

Vascular Parkinsonism with Dystonia in Moyamoya Disease: An Expansion of Movement Disorder Phenomenology

Dear Sir,

Moyamoya disease (MMD) is a chronic occlusive cerebrovascular disease with unknown etiology which is characterized by steno-occlusive changes at the terminal portion of the internal carotid artery (ICA) and a characteristic leptomeningeal collateral vessel at the base of the brain.^[1] About 6% of patients develop movement disorders that include chorea (most common), dystonia, paroxysmal dyskinesia as the presenting symptom or during the course of the illness.^[2,3] Reports of vascular parkinsonism in MMD are scarce. Hereby, we report a 21-year-old lady who had a history of carotid artery transient ischemic attack (TIAs), two episodes of hemiplegia (right and left) with ischemic infarcts in the left internal carotid artery territory, and right capsule-ganglionic bleed. Magnetic resonance angiogram and digital subtraction angiography were suggestive of MMD. She developed lower cranial-cervico-bibrachial dystonia with freezing of gait, appendicular rigidity, and bradykinesia suggestive of vascular parkinsonism.

CASE 1

A 23-year-old lady presented with a history of recurrent neurological symptoms in the form of TIAs – three episodes

lasting less than 30 min (right hemiplegia with dysarthria) 4 years back at the age of 19 years. Six months later, she developed right hemiplegia with motor aphasia which lasted for 2 months and gradually improved. Brain magnetic resonance imaging (MRI) showed an infarct in the left distal internal carotid artery (ICA). Three months after the right hemiplegia, she had a mild headache followed by left hemiplegia which improved completely in 4 weeks. Computed tomography head showed right basal ganglia bleed. There was progressive worsening of gait in the form of slowing in the speed, short-stepped with start hesitation, and motor blocks while walking over the next 3 years. Subsequently, she developed dystonic posturing of upper limbs and torticollis. She had residual motor aphasia. Systemic examination was unremarkable. Neurological examination showed that she was conscious, cooperative, and oriented to time, place, and person. She had motor aphasia. The cranial nerve examination was normal. Deep tendon reflexes were brisk in all limbs with extensor plantar response. There was symmetrical appendicular bradykinesia and rigidity in all limbs. She had jaw opening dystonia, lingual dystonia, cervical dystonia with left torticollis and right laterocollis, and bilateral brachial dystonia [Video 1]. There was difficulty initiation of gait

and was slow, with reduced arm swing, freezing of gait, and postural instability [Video 2].

Complete blood count, renal, liver, and thyroid function tests were normal. Serum vitamin B12, folate, and homocysteine levels were normal. Peripheral smear for sickle cells and hemoglobin electrophoresis was negative. Serum ammonia and lactate levels were normal. Serological tests for human immunodeficiency virus, hepatitis B, and C were negative. Anti-nuclear antibodies profile was negative. The brain MRI showed chronic infarcts in the left anteromedial frontal lobe and chronic hematoma in the right putamen with narrowing of the bilateral supraclinoid internal carotid artery (ICA) with attenuated caliber of bilateral anterior and middle cerebral arteries (ACA and MCA) [Figure 1]. Digital subtraction angiography showed thinning of the supraclinoid portion of both ICA, A1 and M1 segments of ACA and MCA with multiple basal moyamoya collaterals from lenticulostriate arteries and thalamogeniculate arteries suggestive of MMD (Suzuki grading stage II) [Figure 1]. MMD was managed conservatively. There was no response to levodopa challenge (200 mg). She was started on levodopa (maximum tolerated dose of 450 mg/day) and trihexyphenidyl for dystonia with no response at the end of 3 months follow-up.

MMD has a bimodal age distribution with peaks in the first and fourth decades. It is more frequent in Asians and females.^[4] The clinical manifestations include TIA, ischemic

stroke, hemorrhagic stroke, seizures, headache, and cognitive impairment.^[5] Movement disorders in MMD include chorea (most common), dystonia, paroxysmal dyskinesia which can occur as the presenting symptom or during the course of the illness. Reports of vascular parkinsonism (VP) in MMD are scarce. VP is characterized by bradykinesia and rigidity involving mainly the lower extremities associated with gait difficulty. There is one report of vascular parkinsonism in MMD reported by Tan EK, *et al.* (2003).^[6] They reported a 57-year-old lady with slowness in walking and activities of daily living. Examination showed rigidity, bradykinesia, mild postural instability, and brisk reflexes. Perfusion-weighted MRI brain showed hypoperfusion in bilateral basal ganglia and frontoparietal subcortical area. The patient was treated with levodopa 750 mg/day with no improvement in gait. Baik JS, *et al.* (2010)^[4] reported four cases of movement disorders in association with MMD and reviewed 38 patients from other studies. A total of 27 patients (64.3%) had chorea, 4 (9.5%) had dystonia, 4 (9.5%) had both, and 7 (16.7%) had dyskinesia. Out of four patients with dystonia, one patient had hemidystonia, and the remaining three had focal dystonia affecting a leg, an arm, or neck. Lesions were found in the putamen, internal capsule, and thalamus in patients with dystonia. Our patient had bilateral appendicular bradykinesia, rigidity of all limbs, and freezing of gait in the form of start and turning hesitation. She had lower cranial dystonia, cervical and bibrachial dystonia

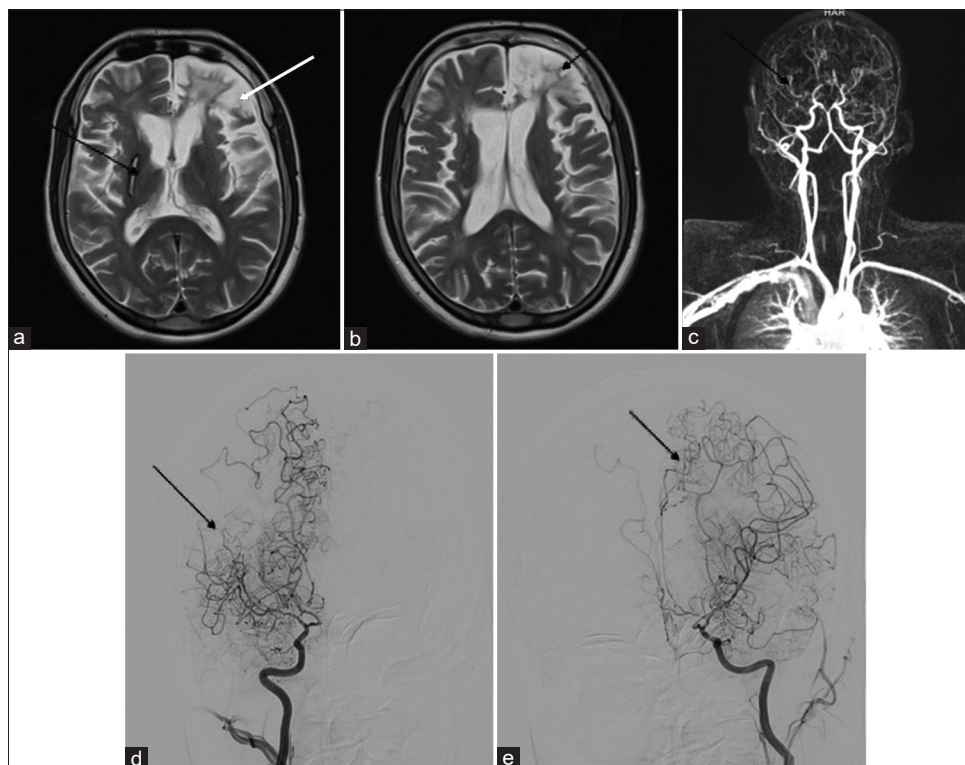


Figure 1: Brain MRI (a) axial T2-weighted image showing gliosis in the left antero-medial frontal lobe (white arrow) and right putamen gliosis with hemosiderin rim (black arrow); (b) axial T2 image showing gliosis in the left antero-medial frontal lobe (black arrow); (c) MR angiogram showing bilateral ICA, ACA, and MCA stenosis (black arrow); DSA right ICA (d) and left ICA (e) showing distal ICA, ACA, and MCA stenosis with extensive collaterals (black arrow)

suggestive of segmental dystonia. Old vascular lesions were found in the left anteromedial frontal lobe and right putamen. Parkinsonism and dystonia are due to the involvement of basal ganglia secondary to ischemia and vascular insult.

The movement disorders described so far in MMD are mainly chorea followed by dystonia in isolation or combination. The combination of vascular parkinsonism and segmental dystonia has not been described so far. This case expands the movement disorder phenomenology in MMD.

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Conflicts of interest

There are no conflicts of interest.

**Rohan R. Mahale, Rohin Dubbal, Hansashree Padmanabha,
Pooja Mailankody**

Departments of Neurology, National Institute of Mental Health and
Neurosciences (NIMHANS), Bengaluru, Karnataka, India

Address for correspondence: Dr. Rohan R. Mahale,
Department of Neurology, National Institute of Mental Health and
Neurosciences (NIMHANS), Bengaluru - 560 029, Karnataka, India.
E-mail: rohanmahale83@gmail.com

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