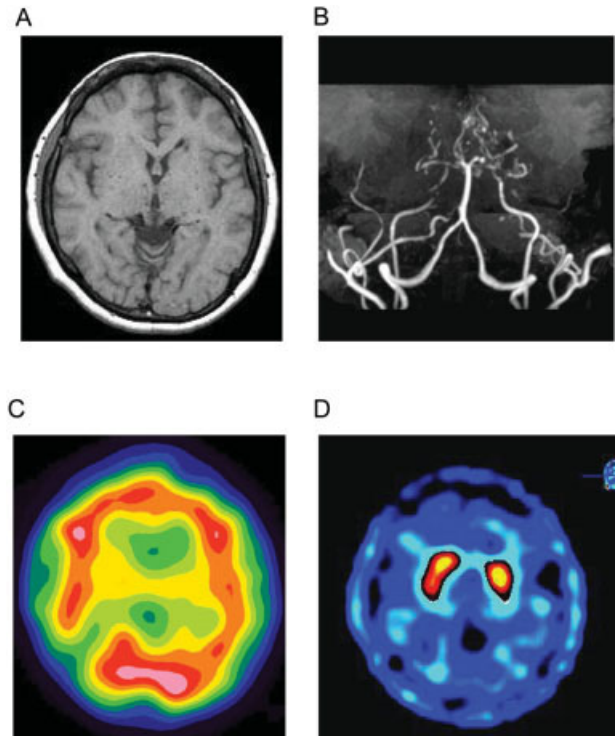


FIG. 1. A: T1-weighted MRI of the brain disclosed multiple dot-like flow voids in the bilateral basal ganglia. B: MRA revealed severe stenosis of both internal carotid arteries at supraclinoid portion with numerous collateral vessels. C: ^{99m}Tc-HMPAO brain SPECT revealed hypoperfusion on the right occipital and high frontal regions. D: TRODAT-1 brain SPECT demonstrated reduced uptake in striatum bilaterally.

cuits. Any interruption of the basal ganglia-thalamocortical circuits might cause the involuntary movements. Some reports have shown evidence of ischemic lesions in the frontal and subcortical structures or basal ganglia.⁵⁻⁷ In our case, although no ischemic changes were found on MRI, ^{99m}Tc-HMPAO SPECT revealed hypoperfusion on the right occipital and high frontal regions. Furthermore, TRODAT-1 brain SPECT demonstrated reduced striatal uptake bilaterally. The choreoathetotic and dystonic movements of left limbs only might be due to relative ischemic changes in right basal ganglia-thalamocortical circuits. To our knowledge, this presentation of MMD with hemichoreoathetosis and hemidystonia in an older adult is very rare.

Acknowledgments: We thank Dr. Anthony E. Lang for providing helpful comments.

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