Short report

McArdle’s sign in multiple sclerosis

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SUMMARY A patient with multiple sclerosis is reported in whom neck flexion produced a reversible deterioration in gait and an increase in the degree of pyramidal weakness of the lower limbs. The name McArdle’s sign is proposed.

Lhermitte’s sign, a peculiar transient sensory disturbance produced by neck flexion, is experienced by about one-third of patients with multiple sclerosis. It seems that Dr MJ McArdle was the first to observe a separate phenomenon, namely that occasional patients with multiple sclerosis would experience a reduction in lower limb strength and impaired mobility during neck flexion, although he never reported this finding (personal communication Drs CJ Earl and P Rudge). We recently saw a case of laboratory-supported definite multiple sclerosis in whom this phenomenon was very clearly demonstrable.

Case report

A 45 year old school teacher was referred in October, 1986 with the diagnosis of a possible cervical cord arteriovenous malformation. At the age of 15 years the patient fell asleep in an awkward position at a cinema; for 1 week thereafter he had a sensorimotor disturbance affecting the right leg and he was admitted to hospital for a lumbar puncture. In 1978 he was seen in a neurology outpatient clinic because of a mild expressive dysphasia which resolved spontaneously after 2 months. In 1981 he noticed that he could not keep apace with his companions during a walk in the Lake District. In 1982 he awoke one morning with considerable neck discomfort and stiffness together with numbness and tingling of both upper limbs. The numbness quickly became confined to the hands and he was unable to write or use ordinary eating implements. Within a few months his dexterity improved so that upper limb disability at the time of admission was limited to picking up small objects, fastening small buttons and identifying coins in his pocket. Progressive difficulty with walking and urgency of micturition began soon after onset of the sensory disturbance. He was admitted to a local hospital for myelography in 1983 and it was noted that he would get an abnormal sensation in the toes of both feet when his neck was flexed. CSF at the time contained 5 mononuclear cells. Since then a neurologist has noted “enormous variability and severity in the patient’s motor symptoms.” On one visit it was noted that “walking was limited to less than 40 yards [36m] after which the patient developed crippling stiffness of the lower limbs with backward extension of the neck.” The patient observed that, when walking, his neck would extend and become stiff. As a result of this he would walk downstairs “blind”. After walking a variable distance he would have to stop “not because of lack of power but because messages don’t seem to be getting through.” After stopping, he would gently bend his neck and massage it. He would then start off again and the whole process would repeat itself. He also noticed reduced exercise tolerance after a hot bath. Gait was spastic and he held his neck in a hyper-extended position. When he bent his neck his gait slowed and he either came quickly to a halt or he stumbled and had to be caught. Romberg’s sign was negative. Cranial nerve examination was normal apart from congenital nystagmus. There was mild bilateral pes cavus. Tone was increased in the lower limbs with sustained ankle clonus. With the neck in extension power was normal but with the neck flexed there was moderate, and mainly proximal, pyramidal weakness of the right leg and he could only just lift the left leg from the bed. The biceps and supinator jerks were symmetric and normal; the triceps and finger jerks symmetric and brisk; the abdominal reflexes preserved; the lower limb reflexes symmetric and very brisk with clonus; both plantar responses extensor. Proprioception was impaired at the
fingers. Vibration sense was absent at the fingers and great toes. There was objective loss of light touch over the fingers.

Full blood count, sedimentation rate, syphilis serology and B12 were normal. Visual and brain-stem auditory evoked potentials were normal. Following both right median nerve and left posterior tibial nerve stimulation there were delayed cortical somatosensory evoked potentials (SEPs). The spinal response was absent following median nerve stimulation. SEPs were not altered by neck flexion. Central motor conduction time (CMCT) was assessed by recording compound muscle action potentials (CMAPs) from abductor digiti minimi (ADM) with electrical stimulation over the C7/T1 interspace and magnetic stimulation of the motor cortex. Following scalp stimulation with the neck in extension, the amplitudes of the CMAPs elicited respectively from right and left ADM were 7% and 6% of that following stimulation of the ulnar nerve at the wrist (normal amplitude following scalp stimulation is 18.6–96.6% of that following wrist stimulation). CMCT was 8.5 ms on the right and 13.4 msecs on the left (normal mean ± SD for CMCT is 6.13 ± 0.89 ms). Neither the amplitudes or latencies of the responses were altered by neck flexion. No response could be obtained when an attempt was made to record from left tibialis anterior following scalp stimulation. Cervical myelography showed minor anterior impressions on the theca at C3/4, 5/6 and 6/7. There was unusually marked cord atrophy at C3/4 which was confirmed on a post-myelogram CT scan. The CSF IgG distribution was oligoclonal. NMR scan revealed discrete lesions in the cerebral hemispheres, around both trigones and in the left pons. There was increased signal in the cord from the lower end of C2 to the lower end of C4 (fig).

Discussion

This patient had cervical spondylosis and multiple sclerosis with clinical, electrophysiological and radiological evidence of demyelination in the upper cervical cord. Brain and Wilkinson observed that in patients with both cervical spondylosis and multiple sclerosis, the disseminated plaques of demyelination were most extensive in those cervical segments which corresponded to the level of the spondylotic bars. They attributed this association to interference with cord blood supply and/or compression of the cord by the bars. In a necropsy study of 18 cases of multiple sclerosis, Oppenheimer found that plaques in the cervical cord were about twice as common as those at lower levels. The plaques were most common in the lateral columns, the typical lesion being fan-shaped in cross-section, tapering upwards and downwards and abutting on the lateral surface of the cord.

During neck flexion the cervical cord is elongated. It has been estimated that the mid-cervical cord is lengthened by 3 cm during neck flexion whilst the length increase measured along the posterior aspect of the cord may be as much as 5 cm. During bilateral hip flexion, as in sitting, the cord is stretched caudally to an even greater extent. Because of tethering by the dentate ligaments, tension is increased in the cord when it is stretched. Nordin et al have demonstrated that it is possible to record bursts of impulses from microelectrodes inserted into median nerve skin fascicles during neck flexion in a patient with Hermitte's sign due to multiple sclerosis, thus supporting the experimental evidence of Smith and McDonald that this phenomenon is due to mechanical stretch-induced impulse generation in axons of primary sensory neurons ascending in the dorsal columns.

In our patient the clinical consequence of neck flexion has been a motor one, the legs becoming stiffer and weaker than they were when the neck was extended. We believe this is due to a stretch-induced increase in the degree of conduction block within demyelinating plaques scattered longitudinally along the corticospinal tracts. Increase in blocking of efferent impulses would be expected to result in a reduction in the amplitude of muscle responses evoked by cortical stimulation or, in the case of severe and preferential blocking of the largest and fastest-conducting myelinated fibres, an increase in latency.
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In our patient, neck flexion did not alter either the amplitude or the latency of responses from ADM following scalp stimulation; however, it was the lower and not the upper limbs which were symptomatically weakened by the change in posture. Unfortunately, we were unable to record CMAPs from the left tibialis anterior of our patient even with the neck in full extension, presumably reflecting further more distal demyelination within the corticospinal pathways.

Why stretch should increase conduction block in a demyelinating lesion is unknown. Alternative explanations could be temporary conduction block produced by cord compression and/or ischaemia due to the associated cervical spondylosis. Compression is more likely to be worse during neck extension when the ligamenta flava are compressed and bulge intraspinally thereby reducing the antero-posterior diameter of the cervical canal. In flexion, the ligamenta flava are stretched and the canal is wider.\(^\text{14}\)^\(^\text{15}\) Compression was excluded in our patient by the normal appearance of the cord on the myelogram taken with the neck in flexion. Ischaemia is considered a less important factor than compression in cervical myelopathy.\(^\text{16}\) In our case the degree of spondylosis was mild and the onset of the clinical phenomenon was too rapid for an ischaemic mechanism.

On the basis of our experience in this and two other patients with probable multiple sclerosis,\(^\text{2}\) we conclude that in occasional patients with slowly progressive cervical cord disease due to multiple sclerosis, neck flexion will produce a reversible deterioration in gait and an increase in the degree of pyramidal weakness of the lower limbs. We suggest that this phenomenon should be called McArdle’s sign since, to our knowledge, he was the first to observe it. As with Lhermitte’s sign, it is likely that McArdle’s sign will not be found to be specific for multiple sclerosis and will also be found to occur in cases of progressive cervical myelopathy due to other causes but in which demyelination, rather than axonal degeneration, is the dominant pathological process.

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**References**
