Axial propriospinal myoclonus misdiagnosed as myoclonic seizures

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Abstract

Propriospinal myoclonus is characterized by brief, repetitive, mainly arrhythmic myoclonic jerks arising from axial muscles and spreading rostrally and caudally to the adjacent myotomes. Propagation is not an obligatory feature and jerks can remain localized to the axial muscles. We present a patient with an axial non-propagated propriospinal myoclonus, misdiagnosed as myoclonic seizures, occurring at rest and at wake-sleep transition phase, and evoked by auditory and somatosensory stimuli. Video-polygraphic recording was useful for a correct differential diagnosis between epileptic and nonepileptic myoclonic events.

KEY WORDS: focal axial myoclonus, myoclonic seizures, propriospinal myoclonus, video-polygraphy.

Introduction

Propriospinal myoclonus (PSM) is a rare movement disorder characterized by spontaneous or, in rare instances, stimulus-sensitive myoclonic jerks, arising from muscles innervated by middle-thoracic myelomes and spreading rostrally and caudally to the other adjacent myotomes with a peculiar slow propagation up and down the spinal cord (propriospinal pattern) (1, 2). PSM can occur in the relaxed wakefulness, during drowsiness preceding sleep, at sleep-wake transition phase and upon awakening in the morning (3, 4). In some cases, jerks remain confined to the abdominal muscles without spreading to the cervical and leg muscles, thus resembling a spinal segmental axial myoclonus (5).

We report a patient with a focal axial propriospinal myoclonus, initially misdiagnosed as myoclonic seizures. We emphasize the role of video-polygraphic recordings for an appropriate diagnosis.

Case history

At the age of 59 years, a right-handed man came to our Epilepsy Centre because of a history of axial jerks, initially misdiagnosed as myoclonic seizures, resistant to levetiracetam (up to 3000 mg/day). Since the age of 35 years, he developed a motor disorder, represented by involuntary repetitive jerks involving the abdominal wall. The jerks occurred in the lying position, at relaxed wakefulness, during the wake-sleep transition phase and recurred every 10-30 seconds. Jerks were also elicited by auditory and somesthetic stimuli. No periods of spontaneous remission were reported. At the hospital admission, the patient underwent multiple sessions of video-polygraphic monitoring during relaxed wakefulness, and focal axial myoclonus was recorded (Video, Fig. 1). EEG showed no abnormalities throughout the recordings; back-averaging of the EEG disclosed no time-locked cortical potentials in the first second preceding and following the spontaneous jerks. On the electromyographic recording, isolated or recurrent bursts of fast activity were recorded in the left external oblique abdominal muscle (T5-T12 innervation) with subsequent propagation to adjacent muscles, i.e. left rectus abdominis (T5-T12 innervation) and, less frequently, to left dorsal muscles (i.e. left latissimun dorsi, C6-C8 innervation). EMG bursts had a variable duration of 100-300 ms. The delay between the first activated muscle (i.e. left external oblique abdominal) and the rostral one (i.e. left rectus abdominis) ranged from 50 to 150 ms. Repetitive myoclonic jerks were also elicited by touching the patient’s left arm (Video, Fig. 2). Long latency reflex and transcranial magnetic stimulation were normal. No abnormalities were found with conventional spinal and brain MRI sequences.
Clonazepam (2 mg/day at bedtime) was ineffective, whereas rotigotine (2 mg/day at bedtime) reduced jerks frequency.

Discussion

Clinical, neurophysiological and neuroimaging features of our patient suggested an idiopathic focal axial myoclonus resembling a PSM. Myoclonic jerks did not spread to the other myotomes and remained localized to the abdominal muscles. We can thus consider it as a focal axial propriospinal myoclonus. Vetrugno at al. (5) reported a patient with both focal axial myoclonus and propriospinal myoclonus. Focal and propagated jerks could be spontaneous or triggered by sensitive stimuli. The Authors concluded that the two forms may coexist and hypothesized that the same spinal generator responsible for the monomeric segmental myoclonus may cause a multimeric propriospinally propagated muscular activation. In our patient there was not the coexistence of both phenomena, and focal axial myoclonus was exclusively recorded. The pathogenic mechanisms involved in the axial PSM have not been conclusively established. Nevertheless, in our patient we hypothesize that a functional spinal reflex activity limited to the thoracic spinal tract was involved. Our case highlights the difficulty to differentiate movement disorders from epileptic seizures and proves the usefulness of prolonged video-polygraphic recordings.

Disclosure

The Authors report no disclosures relevant to the manuscript. All Authors have read and agreed to the content of the manuscript. The paper is not simultaneously under consideration by any other journal and has not been previously published. Any financial or commercial involvement or other conflicts of interest by any Author. Any contribution of industry-sponsored research or of corporate participation in preparing the manuscript. Signed consent forms authorizing publication have been obtained.
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Figure 2 - Video-EEG/polygraphic recording. Repetitive myoclonic jerks elicited by touching the patient’s left arm.

References