Limb shaking presenting as rhythmic involuntary hyperkinetic movements may represent as severe bilateral occlusive carotid disease. This unusual form of transient ischemic attack is often misdiagnosed as focal motor seizures. However, careful assessment reveals a lack of usual seizure characteristics such as a jacksonian march or facial involvement. The movements also appear to be precipitated by activities that lower blood pressure. We present two cases of patients with severe bilateral carotid stenosis leading to limb-shaking transient ischemic attacks. There was complete stenosis in the internal carotid artery (ICA) contralateral to the jerking limb, combined with significant stenosis in the ipsilateral ICA. Cerebral perfusion on the occluded ICA side was maintained through collateral circulation from the opposite ICA and posterior circulation. When blood pressure was lowered orthostatically or by medication, the resulting cerebral hypoperfusion manifested as limb jerking. Recognition of limb shaking as a rare form of transient ischemic attack and differentiating it from focal motor epilepsy can facilitate early identification of critical carotid stenosis, allowing for appropriate interventions and thus reducing the risk of a disabling stroke. We recommend that clinicians should consider carotid disease in elderly patients presenting with orthostatic or episodic movement disorders. [International Journal of Gerontology 2010; 4(1): 47–50]

Key Words: carotid stenosis, limb jerking, limb shaking, low-flow transient ischemic attacks

Introduction

Transient ischemic attacks usually present with motor and/or sensory deficit or with visual loss. Involuntary hyperkinetic movements of the limbs are not generally regarded to be a feature of cerebral ischemic episodes. Moreover, carotid occlusive disease is not generally considered in the differential diagnosis when patients present with such movement disorders. Because damage from cerebral ischemia is the most common cause of symptomatic epilepsy in the elderly, this diagnosis is most often suspected when encountering such stereotypical movements.

In 1962, however, Fisher described a syndrome of transient limb shaking associated with contralateral carotid stenosis. Since then, this phenomenon has been reported in over 50 cases worldwide. Early recognition of this unusual form of transient ischemic attack is important, as these patients with severe occlusive carotid disease are at high risk of stroke.

Case Reports

Case 1

A 76-year-old woman presented with a 1-day history of transient left-sided weakness and slurred speech. She had been started on bendroflumethiazide for hypertension a few days previously. Prior to this, she had noticed intermittent jerking of her left arm upon standing; this was after she had commenced taking an angiotensin-converting enzyme inhibitor. The antihypertensive treatment was part of her secondary prevention...
regime following an episode of left-sided hemiparesis immediately after an aortic valve replacement some 2 months previously.

Her medical history was remarkable for ischemic heart disease and hypertension, and she had been warfarinized because of her prosthetic valve. On initial examination, she had left hemiparesis that resolved a few hours later, and auscultation revealed a systolic flow murmur. Her international normalized ratio was subtherapeutic on presentation at 1.9, and her brain computed tomography showed only an old right frontoparietal infarct. Carotid Doppler and computed tomography carotid angiography demonstrated complete right internal carotid artery (ICA) occlusion with a 60% stenosis of the left ICA (Figures 1 and 2). Posterior circulation was patent, and an echocardiogram did not reveal any vegetation.

Initial treatment comprised heparin infusion and withdrawal of bendroflumethiazide, with gradual resolution of her symptoms. She was referred to our local vascular surgeons given her symptomatic left ICA, but no surgery was planned at the time of writing. She was managed conservatively, with a reduction in antihypertensive treatment resulting in good symptom control.

Case 2
A 69-year-old man presented with intermittent jerking of the left arm and leg upon standing up. Each jerking episode lasted about 30 seconds, and would resolve once the patient sat down. His symptoms were particularly bad whilst on holiday in a warm climate, and he described a particular episode of his left leg giving way when he alighted from a hot bus. Further exacerbation of the jerking episodes followed after the introduction of an angiotensin-converting enzyme inhibitor, and continued when treatment was switched to an angiotensin II antagonist. Frequency of these episodes declined following the withdrawal of these agents.

Past medical history included a right temporal lobe infarct 8 years previously, ischemic heart disease, spinal stenosis, and a colonic resection for carcinoma. He was on several antihypertensive medications and had a noticeable postural drop. Examination was unremarkable except for left central facial palsy. Carotid ultrasound revealed complete stenosis of his right ICA, with 70% stenosis on the left side (Figure 3). He had also undergone neurosurgical review regarding potential intervention for his symptomatic left ICA stenosis, and again a conservative approach was adopted.
Discussion

The jerking movements in limb-shaking transient ischemic attacks can be easily mistaken for focal motor epilepsy. The absence of a jacksonian march and the precipitation of jerking with postural changes (lowering of blood pressure) are some features that can be used to distinguish this phenomenon from focal seizures. Facial muscles are not usually affected, and there is a reported preferential involvement of upper limbs. No epileptic discharge is evident on electroencephalography, and no improvement is demonstrated with anticonvulsant therapy.

The underlying mechanism is speculated to be due to transient focal hemodynamic failure. Cerebral hypoperfusion above the carotid narrowing in the hemisphere contralateral to the side of limb jerking has been described, with loss of cerebral vasoreactivity to hyperventilation in this territory similarly demonstrated.

In our two cases, the ICA contralateral to the jerking limb was completely occluded. In addition, there was significant stenosis in the ipsilateral ICA. To attribute the recurrent limb-jerking events to embolic events, these emboli would need to travel up the partially stenosed ICA ipsilateral to the jerking limb, recurrently crossing over at the circle of Willis, to lodge only in the hemisphere contralateral to the jerking limb.

A more likely mechanism is cerebral hypoperfusion in the hemisphere contralateral to the jerking limb. Cerebral perfusion is maintained through collateral circulation from the partially occluded ICA and posterior arteries. Perfusion is compromised, however, when blood pressure is lowered orthostatically or by medication. The resulting cerebral hypoperfusion manifests as limb jerking on the contralateral side, which may resolve upon sitting or lying down. Although a reduction of blood pressure through postural changes appears to decrease cerebral perfusion, orthostatic hypotension does not seem compulsory for the development of limb jerking.

Management is aimed at improving/maintaining cerebral perfusion through careful blood pressure control and surgical revascularization. In the absence of cardiac or renal disease, blood pressure may even be allowed to rise, leading to an improvement in symptoms. Addressing other stroke risk factors with cholesterol lowering and antiplatelet agents is also beneficial.

Alternatively, surgical revascularization can be considered if comorbidities do not contribute to increases in blood pressure. Carotid endarterectomy can improve symptoms and lower stroke risk in the context of incomplete carotid stenosis. For complete carotid stenosis, however, extracranial–intracranial bypass surgery is suggested, and can lead to a resolution of symptoms but does not reduce the risk of a future stroke.

Carotid surgery, however, is not without risk. A much higher incidence of postoperative intracerebral hemorrhage following carotid endarterectomy has been reported in patients with limb shaking because of severe carotid stenosis (23%), compared with an overall risk of about 0.5% following carotid endarterectomy in general. A reperfusion syndrome due to the combination of an increase in cerebral blood flow with the associated loss of vasomotor reactivity above the stenosis is likely to contribute to the higher risk of bleeding. Accordingly, use of transcranial Doppler ultrasound has been recommended to help assess cerebral perfusion prior to surgery.

In conclusion, limb-jerking transient ischemic attacks are a rare presentation of carotid occlusive disease. It is important to recognize this phenomenon correctly and differentiate it from focal motor epilepsy. This should facilitate early identification of critical carotid stenosis, allowing for appropriate interventions and thus reducing the risk of a disabling stroke. We recommend that clinicians should consider carotid disease in elderly patients presenting with orthostatic or episodic movement disorders.

References