SHORT REPORT

Propriospinal myoclonus in multiple sclerosis

R Kapoor, P Brown, P D Thompson, D H Miller

Abstract

The clinical and electrophysiological features of segmental myoclonus affecting the right arm and upper trunk are described in a patient with multiple sclerosis. Electrophysiological studies suggested that the myoclonus was propagated from a generator site in the cervical cord, where lesions were found using MRI. The spread of electromyographic activity in each myoclonic jerk was slow and variable. These findings are characteristic of propriospinal myoclonus, which has not been associated with multiple sclerosis previously.

(*J Neurol Neurosurg Psychiatry* 1992;55:1086-1088)

The types of myoclonus that arise from various generator sites in the nervous system can be distinguished on clinical and neurophysiological grounds. In cortical myoclonus, for example, discharges in the sensorimotor cortex are conducted rapidly in the pyramidal tracts,¹ leading to focal or multifocal jerks. In myoclonus of brainstem origin, reticular firing causes more generalised axial jerks due to rostral and caudal spread of activity in reticulospinal pathways.^{2,3} In contrast, spinal myoclonus tends to be focal and segmental, with little spread of activity from spinal generator sites, which produce jerks of longer and more variable duration than those of supraspinal origin.^{4,5}

Recently, an additional form of spinal myoclonus has been described in which a spinal generator recruits axial muscles via slowly conducting propriospinal pathways to produce extensive jerks of the trunk.⁶ Whereas segmental spinal myoclonus can occur with numerous disorders including multiple sclerosis,⁵ few cases of propriospinal myoclonus have been described. In one, a cervical cord lesion was present, but lesions were not found in the others.⁶ We now report a patient with multiple sclerosis in whom propriospinal myoclonus was documented and in whom associated lesions of the cervical cord were found on MRI.

Case report

A right handed white woman developed right optic neuritis when aged 23 years. The following year there was an episode of left optic neuritis. She then noted a fine postural tremor

(which responded to propranolol), together with irregular, involuntary jerking of the right shoulder and arm, and sometimes of the leg. This could be preceded by a tingling sensation over the back of the neck. The muscle jerking persisted, and when aged 25 years she also developed Lhermitte's symptom and altered sensation over the left side of her face. This progressed over four weeks to include urinary urgency, an unsteady gait and finally a burning discomfort of the right leg. There was no additional past or family history of neurological disease, she was otherwise asymptomatic and took no regular medications.

At this point the general and state examinations were normal. The corrected visual acuities were 6/6 on the right and 6/9 on the left, with impaired colour perception on the left (9 of 13 Ishihara plates were identified). There were bilateral, paracentral scotomata and both optic discs were pale. A left relative afferent pupillary defect and convergence nystagmus affecting only the left eye were noted. Spinothalamic sensation was impaired in the first and second divisions of the left fifth cranial nerve, but the corneal reflexes and remaining cranial nerves were normal.

In the limbs, there were spontaneous, irregular jerks of the right shoulder and arm leading to elevation and abduction of the right shoulder associated sometimes with a grunt. They were preceded by a tingling sensation over the neck but could not be suppressed voluntarily. Similar jerks were elicited by taps to the right shoulder and by pinprick over the proximal right arm. More violent jerks spread to involve trunk and hip flexion on the right. Limb tone and coordination were normal but there was mild pyramidal weakness of the left leg. The tendon reflexes were uniformly brisk, the abdominal reflexes present and the left plantar response was extensor. Sensory testing revealed no objective abnormalities in the limbs.

Investigations

All routine haematological and biochemical tests were normal. After the episode of left optic neuritis the auditory evoked potentials were also normal, but the visual evoked potentials were delayed bilaterally (full field pattern evoked P100 latencies 125 ms on the right, 131 ms on the left). The CSF contained 0.58 g/l protein, glucose 3.3 mmol/l (blood glucose 5.1 mmol/l) and 11 white blood cells/mm³ (all mature lymphocytes). Electro-

University
Department of
Clinical Neurology,
National Hospital for
Neurology and
Neurosurgery, Queen
Square, London, UK
R Kapoor
P Brown
P D Thompson
D H Miller

Correspondence to:
Dr Kapoor

Received 18 November
1991.

Accepted 23 January 1992

phoresis revealed IgG oligoclonal bands in the cerebrospinal fluid but not in the blood, and a diagnosis of multiple sclerosis was made.

Electrophysiological studies: Polymyography, somatosensory evoked potentials (SEPs) and magnetic stimulation of the brain were performed using methods described previously.⁶ In spontaneous jerks EMG activity was recorded in the latissimus dorsi, deltoid, triceps, diaphragm, and sometimes trapezius and sternocleidomastoid muscles on the right. In the more violent spontaneous jerks EMG activity was also recorded in the forearm flexor muscles, rectus abdominis and tibialis anterior on the right. The duration of the bursts of EMG activity in the various muscles during spontaneous jerks ranged from 100 to 1000 msec. There was considerable jitter in the relative latencies to onset of EMG activity in different muscles. For example, the interval between the onset of EMG activity in diaphragm and latissimus dorsi ranged from -19 to +19 ms. It was not possible to distinguish a recruitment order in the relative latencies of sternocleidomastoid, trapezius, latissimus dorsi and diaphragm. However, in the more violent and extensive jerks the median latency to onset of EMG activity in rectus abdominis was about 10 msec longer than that in latissimus dorsi or diaphragm, and the median latency to onset of EMG activity in tibialis anterior was about 32 ms after that in latissimus dorsi or diaphragm (fig 1). No time-locked cortical correlate was recorded in the backaveraged EEG activity preceding spontaneous jerks in latissimus dorsi.

The latency to onset and order of recruitment of EMG activity in the reflex response to

taps over the right shoulder was variable. Reflex EMG activity was usually recorded first in the right trapezius and latissimus dorsi, at latencies ranging from 62 to 218 ms (median 130 ms). The latency to onset of EMG activity following cutaneous electrical stimulation of the proximal right arm was also variable. The earliest reflex EMG activity was again usually recorded in the right trapezius and latissimus dorsi, at latencies ranging from 40 to 159 ms (median 83 ms) after each stimulus. Reflex EMG activity was recorded about 15 ms after that in diaphragm following either taps to the right shoulder or cutaneous electrical stimulation of the proximal right arm. EMG activity was not recorded in tibialis anterior in reflex jerks.

SEPs following electrical stimulation of the right median nerve at the wrist were of normal latency (Erb's point potential, cervical potential and N19 at 8.4, 12 and 17 ms respectively) and amplitude (P25-N30 5.6 uV). Magnetic stimulation of the motor cortex elicited responses of normal latency (right deltoid, biceps, diaphragm and tibialis anterior at 10.4, 11.3, 14.0 and 28 ms respectively, left deltoid at 10.7 ms).

MRI: T2 weighted brain MRI (SE 2000/60) revealed multiple white matter lesions characteristic of MS. T1 weighted MRI of the cervical cord was normal. T2 weighted (SE 1500/80) sagittal MRI of the cervical cord revealed intrinsic high signal lesions on the left side of the cord at C3 and C4 (fig 2a) which were confirmed on axial imaging (fig 2b). There was also a lesion on axial images alone in the right lateral cord at C2.

Discussion

The muscle jerks in the present case lacked both the suppressibility and compulsive nature of tics, and were therefore considered to be myoclonic. Several features argued against the myoclonus being of either cortical or reticular origin.¹⁻³ There were no time-locked cortical correlates in the back-averaged EEG activity preceding spontaneous jerks, and SEPs were of normal amplitude. In addition, the duration of EMG activity in the jerks was longer than that seen in cortical or brainstem reticular reflex myoclonus. Ascending activation of cranial nerve innervated muscles in the generalised jerks, which sometimes occurs in reticular myoclonus, was also absent. Finally, the caudal spread of myoclonic activity was relatively slow: the difference in latency to onset of EMG activity between latissimus dorsi or diaphragm and tibialis anterior was about 32 ms in spontaneous jerks, about 15 ms longer than the difference in latency between these muscles following magnetic stimulation of the motor cortex (fig 1).

Conversely, the myoclonus was largely confined to segments innervated by the cervical cord and each spontaneous jerk was preceded by cervical paresthesiae. Furthermore, the finding of intrinsic cervical cord lesions compatible with MS plaques on MRI suggests that the segmental myoclonus arose from this level.

Figure 1 A spontaneous myoclonic jerk compared with the responses to magnetic stimulation of the motor cortex. A): Rectified EMG record of a single spontaneous jerk in the patient. B): Unrectified EMG record of response to a single magnetic stimulus to the motor cortex. It is clear that the difference in latency to onset of EMG activity between latissimus dorsi (or diaphragm) and rectus abdominis, and between latissimus dorsi (or diaphragm) and tibialis anterior in the spontaneous jerk exceeds that following magnetic stimulation of the motor cortex. Arrows mark the onset of EMG activity in diaphragm, rectus abdominis and tibialis anterior. The response to magnetic stimulation of the motor cortex was not recorded in the trapezius. All muscles shown in A) and B) were right sided. Horizontal calibration line: 25 msec; vertical calibration line: 100 uV in A) and 500 uV in B).