Spinal myoclonus with giant somatosensory evoked potentials and enhanced long-loop reflex: a case report

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Summary

We describe a patient with an ischaemic lesion of the cervical spinal cord who presented with clinical evidence of stimulus-sensitive, multisegmental myoclonic jerks restricted to the truncal and proximal limb muscles and accompanied by electrophysiological features (giant somatosensory evoked potentials and enhanced long-loop reflex) of cortical myoclonus. We hypothesize that these features might result from a loss of inhibitory influences on the sensory input to cortical structures: a concomitant contribution of spinal and cortical hyperexcitability seems to have played a crucial role in inducing myoclonus in our patient.

KEY WORDS: spinal myoclonus, giant SEPs, LLR, ischaemic myelopathy.

Introduction

Myoclonus is a heterogeneous condition of varying aetiology which, applying clinical and electrophysiological (1) criteria, can be classified according to the site of its generation and the nature of its appearance (spontaneous or triggered). Myoclonus may originate from the cerebral cortex, subcortical structures, brainstem, spinal cord and peripheral nerves (1-6). Spinal myoclonus is a rare type of myoclonus that arises from the spinal cord, and shows segmental distribution in one or more spinal nerve-innervated parts of the body. It occurs in two forms, spinal segmental myoclonus (SSM) and propriospinal myoclonus (PSM) (7). SSM is the result of abnormal spontaneous discharges of motor neurons in a limited area of the spinal cord. PSM is a spontaneous or stimulus-sensitive jerking that, through cranio-caudal or caudo-cranial propagation in the spinal cord, via slow-conducting propriospinal pathways (8,9), extends to many segments. Spinal myoclonus has been associated with infectious, structural, degenerative, vascular, neoplastic, and post-traumatic aetiologies (10-14).

We describe a patient with an ischaemic lesion of the cervical spinal cord who presented with clinical evidence of stimulus-sensitive, multisegmental myoclonic jerks originating in the cervical muscle and spreading, showing a characteristic propriospinal pattern, to the proximal limb and abdominal muscles. These clinical signs were accompanied by electrophysiological characteristics (giant somatosensory evoked potentials, SEPs and enhanced long-loop reflex, LLR) of cortical myoclonus.

Case report

Clinical evaluation

A 67-year-old woman was referred to our hospital following the appearance of spontaneous and evoked myoclonic jerks in the upper limbs. Onset of the symptoms had occurred, in a subacute manner, 4 months before her admission. The patient neither smoked nor drank alcohol. She had been suffering from arterial hypertension for 20 years, which was being treated with oral antihypertensive drugs (enalapril 20 mg/day). There was no personal or family history of neurological disease. At admission, the neurological examination was negative, except for the presence of myoclonus. Spontaneous, non-rhythmic myoclonic jerks of varying amplitude were observed in the patient’s limbs, particularly in the proximal muscles. These jerks were easily elicited by tactile stimuli, such as tapping the side of the neck, whereas tendon stretch was less effective. Sudden auditory stimuli were not effective in eliciting the myoclonus.

The patient was unable voluntarily to suppress jerking. Tapping the region on the left of her neck evoked myoclonic jerks of axial and proximal limb muscles innervated by spinal nerves from adjacent segments; the trapezius, deltoid, biceps, and abdominal muscles were particularly affected, whereas there was no jerking of muscles in the extremities. There was no habituation to repeated stimulations. Laboratory investigations consisting of a full blood
Neurophysiological findings

The patient was submitted to neurophysiological investigations that included routine electroencephalogram (EEG), surface electromyogram (EMG) recording of the upper limb muscles, back-averaging of the EEG, SEPs, and LLR. The SEP and EMG findings were also evaluated in 10 healthy subjects matched with the patient for sex and age (10 females; age range 60-70 years).

Routine EEG was recorded from 11 silver/silver chloride electrodes placed on the scalp and referred to the linked earlobes in accordance with the 10-20 International System. The time constant was 0.3 sec. Electromyographic recordings of the myoclonic jerks were performed by placing a pair of disc electrodes, 1 to 3 cm apart, over the trapezius, deltoid, biceps, triceps, flexor carpi ulnaris, opponens pollicis, and abdominal muscles. EEG was carried out concomitantly with the EMG recordings. The technique of jerk-locked back-averaging was used to observe events both before and after the occurrence of the spontaneous and evoked myoclonic jerks. EEG activity was collected from nine scalp electrodes referred to the linked earlobes, again positioned in accordance with the 10-20 International System. For spontaneous jerks, EEG events (50 jerks) before and after the trigger point were averaged. The trigger point corresponded to the moment of EMG discharge in the left trapezius and deltoid muscles.

For evoked jerks, the onset of the stimulus application served as the zero point, and the myoclonus latencies in individual muscles were calculated for each jerk. Reflex myoclonic jerks were elicited by applying a single square electrical pulse on the left side of the neck, at twice the perception threshold (0.5 ms pulse duration). Silver chloride discs in a bipolar montage, 2 cm apart, were used for the EMG recordings. The high and low-pass filter settings of the amplifiers were 20 Hz and 20 kHz, respectively. To ascertain the earliest latency of the EMG discharge for each muscle, a series of eight individual recordings as well as averaged rectified recordings were collected using a 500-ms sweep. Reflex latencies were determined both from individual trials and from the averaged rectified recordings. SEPs were recorded following both right and left median nerve (MN) stimulation at the wrist. Conduction along the peripheral and central somatosensory pathways was evaluated recording from Erb’s point, seventh and second cervical spinous processes, and contralateral parietal (P3 and P4) region. The recording electrodes were again placed according to the conventional 10-20 International System. Electric shocks were delivered to the nerve as square wave pulses of 0.2 ms duration at a rate of 1 Hz. The stimulus strength was adjusted to 50% above the motor threshold, and 500 traces were averaged.

For the other studies, including peripheral motor and sensory nerve conduction velocity (MCV and SCV) and F wave measurements, conventional methods were used.

The long-loop reflex (LLR) was recorded by stimulating the median nerve at the wrist. Muscular responses were recorded from voluntarily contracted thenar muscle, while the median nerve was stimulated using a percutaneous square electrical pulse (duration 1 ms) at near-motor threshold intensities (12-16 mA). Muscular responses were recorded through Ag/AgCl disc electrodes 8 mm in diameter, with a belly-tendon montage. LLRs were labelled according to the Upton nomenclature (16) as V1 (=short latency) and V2 (=long latency). Raw as well as fully rectified EMG signals were simultaneously acquired and 20 traces were averaged in the 200 ms following nerve stimulation. Peak-to-peak amplitude of the V1 and V2 components and the V1/V2 amplitude ratio in the different experimental conditions were measured and compared. LLR latencies were measured at the onset of the V2 component.

Figure 1 - T2-weighted MRI image (TR 2500 - TE 120) shows a linear area of high signal intensity within the spinal cord from the C3 to C5 spinal segments, which is consistent with cervical spinal cord ischaemia probably secondary to a spondylogenic or thromboembolic disease.
Results

Healthy subjects

The mean peak-to-peak amplitude of the N20/P25 for both sides was 3.6±2.2 (range 2.4-5.0). The mean peak-to-peak amplitude values of the LLR for the V1 and V2 components were 0.78±0.3 mV (range 0.4-1.0) and 1.2±0.5 (range 0.5-2.0) mV, respectively.

Patient

Tapping on the left side of the neck elicited muscle jerking which began in the trapezius muscle bilaterally (showing a synchronous pattern) at an average latency of 100 ms. The time difference in EMG activity between the same muscles on the right and left sides was small (on average 1.5 ms in the trapezius and 2.2 in the biceps). The latencies of the jerks from the other muscles increased in proportion to the distance from the trapezius muscle, ranging from 10 to 50 ms (Fig. 2). Intraspinal propagation velocity was calculated from cervical and thoracic level, and it ranged from 8.1 to 11.4 m/sec. Back-averaging of the EEG, in which activation of the left trapezius served as a trigger, did not show either positive or negative spikes. Following left and right median nerve stimulation a typical giant SEP with increased N20-P25 amplitude (33.6 µV and 32.0 µV, respectively) was found over the contralateral parietal region (Fig. 3). The amplitudes of both V1 and V2 LLR components were markedly enhanced, particularly the latter (Fig. 3), when compared with the maximal value found in 10 normal subjects (V2 amplitude: 2.8 mV vs 1.2 mV, respectively).

Discussion

There is strong evidence that our patient’s myoclonic jerks, both spontaneous and elicited, are of spinal origin: a) the neuroimages revealed an ischaemic lesion in the cervical spinal cord; b) the myoclonic jerks were elicited by tactile stimulation to the lateral region of the neck corresponding to the level of the spinal lesion (C3-C5); c) the myoclonus was multisegmental and bilaterally synchronous; d) the temporal relationship between muscles was found to be similar in each jerk, i.e., EMG activity occurred in the trapezius muscles first, then extended to the upper limb muscles in a rostro-caudal manner. Furthermore, the clinical characteristics and the velocity at which the jerking spread were compatible with other cases of spinal myoclonus reported in the literature (10-14). In particular, our case presented some of the clinical characteristics reported in so-called “propriospinal myoclonus”, such as the stimulus-sensitive activity, the multisegmental involvement, and the low-velocity (8.1-11.4 m/sec) cranio-caudal progression (9,12,14). However, in our patient the presence of giant SEPs and an enhanced LLR point to an abnormality in the motor-sensory cortex, suggesting a possible role of the latter in the generation of the myoclonus. Moreover, the latencies between the trigger stimulus and jerking in adjacent muscles were in the range of 90-100 ms, a time compatible with a polysynaptic loop and suggestive of a spinocortical or subcortical pathway. The physiopathological mechanism producing myoclonic jerks in our patient may only be supposed. A lack of functional activity of the inhibitory spinal interneurons has been hypothesized in the generation of spinal myoclonus (13,15). By contrast, it has been demonstrated that giant SEPs and an abnormal LLR, associated with cortical myoclonus, indicate an enhanced response to sensory stimuli from the motor-sensory cortex, which suggests that it is abnormal activation of the motor-sensory cortex that produces spontaneous myoclonic jerks. Thus, the neurophysiological abnormalities observed in our patient seem to reflect both cortical and spinal hyperexcitability. However, it is known that the giant SEP is not necessarily linked to myoclonus, which means

![Image](https://example.com/image1)

**Figure 2** - Electromyographic correlates of myoclonic jerks from the left side of the body recorded simultaneously following an electrical pulse on the right side of the neck. The latencies become longer as the distance between the recording site and the stimulated segment increases.

![Image](https://example.com/image2)

**Figure 3** - **Upper trace.** Marked enhancement of the V2 components in LLR response after median nerve stimulation. **Lower traces.** A typical giant SEP with N20-P25 amplitude in the region of 30.0 µV over the contralateral parietal region after both right and left median nerve stimulation.
that hyperexcitability of the central cortex per se does not always cause myoclonic jerks (5). Moreover, the lack of jerk-related spikes on the contralateral surface EEG does not support the hypothesis of cortical generation of myoclonic jerking. However, this does not exclude a contribution of the cortical reflex loop to the generation of myoclonic jerks in our patient.

It may be hypothesized that giant SEP amplitude and enhanced LLR result from a loss of interneuronal inhibitory influences on the sensory input to cortical structures (17). Vice versa, cortical structures might be responsible for an abnormal modulation of the excitability of the spinal interneurons. Considering that a small proportion of patients with cortical or spinal lesions display a myoclonus syndrome, the concomitant contribution of spinal and cortical hyperexcitability seems to play a crucial role in inducing myoclonic jerks in our patient.

In view of the findings recorded in these studies, further studies may help to clarify the concomitant impairment of the excitability of the sensory-motor cortex and spinal neurons in patients with myoclonus.

References