CASE REPORT

Alternating Paroxysmal Hemiballism-Hemichorea in Bilateral Internal Carotid Artery Stenosis

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Abstract

A 72-year-old man presented with paroxysmal and transient involuntary movements, or “limb shaking”. The attacks occurred alternately from one side of the body to the other and ceased spontaneously. Surface EMG study showed synchronous grouping discharges in multiple limb muscles, being compatible with hemiballism-hemichorea. Cerebral angiography demonstrated marked stenosis of the bilateral internal carotid arteries. Bilateral carotid endarterectomy led to complete disappearance of the involuntary movements. Alternating paroxysmal hemiballism-hemichorea might be a transient ischemic attack, and alternating striatal dysfunction induced by cerebral hemodynamic or microembolic ischemia probably plays a central role in the occurrence of such involuntary movements. (Internal Medicine 40: 808-812, 2001)

Key words: limb shaking, transient ischemic attack, carotid endarterectomy

Introduction

Hemiballism-hemichorea is well known to occur in patients with poorly controlled diabetes mellitus, cerebrovascular diseases and other CNS events, and is attributed to lesions in the basal ganglia such as the caudate nucleus and putamen or in the subthalamic nucleus (1-6). This syndrome usually appears in unilateral limbs contralateral to CNS lesions, and bilateral or alternating ballism-chorea is a very rare manifestation (7, 8). On the other hand, “limb shaking” has been described as a syndrome of carotid transient ischemic attacks (TIAs) (9-15). We report herein a patient with alternating paroxysmal hemiballism-hemichorea, or alternating limb shaking, as a manifestation of a TIA induced by bilateral internal carotid artery stenosis, with a discussion on its pathomechanism.

Case Report

A 72-year-old man was admitted to our hospital in November 1999 because of alternating paroxysmal attacks of involuntary movements in the upper and lower extremities since December 1998. The involuntary movements first occurred suddenly in his left hand lasting a few minutes. There was no trigger for the movements, and he could not induce the movements by himself. In July 1999, oral dyskinesia started continuously. The attacks of involuntary movements gradually worsened, and in September 1999, the attacks consisted of twitching or shaking of the right upper and lower extremities, with a duration of about half an hour. The attacks occurred two to three times a day, and in October 1999, they also occurred in the left extremities, subsequently progressing to alternating paroxysmal attacks. He was a non-smoker. He had an eight-year history of hypertension with administration of a calcium channel blocker. There was no history of cardiac arrhythmia, diabetes mellitus, hyperlipidemia or prior neuroleptic exposure.

On admission in November 1999, his blood pressure was 150/90 mmHg and his pulse was regular at 82/min. The patient was alert and non-demented. There were bilateral cervical vascular bruits. Cranial nerves were intact, but he showed continuous oral dyskinesia. He had no weakness or spasticity; his plantar responses were flexor and the sensory function normal. His gait was intact. During the attacks, his proximal arm twitched or jerked rapidly, his forearm pronated and supinated, and his fingers twisted. His leg on the same side was also shaking, all of which indicated hemiballism-hemichorea. His eyes often twitched conjugately to the right or left. There were no obvious precipitating conditions. One series of the attack lasted about one to three hours and ceased spontaneously. The movements were usually limited to one side of his body, but the side alternately changed in each attack, and occasionally changed from one side to the other side during one series of the attack. The patient had no orthostatic hypotension. Normal laboratory studies included complete blood cell count, urinalysis, routine blood chemistry, hemoglobin-A1c, thyroid and parathyroid functions, clotting and coagulating studies and CSF. Anti-
nuclear, anti-DNA and anti-cardiolipin antibodies were negative. His chest X-ray film was normal, and ECG showed sinus rhythm with occasional ventricular premature beat. EEG was unremarkable without evidence of epileptiform discharges. Surface EMG analysis of the left upper and lower limb muscles during the attack showed involuntary grouping

<table>
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<th>Muscles</th>
<th>Activity</th>
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<td>Deltoid</td>
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<td>Biceps brachii</td>
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<td>Triceps brachii</td>
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<td>Flexor carpi radialis</td>
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<tr>
<td>Extensor carpi radialis</td>
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<td>1st dorsal interosseus</td>
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<td>Abductor pollis brevis</td>
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<td>Rectus femoris</td>
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<td>Hamstring</td>
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<td>Tibialis anterior</td>
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<td>Gastrocnemius</td>
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<td>Extensor digitorum brevis</td>
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<td>Flexor hallucis brevis</td>
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Figure 1. Surface EMG findings in the left upper and lower limb muscles during the involuntary movements. Relatively large EMG bursts occurred synchronously in the upper and lower limb muscles, and small and repetitive bursts were seen in the forearm and hand muscles.

Figure 2. T2-weighted brain MR images of the patient showing multiple small high intensity spots in the cerebral white matter and basal ganglia. R and L represent right and left, respectively.
discharges synchronously occurring in multiple muscles (Fig. 1). In the upper extremity, a relatively large EMG burst of several hundred msec in duration occurred synchronously in all upper limb muscles. In addition, small and irregularly repetitive bursts were seen in the forearm and hand muscles. In the lower extremity, the EMG bursts lasted for about one hundred msec to several seconds, with synchronicity for multiple muscles.

Brain T2-weighted MR images demonstrated multiple small high intensity spots in the cerebral white matter and basal ganglia without Gd-enhancement, suggesting chronic cerebral ischemia (Fig. 2). The subthalamic nucleus appeared intact. Brain single photon emission computed tomography (SPECT) by 740 MBq 99mTc-ECD (ethyl cysteinate dimer) showed mild bilateral scattered cortical hypoperfusion (Fig. 3). Four vessel cerebral angiography showed marked stenosis of the bilateral internal carotid arteries immediately distal to the bifurcation (Fig. 4). The right vertebral artery was also diffusely stenotic due to arteriosclerosis, and the left vertebral artery seemed unremarkable. The anterior choroidal arteries and perforators of the middle cerebral arteries appeared intact.

At first, ticlopidine hydrochloride was orally administered,
which resulted in a tendency to reduce the attack frequency. After obtaining the written informed consent from the patient and his wife, right carotid endarterectomy was performed in December 1999. The hemiballism-hemichorea and oral dyskinesia disappeared after the operation, but his subjective sense of moving of the left hand remained. Left carotid endarterectomy was done in January 2000, leading to complete disappearance of the involuntary movements. Over a 6-month follow-up after the operation, he has been completely free from the involuntary movements.

**Discussion**

From the clinical observation, the abnormal involuntary movements could be expressed as ballism, chorea or athetosis, manifestations of basal ganglia disorders, which were also consistent with the surface EMG findings (16). Hemiballism-hemichorea is known to occur in non-ketotic hyperglycemia or CNS lesions such as stroke and encephalitis, and is usually limited to one side of the body contralateral to the lesions (3–6). These types of abnormal movements are often continuous, lasting for weeks or months. The unilateral striatum or subthalamic nucleus is considered as the causative region. On the other hand, “limb shaking” is a term used for a rare manifestation of carotid TIAs (9–15). Detailed reports of neurophysiological evaluation of limb shaking have been few, but many papers have described the involuntary movements of “limb shaking” as ‘jerking,’ ‘shaking,’ ‘swinging,’ ‘twitching’ or ‘wavering’ of the extremities. In which categories of involuntary movements these movements should be classified, however, remains to be clarified, although tremor or reciprocal rhythmic movement can be at least ruled out. Yanagihara et al used the term ‘repetitive involuntary movement,’ but this also does not precisely express the phenotype of the movements (10). The involuntary movements in our patient could be defined as hemiballism-hemichorea, which was, to the best of our knowledge, reported first as a phenotype of the movements (10). The involuntary movements of “limb shaking” as ‘jerking,’ ‘shaking,’ ‘swinging,’ ‘twitching’ or ‘wavering’ of the extremities should be classified, however, remains to be clarified, although tremor or reciprocal rhythmic movement can be at least ruled out. Yanagihara et al used the term ‘repetitive involuntary movement,’ but this also does not precisely express the phenotype of the movements (10). The involuntary movements in our patient could be defined as hemiballism-hemichorea, which was, to the best of our knowledge, reported first as a phenotype of the movements (10).

Alternating paroxysmal hemiballism-hemichorea is a very rare manifestation. Limb shaking as a carotid TIA usually occurs unilaterally even though patients have bilateral stenosis or occlusion in the carotid arteries. A few cases reported previously involved bilateral limb shaking (9, 10, 14), but there have been no cases of alternating limb shaking. Although pathomechanisms for the alternation of movement side could not be proven, the side alternation suggests that ischemia affects only one hemisphere and that perfusion within the contralateral hemisphere is preserved during the attack. When considering the exhausted vasomotor reactivity reported in patients with limb shaking (14), however, reactive blood flow increment within the ipsilateral hemisphere to the limb shaking side is unlikely. To date, the main discussion related to limb shaking has emphasized a hemodynamic mechanism, but the asymmetry might also suggest an additional microembolic mechanism from atheromatous foci of carotid arteries.

The reason as to why hemiparesis seldom occurs during limb shaking remains to be solved. Although previous radioisotope perfusion studies during an interictal phase showed hyperperfusion of a relatively wide brain area compatible with decreased carotid flow as in our patient (11, 12, 15), there have been no reports of a perfusion study during the ictal phase. The striatum is known to be vulnerable to ischemia or hypoxia. Hypoxia-ischemia induces the release of neurotransmitters including dopamine, which plays a central role in neuronal death during cerebral ischemia (17, 18). After hypoxia-ischemia, neurons in the neonatal striatum show membrane depolarization and rapid degeneration (19, 20). The extrapyramidal manifestations or hemiballism-hemichorea, without hemiparesis, in carotid TIAs might be caused by the selective vulnerability of striatal neurons to oxygen deprivation and their peculiar sensitivity to energy metabolism failure, accompanied by excessive dopamine release (19).

The subthalamic nucleus might be another candidate for the causative lesion of hemiballism (1, 5). This nucleus receives its blood supply from the anterior choroidal arteries branching from the internal carotid arteries, or the posterior communicating arteries, and infarction of the subthalamic nucleus induces continuous hemiballism. However, the fact that the oral dyskinesia in the present patient also disappeared after the operation suggests that the responsible lesion for the attacks was the striatum rather than the subthalamic nucleus (21).

To argue from a neurophysiological point of view, defective putaminal inhibitory outflow to the lateral pallidum induces oversuppression of the subthalamic nucleus and a concomitant reduction in pallidothalamic inhibitory outflow. This results in an overactivity of thalamocortical excitatory projections and consequent hyperkinetic syndrome.

Limb shaking, or paroxysmal hemiballism-hemichorea, is a syndrome of carotid TIA that should not be misdiagnosed as other neurological disorders such as simple partial seizures (22) or Sydenham’s chorea. Conservative treatments such as administration of antiepileptic agents or haloperidol are not effective. Clinicians should always consider the existence of underlying carotid occlusive disease and perform surgical intervention as a possible radical treatment.

**References**

8) FURIE R, Ishikawa T, Dhawan V, Eidelberg D. Alternating hemichorea in


