Vertebrobasilar Insufficiency Presenting as Orthostatic Cerebellar Ataxia

Sir,

Limb-shaking episodes are a rare, but well-known presentation in transient ischemic attack (TIA).[1] Owing to their clinical similarity to seizures or convulsive disorder, they are often mistaken for one of these conditions. The etiology is widely attributed to an impairment of blood supply to the central nervous system as a result of vascular stenoocclusion. Limb-shaking TIA has been linked to vascular stenoocclusion, but is less frequently linked to changes in the body position from supine to upright.[2-4] In addition, isolated postural whole-body shaking movement has not been reported with a video clip. Herein, we report a patient with repetitive truncal and limb ataxia induced by orthostatic positioning, which was attributable to poor vertebrobasilar circulation.

A 79-year-old man was admitted to the emergency room with sudden onset of dysarthria with body shaking, which only occurred when standing up, and soon disappeared after lying down. He had been diagnosed with hypertension, diabetes mellitus, ischemic stroke, and ischemic heart disease, and was taking dual antiplatelet (aspirin 100 mg and clopidogrel 75 mg) and lipid-lowering (atorvastatin 10 mg) medication. He denied any recent head trauma or previous seizure-like episodes. Physical and neurological examinations were unremarkable while the patient was in the supine position. The symptoms occurred specifically in head-up body positions

such as sitting or standing [Video Clip 1]. The symptoms were so severe that he had to grab the side-rail of this bed to keep himself from falling and were aggravated when he attempted to maintain his head-up posture. All the phenomena, however, gradually disappeared a few minutes after changing to a supine position.

Neurological examination revealed that he had profound cerebellar dysfunction, including cerebellar dysarthria and generalized ataxia, with limb dysmetria and truncal rocking. His gait showed severe disequilibrium, wherein he ended up collapsing and falling after trying a few steps with a wide base and titubation. The results of laboratory tests, including complete blood count, routine biochemical analysis, and tests for glucose levels and thyroid function, were all unremarkable. There was no evidence of suspected orthostatic hypotension in a 3-position blood pressure test. Electroencephalography in the middle of an ongoing orthostatic episode showed simple intermittent slowing in bilateral hemispheres without epileptic discharges. Brain diffusion-weighted image (DWI) showed a single small diffusion restriction in the right cerebellum [Figure 1a]. Magnetic resonance angiography (MRA) showed severe stenoocclusion involving the whole vertebrobasilar vasculature [Figure 1b], which was difficult to trace on time-of-flight images. The severity of stenosis was markedly progressed than that on images taken 7 years before.

He received treatment for an ischemic stroke, including hydration therapy, antiplatelet, and lipid-lowering medication under the diagnosis of severe vertebrobasilar insufficiency (VBI) with orthostatic ataxia. After massive volume expansion with 2 L of normal saline daily and increasing the dose of atorvastatin from 10 to 80 mg, these symptoms partially recovered to some extent so he could walk by holding the side-rail of the bed, but the orthostatic ataxia lasted for several days. Follow-up DWI at 5 days [Figure 1c, 1d] revealed a further extension of the previous acute infarction in the right cerebellum and newly developed diffusion restrictions in both cerebellar hemispheres and pons. On hospital day 6, cilostazol (50 mg twice daily) was administered, and all manifestations began to disappear 3 days later. He was discharged without neurological abnormalities. A year later, he reported a brief episode of orthostatic ataxia. Follow-up brain imaging was performed to identify a possible new cerebral event, but DWI did not show any new acute ischemic lesions, while MRA showed that the patency of the vertebrobasilar vasculature was much improved [Figure 1e].

Orthostatic cerebral ischemia in the posterior circulation has been rarely reported so far. Caplan and Sergay described two cases with repetitive presentation of sudden dizziness, diplopia, loss of consciousness, Horner's syndrome, or unilateral ataxia.[5] Both cases showed a clear orthostatic nature and had severe basilar artery stenosis in common. Markedly decreased blood perfusion on the basis of severe stenosis in relevant arteries is believed to be responsible for the emergence of orthostatic cerebral ischemia. Under normal conditions,

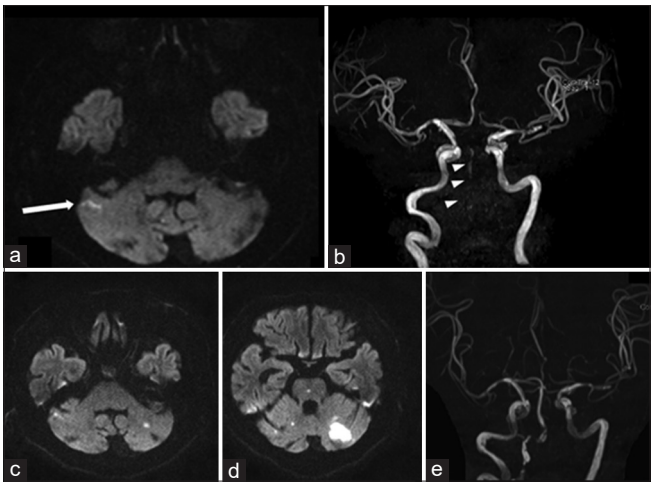

Figure 1: Initial brain magnetic resonance image revealed tiny acute infarction in the right cerebellum on the day of admission (arrow) (a) and severe stenoocclusion throughout the whole vertebrobasilar artery system (white arrowhead) (b). Follow-up diffusion-weighted images (DWI) on the hospital day 5 showed extension of preexisting acute infarction in the right cerebellum (c) and additional new acute infarctions in both cerebellar hemispheres and pons (d). Follow-up magnetic resonance angiography at 1 year after admission demonstrated much-improved patency of vertebrobasilar vasculature (e)

cerebral autoregulation copes with the hemodynamic variations caused by postural changes to ensure a stable cerebral blood perfusion. However, cerebral vasomotor activity is impaired in patients with limb-shaking TIA.[6] Single-photon emission computed tomography and ultrasound examination demonstrated marked attenuation of cerebral blood perfusion along with impaired vasoreactivity in relation to standing in patients with limb-shaking TIA.[7-9]

The clinical improvement achieved by efforts to restore cerebral blood perfusion, including stenting or endarterectomy, maintenance of a stable blood pressure (BP) level, and reduction of antihypertensive agents, supports the critical role of low perfusion in orthostatic cerebral ischemia.[7,10] A majority of patients reporting orthostatic cerebral ischemia underwent treatment with either endovascular revascularization or maintenance of stable BP. We observed favorable clinical outcomes, no sequelae with few recurrences, and spontaneous revascularization of the vertebrobasilar vasculature by stepping up the triple antiplatelet therapy. In addition to adequate hydration, stable BP control, and interventional procedures, an aggressive antiplatelet treatment regimen might be beneficial in clinical TIA patients with severe vertebrobasilar stenosis.

Thus, isolated generalized involuntary shaking movement with an orthostatic nature is an uncommon form of limb-shaking TIA in the posterior circulation that should be promptly recognized and thoroughly evaluated to ameliorate the recurrent attacks and reduce permanent damage in the brain. Active treatment approaches, including adequate hydration, stable BP control, interventional procedures, and intensification of the antiplatelet regimen, should be fully considered.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

Geun Soo Kim, Hong-Kyun Park, Pamela Song, Jae Jung Lee, Joong-Yang Cho

Department of Neurology, Inje University Ilsan Paik Hospital, Inje University College of Medicine, Goyang, South Korea

Address for correspondence: Dr. Joong-Yang Cho, Department of Neurology, Inje University Ilsan Paik Hospital, Inje University College of Medicine, 170, Juhwa-ro, Ilsanseo-gu, Goyang-si, Gyeonggi-do, 10380, South Korea.
E-mail: joongyangcho@gmail.com

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