

CAROTID TRANSIENT ISCHEMIC ATTACKS PRESENTING AS LIMB-SHAKING SYNDROME

Report of two cases

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ABSTRACT - Limb shaking syndrome (LSS) is a rare presentation of transient ischemic attacks (TIAs), usually secondary to a critical carotid stenosis compromising intracranial circulation, first described 40 years ago. Two additional cases are described herein, aiming to add on to previous descriptions, and to warn physicians about this potentially harming and rather uncommon condition.

KEY WORDS: limb-shaking syndrome, carotid stenosis, tremor, paroxysmal dyskinesias, cerebral hypoperfusion, transient ischemic attacks.

Ataques isquêmicos transitórios apresentando-se como síndrome do membro tremulante: relato de dois casos

RESUMO - A síndrome do membro tremulante, inicialmente descrita há 40 anos, é apresentação rara de ataques isquêmicos transitórios, sendo usualmente secundária a estenose carotídea crítica comprometendo a circulação intracraniana. Dois novos casos são aqui descritos, visando ilustrar e discutir esta condição incomum e com alto potencial de morbidade.

PALAVRAS-CHAVE: síndrome do membro tremulante, estenose carotídea, tremor, discinesias paroxísticas, hipoperfusão cerebral, ataques isquêmicos transitórios.

Transient ischemic attacks (TIAs) are signs of impending definite brain infarction in persons with atheromatous stenosis in any of the supplying vessels of the brain or with heart disease (valvular disease, atrial fibrillation and mural thrombi). Symptoms usually comprise visual or hemispheric rapidly resolving deficits, such as transient monocular blindness (*amaurosis fugax*) and motor and/or sensitive deficits¹. Thrombi development, embolization and clot resolution underlie this clinical presentation, hypotension and global reduction of cerebral blood flow (CBF) playing only a limited role. Transitory movement disorders have been described in the chapter on TIAs as position-related shaking of arms and legs, associated with a high-grade stenosis of contralateral carotid artery^{2,3}. We present two additional cases, calling attention to the relevance of the diagnosis.

CASE

Patient 1: A 73 year-old woman, three months before the first evaluation, started presenting brief spontaneous episodes of rapid shaking of the left arm, described as a "tremor". These movements, initially sparse, progressed to daily attacks lasting a few seconds. She also reported a single short-lasting episode of spatial disorientation,

with spontaneous resolution and a single episode of syncope while inside a church. Medication used included telmisartan 40mg/day, atorvastatin 10mg every other day and clorthalidone 12.5mg/day. Vital signs and general physical examination were normal, and there was no orthostatic hypotension. Neurological examination disclosed a +++/4 bruit over right carotid artery, but was otherwise unremarkable, with no signs of abnormal postures, gestures or movements. MRI demonstrated lacunar infarctions on the right hemisphere and extracranial Doppler scanning disclosed a 95% stenosis of the right internal carotid artery. Without further investigation but with brain protection therapy, the patient underwent an endarterectomy and remained asymptomatic in the following five-month follow-up period.

Patient 2: A 73 year-old man, who regularly came to our outpatient clinic for diabetic neuropathy, complained in July of 1999 a single episode of right arm paresis, self-limited to 15 minutes, with normal muscular strength being restored thereafter. At that time, he used to take fludrocortisone 0.1 mg thrice a day. On physical examination, his supine blood pressure was 100/60 mmHg, whereas at a standing position it was 70/50 mm/Hg. During orthostatic blood pressure measurement, a right leg gross tremor was noted, subsiding when the patient sat down. The patient denied ever noticing such an abnormal movement. The rest of the neurological examination disclosed deep

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tendon areflexia, decreased superficial and vibration senses in the lower limbs and a Romberg sign, all interpreted as related to diabetic polyneuropathy. Magnetic resonance angiography disclosed a critical stenosis of the left cavernous carotid artery, and a 60% stenosis of the right cavernous carotid artery. The patient was submitted to left carotid stenting and became asymptomatic both to right arm TIAs and to right leg limb-shaking after two years of follow-up.

DISCUSSION

Carotid stenosis is not a relevant clinical finding in most instances of movement disorders, and paroxysmal hyperkinesias do not constitute an exception. Moreover, clinicians usually relate transient focal clonic jerks in patients with cerebrovascular disease to symptomatic epilepsy. Nevertheless, since Miller-Fisher's first description of temporary limb-shaking syndrome (LSS) associated to contralateral carotid stenosis¹, this diagnosis has been regularly reported throughout the years. Nowadays, about 45 cases of this condition are found in the literature^{2,3}.

The clinical features of LSS comprise a group of rhythmic or arrhythmic involuntary hyperkinesias affecting the hand, arm, leg, hand-arm or hand-arm-leg unilaterally. When the paroxysms are present on hand-arm-leg, there is no Bravais-Jacksonian march and it always spares the face muscles; upper limbs are more evidently affected. Occasionally, jerking may be present in both arms, although in an asymmetrical fashion. Isolated leg involvement is also uncommon. Most often, limb-shaking is regular in frequency⁴. Patients describe movements as "jerking", "swinging", "shaking", "trembling", "twitching", "flapping", "wavering", "inability to control the arm" and "lack of coordination"⁵. When observed by an examiner, LSS may assume either a choreic⁶ or a coarse tremor-like appearance. Limb-shaking spells occur with variable frequency: patients report abnormal movements from a single episode to many times a day⁴. Associated symptoms may encompass ataxia, myoclonic jerks, dystonic limb posturing, and parkinsonism, the latter manifested as micrographia, paroxysmal tremor and rigidity, in addition to ataxia, dysarthria and nistagmus³. According to Gálvez-Jiménez et al., LSS is considered to be one of the various secondary or symptomatic dyskinesias of vascular etiology³.

An almost invariable clinical clue is that symptoms arise after maneuvers that theoretically provoke cerebral blood hypoperfusion, such as arising from a bed or a chair, as well as hyperextending the neck. A patient noticed a repeated tendency to drop the keys when leaving his car, with an associated tremor-like sensation and clumsiness of his left arm². There is a short latency between standing and symptom starting, usually of a few seconds. Conversely, limb-shaking, which lasts from a few seconds to a few minutes, ceases when the patient sits or lies down. These characteristics were lacking in one of our patients and, in the other, limb-shaking was first noticed at the neurological clinic. Other combined neurological features to remind the clinician of the vascular nature of LSS are transient

dysphasia and/or dysarthria, numbness of the shaking extremity and ipsilateral hemiparesis.

There is no single pattern of lesion on imaging studies, which eventually disclose signs of small vessel disease, both bilaterally or confined to the hemisphere contralateral to the abnormal movements. Nevertheless, these studies may range from normal to old infarcts in regions topographically related to limb-shaking. In a series of 12 patients analyzed by Yanagihara et al.⁴, from nine patients who underwent brain CT scan, four had normal exams, one had generalized atrophy and the remaining had single small lesions contralateral (three patients) and ipsilateral (one patient) to the shaking limb. Similarly, Baquis et al. found six normal CT scans in eight patients, while the other two had old infarcts⁵.

Electroencephalographic studies failed from the start to show epileptiform activity associated with LSS, although some patients had focal contralateral slow activity⁶. Induction of repetitive involuntary movements in nine electroencephalographically monitored patients rendered abnormal findings in two. One had diffuse delta slowing, and the other temporal slowing of the same type⁴.

The most striking feature of LSS is severe stenosis of the internal carotid artery, raising the recommendation to look for carotid bruits in the elderly with orthostatic and/or episodic movement disorder. Most patients have critical stenosis, while some present an occluded carotid artery. All 12 patients described by Yanagihara et al had occluded or critically compromised contralateral carotid artery. The ipsilateral artery had less than 50% stenosis in seven and severe stenosis in five⁴. However, small vessel disease with normal carotid angiography and discrete thalamic and midbrain infarction have also been reported as a cause of LSS³. For defining stenosis, noninvasive methods might be preferable, since LSS may be triggered by carotid angiography³.

Other paroxysmal dyskinesias than limb shaking may be presented as a symptom of vascular insufficiency⁷. Choreoathetosis^{8,9}, limb spasms¹⁰ and hyperkplexia¹¹ have been reported³.

Limb shaking, like the other vascular paroxysmal dyskinesias, can be explained by the "hypoperfusion theory", in which carotid stenosis and orthostatism lead to decreases in cerebral blood flow in critical watershed territories. Bogousslavsky and Regli, in an assessment of 51 patients with infarcts in watershed cerebral territories, found 12% of focal limb-shaking⁶. The association between LSS and watershed area hypoperfusion was further analyzed by physiological studies. Using xenon-133 for detection of regional decreases of cerebral blood flow during CT scan, Tatemichi et al found significant hypoperfusion of the right dorsofrontal and upper rolandic regions contralateral to the shaking limb in a 63 year-old patient¹². Similarly, by means of transcranial Doppler sonography, vasomotor reactivity has been shown to be impaired on physiological stimulus with hypercapnia¹³. PET scan imaging in a

patient with LSS disclosed acetazolamide-induced hypoperfusion of corresponding cerebral territories².

It ought to be pointed out that although some patients present systemic orthostatic hypotension, this is not the rule and some series do not even report this phenomenon among their patients^{4,13}. The one described by Zaidat et al showed no improvement after reducing the anti-hypertensive drugs². The patient reported by Leira et al, a young male with many comorbidities, ceased LSS after the same measure, but such amelioration was doubtful, because the patient died soon after¹⁴.

The treatment of LSS is aimed to restore the cerebral blood flow, since medical measures, as stated above, are usually of little value¹³. Superficial temporal artery (STA) - medial cerebral artery (MCA) bypass, a surgical technique seldom indicated for secondary prevention of stroke, may be beneficial in such cases¹⁵. The following numbers assemble patients reported by Baumgartner and Baumgartner, Yanagihara et al. and Baquis et al: nine patients underwent endarterectomy: seven with complete symptom resolution, while two died of surgical complications. Ten patients were submitted to STA-MCA bypass, eight of these with good results in limb-shaking and two without it. Vertebral artery balloon angioplasty has been reported as effective in one patient³. From seven persons kept on conservative treatment, five had a good outcome and two died, one of lung cancer and the other of a large brain infarct^{3,4,5,13}. We add to the above list the first patient treated with carotid stenting, who remains asymptomatic after two years of follow-up.

Limb-shaking TIA is a rare syndrome, and its diagnosis ought to be suspected in patients with paroxysmal movement disorders, especially when risk factors for cerebrovascular disease are present. To the moment, there are few data to support conservative measures, and any patient with LSS symptoms and a carotid artery stenosis greater than 70% should be considered for revascularization.

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