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A Case of Hemiabdominal Myoclonus

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Abstract

Myoclonus consists of sudden, brief, involuntary jerky muscular contractions. Central and peripheral nervous system lesions are involved in the pathogenesis of this movement disorder. Symptomatic or secondary spinal myoclonus is the most common form. A 68-year-old woman was diagnosed with hemiabdominal spinal myoclonus. Occasional and very mild involuntary repetitive movements of the hemiabdomen began immediately after surgery for uterine cancer. After surgery for laparocele, secondary to the uterine cancer surgery, performed under spinal anesthesia, there was severe worsening of movements. Neuroradiological investigations failed to demonstrate spinal injury, while neurophysiological studies showed impairment of the right central somatosensory pathway. Considering the low resolution of magnetic resonance imaging in the evaluation of thoracic level, we suggest an extensive neurophysiological evaluation in patients with spinal myoclonus.

Keywords

spinal myoclonus, hemiabdominal myoclonus, video-EEG, electromyography

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Introduction

Myoclonus is a movement disorder with a wide differential diagnosis, but with common semiological elements: brief, shock-like movements caused by muscle contraction, or inhibition of ongoing muscle activity. Myoclonus can be subclassified by site of origin into cortical, subcortical, spinal, and peripheral.[1] Spinal myoclonus can be subdivided spinal segmental and propriospinal myoclonus.[1]

The spinal motor system is organized on 2 levels: the spinal segmental and propriospinal systems. Lesions in each can cause myoclonic syndromes, with substantially different clinical features. Spinal segmental systems may become hyperexcitable, often by viral infections, or due to isolation of anterior horn cells from inhibitory influences in disorders such as syringomyelia, glioma, and spinal ischemia.[2] The result is a myoclonus, involving 1 or 2 contiguous spinal myotomes, that is peculiarly resistant to supraspinal influences, such as voluntary movement or sleep. Conversely, the propriospinal system is a slowly conducting intraspinal pathway, that connects multiple segmental levels. Involvement of this system leads to predominantly axial jerks that, unlike brainstem myoclonus, spare the face and are not provoked by stimuli. Electrophysiologically, propriospinal myoclonus is characterized by long-lasting electromyographic bursts that generally spread relatively slowly rostrally and caudally from the level of spinal hyperexcitability.[2] Diagnosis of propriospinal myoclonus is based on clinical features and polymyography.[1] Despite the hyperexcitability of a specific myelomere (myoclonic generator), conventional MRI of the spinal cord is usually normal.[1]

Spinal myoclonus can be symptomatic or idiopathic. Symptomatic or secondary spinal myoclonus is the most common form, and it occurs after spinal cord lesions, drug use, malignancy, or infection. Essential myoclonus (primary and nonprogressive) affects only a small proportion of patients.

Here, we describe a case of hemiabdominal myoclonus with unusual clinical and neurophysiological features that, in our opinion, support the diagnosis of segmental spinal myoclonus.

Case Report

A 68-year-old woman presented with a 1-month history of spontaneously developed involuntary repetitive movement of the hemiabdominal right muscles.

The movements began in November 2009, immediately after surgery for uterine cancer, but were occasional and very mild. The surgery was performed after general intravenous

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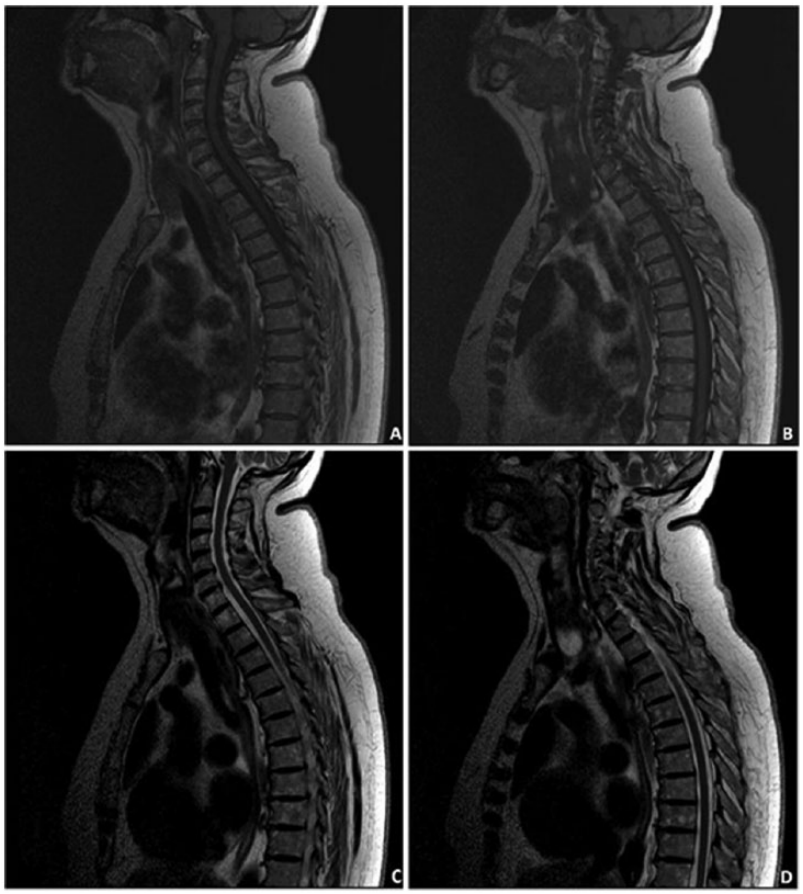

Figure 1. Spinal cord magnetic resonance imaging. Sagittal cervical (A) and dorsal (B) spinal cord—T1-weighted images and sagittal cervical (C) and dorsal (D) spinal cord—T2-weighted images are normal.

anesthesia, which was not well tolerated by the patient, who had marked side effects (dyspnea, nausea, and vomiting). In November 2010, after plastic surgery, of a laparocele secondary to the previous uterine cancer surgery, performed by spinal anesthesia, she developed severe worsening of involuntary repetitive movements of the right abdominal muscles. In fact, these movements became continuous, increasing in the supine position and presleep. Her husband stated that the involuntary movements persisted during sleep. After surgery the patient did not undergo chemo-, radio- or antibiotic therapy.

No family history of myoclonus or epilepsy was identified. No drugs known to induce myoclonus[3] were consumed by the patient, and there was no history of spinal cord trauma, infections or psychological disorders.

On neurological examination, asymmetrical myoclonus of the abdomen was observed with a frequency of approximately 0.2 to 0.3 Hz. The myoclonus was brief, periodic, and continuous, and it also occurred during sleep. It was enhanced by supine position, but not by cutaneous sensory stimuli; it was not preceded by a premonitory sensation and it was suppressed by volition for few seconds, soon followed by a rebound myoclonus. The characteristics and frequency of myoclonus were unaffected by distracting maneuvers. No other neurological signs were found.

All laboratory investigations were normal, including blood tests cell counts, sedimentation rate, C-reactive protein, fibrinogen, hepatic enzymes, free triiodothyronine and free thyroxine, thyroid-stimulating hormone, iron studies, and electrolytes; HIV, human T-cell lymphotrophic virus, Lyme, syphilis, and herpes viridae serology; antinuclear, anti-DNA, anti–extractable nuclear antigens, antigliadin, anti-endomysium, anti-thyroid, and anti-phospholipid antibodies; serum levels of B12, folic acid, copper, ceruloplasmin, homocysteine, and angiotensin-converting enzyme, and tumoral markers. Also anti-neural autoantibodies (anti-Hu, -Yo, -Ri, -CV2, -Amphyphisin, -GAD65, -$NMDA_2$, -Lgi-1, -Caspr-2, -GABAb, -AMPA) were negative. The patient refused lumbar puncture.

Brain (data not shown) and spinal cord MRI was normal (Figure 1). A total-body computed tomography scan, performed to screen for occult malignancies, was normal.

Nerve conduction studies of lower limbs, and electromyographic (EMG) examination of rectus femori, tibialis anterior, extensor longus hallucis and gastrocnemius bilaterally were

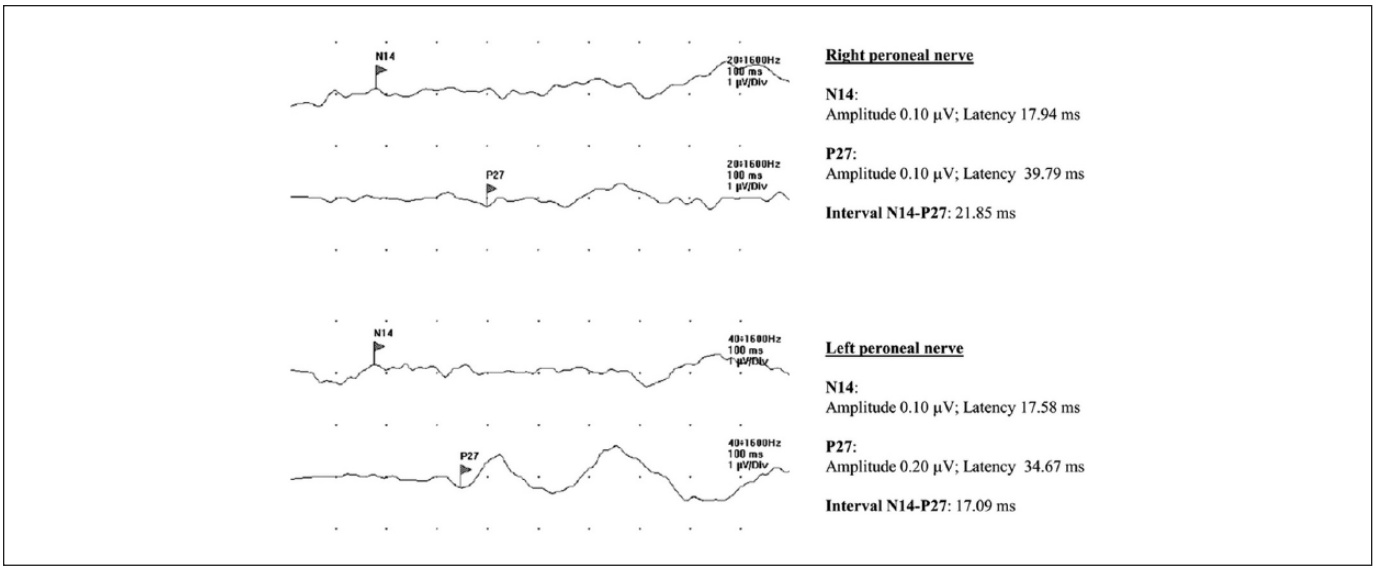

Figure 2. Somatosensory-evoked potentials (SEPs) obtained after peroneal nerve stimulation.
For peroneal nerve SEP studies, a 2-channel recording was used. Channel 1 recorded the lumbar potential (N14) with the active electrode positioned between the T12 and L1 spinous process, in the midline, and the reference electrode over the iliac crest. Channel 2 recorded the scalp potential (P27) with the active electrode positioned at Cz and the reference electrode at Fz. Values of amplitude and latencies of both waves and of the N14-P27 interval are show on the right side.

unremarkable. EMG of right rectus abdomini confirmed the presence of bursts of motor unit potentials with a frequency of 0.2 to 0.3 Hz and a duration between 500 and 800 ms; conversely, EMG of left rectus abdomen did not disclose any muscular activity. The patient refused needle EMG of paravertebral muscles.

Somatosensory-evoked potential from peroneal nerve bilaterally revealed a prolonged somato-sensory conduction time from the right lower limb (Fig. 2) in comparison to the contralateral one. Motor evoked potential recorded from all four limbs muscles were unremarkable.

Video-EEG–EMG polygraphic recording was performed, with 2 couples of surface electrodes over the abdominal muscles bilaterally, and with a needle electrode in the right rectus abdominis, and confirmed myoclonic periodic activity in the right abdominal muscles (see video at http://eeg.sagepub.com/supplemental). Distracting maneuvers did not affect the polymyographic findings, such as frequency, burst duration, or the velocity of propagation. Back averaging study did not reveal any cortical focus of myoclonus; a bereitschaft potential that preceded the movements was absent (see video). Since all the tests were normal, we made a diagnosis of spinal segmental myoclonus. Pramipexole, prolonged release 0.52 mg/d, was started with partial benefit on the myoclonus, followed by add-on therapy, of clonazepam 0.5 mg 3 times a day, with marked improvement of the movement disorder.

Discussion

In this case, the clinical and electrophysiological features of the abnormal abdominal movement were consistent with a diagnosis of myoclonus and, in particular, of spinal myoclonus. The peculiarity of this case is the exclusive involvement of a hemiabdomen.

Our patient stated that the onset of the abnormal myoclonus was after surgery for uterine cancer. Suspecting a surgery-related central nervous system injury, we performed brain and spine contrast-enhanced MRI that were unremarkable. Moreover, a paraneoplastic etiology was excluded because both antineural antibodies and total-body contrast-enhanced computed tomography results were negative.

Normal results of spinal cord imaging ruled out direct spinal cord traumatic injury during the lumbar puncture. We cannot exclude that the myoclonus could be a side effect of the anesthetic drugs[1] even though the myoclonus gradually improved.[4] However, interestingly, somatosensory-evoked potentials revealed a prolonged somatosensory conduction time, after right lower limb stimulation, suggesting a functional impairment of spinal cord sensory pathways.

Several cases of abdominal myoclonus have been described.[5-8] In all, myoclonus was related to a spinal generator.[5-8] In some cases, a spinal cord cause has been identified,[7] while in the majority the etiology remained unknown.[5] Generally a diffuse and symmetric involvement of abdominal muscles has been reported.[5] Only in 1 case, myoclonic activity was confined to right abdominal side.[6]

In our case, the absence of bereitschaft potential suggests an organic disorder. Considering that the myoclonus was rhythmic, persistent during sleep, symptomatic, and with long-lasting EMG bursts, a diagnosis of spinal segmental myoclonus is the most likely. Furthermore, the absence of a spinal generator recruiting axial muscles up and down the spinal cord, via long propriospinal pathways, seems to exclude a propriospinal myoclonus. Considering the muscles involved we can

hypothesize a thoracic spinal generator; unfortunately the patient refused EMG of paraspinal muscles and so we cannot define the metameric level in more detail.

In conclusion, we report a further case of right hemiabdominal myoclonus, in which neuroradiological investigations failed to demonstrate a spinal injury, while neurophysiological studies showed an impairment of right-side spinal somatosensory pathway. Based on the clinical history characterized by sudden onset of myoclonus after surgery, we hypothesize that the patient may have suffered a minimal spinal injury, probably of ischemic nature, unrecognized by MRI imaging, that was documented by neurophysiology. Considering the low resolution of conventional MRI in the evaluation of thoracic level,[1] we strongly suggest an extensive neurophysiological evaluation in patients with spinal myoclonus.

Declaration of Conflicting Interests

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