CASE REPORT

A Rare Case Report: Abdominal Muscle Myoclonic Jerks

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ABSTRACT

Propriospinal myoclonus (PSM) is a rare disorder that is characterized by hyperkinetic movements of flexion or extension in the axial muscles. It can be idiopathic, secondary, or psychogenic. The psychogenic form, or functional movement disorder, has been increasingly recognized in patients previously diagnosed with idiopathic spinal myoclonus. This case report introduces a patient who presented with isolated abdominal muscle contractions that occur usually when a patient lies down, is about to fall asleep, or has just woken up. We discuss the possible differential diagnosis for this rare presentation as well as current knowledge regarding PSM.

Key words: Myoclonic jerk, truncal myoclonus, propriospinal myoclonus, functional movement disorder

INTRODUCTION

Myoclonus is produced by muscular contraction, characterized by brief, shock-like movements. Patients use descriptions such as "jerks," "shakes," or "spasms" for myoclonus. There are multiple ways to classify myoclonus, including by clinical presentation, anatomical location of signal origin, neurophysiology testing, and etiology. The four main etiologies of myoclonus are physiologic, essential, epileptic, and secondary. Physiologic myoclonus is a normal phenomenon in healthy people with minimum association with disability or disease. In essential myoclonus, the myoclonus is the most prominent or sole clinical finding where patient usually experiences mild disability. Epileptic myoclonus occurs in the setting of epilepsy. Finally, symptomatic or secondary myoclonus, the most common type, occurs secondary to disorder that can be neurologic or non-neurologic.

CASE REPORT

A patient is a 59-year-old right-handed male with hypertension, hyperlipidemia, and type 2 diabetes who presented to clinic with myoclonic jerks. 3 weeks ago, he began experiencing sudden jerks in the body, usually when he lies down, about to fall asleep, or has just woken up to initiate the first movements of the day; however, it does not occur every single time with the activities described above. The jerks had woken him up from sleep twice since onset but had not decreased his sleep quality as he does not feel tired throughout the day. He described the jerks to be a sudden severe contraction of his abdominal muscles, making him flex at the waist symmetrically without leaning toward one side or the other. The jerks are usually singular per episode, lasting 1–2 s, occur around 1–2 times a day, independently without pain, tingling, numbness, weakness, loss of consciousness, or seizure-like activity (rhythmic, repetitive jerking movements). The patient felt that the intensity and location of the jerks were consistent every time. The jerks occurred without warning signs or aura. The jerks had not increased in frequency or intensity since onset. Although no episodes had occurred in the public thus far, the patient was concerned they will. No associated symptoms such as headache, nausea, neck pain, back pain, unintentional weight loss, insomnia, changes in cognition, memory loss, mood, personality changes, or changes in sleep were reported. The

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patient denied personal or family history of seizure, cancer, neuromuscular disease, movement disorder, or dementia. There was no history of measles, recreational/IV drug use, recent illness, trauma to head or spine, change in medication, stressful events, and recent travels.

Physical examination was grossly unremarkable. His vital signs were within normal limits, and cardiovascular, pulmonary, and musculoskeletal examinations were unremarkable. No myoclonic jerk was elicited when the patient laid to a supine position. A full neurologic examination was performed, which the patient was alert and oriented with no deficit in speech and language, cranial nerves were intact, and motor function, reflex responses, sensory function, and coordination were all within normal range. Due to limitations of the clinic, no laboratories or imaging was performed at the time of assessment.

Assessment

A 59-year-old right-handed male with hypertension, hyperlipidemia, and type 2 diabetes presented with 3 weeks history of painless, non-progressive, and symmetric truncal jerks occurring occasionally with supine position, before sleep and upon awakening. He had been woken up twice by the jerks. No associated symptoms and no inciting events had been identified. Physical examination was unremarkable, and no jerks were elicited with supine position during visit. Propriospinal myoclonus (PSM) is suggested by the acute onset, isolated truncal myoclonic jerk that occurs with supine position, sleep onset and upon awakening. Other differential diagnoses include but are not limited to functional movement disorder (FMD), essential myoclonus, primary myoclonus of aging, myoclonic seizure, sleep myoclonus, paraneoplastic syndrome, and Creutzfeldt-Jacob disease.

Differential Diagnosis

PSM (see detailed discussion in next section)

Patient's acute onset of isolated truncal myoclonic jerk that is associated with supine position, sleep onset and awakening without any other symptoms is consistent with PSM. Myoclonic jerk is consistent in intensity and duration and always occurs in the abdominal region without involving any other muscle groups. Jerks can also occur during sleep. Patient's absence of loss of consciousness, change in memory, mood or personality, rhythmic contractions, recent illness, or trauma suggests PSM. However, 28% of the cases in a study noted that the jerks are sensitive to positional stimulation in which, in this patient, the myoclonus was not always elicited when he lies supine or with sleep onset.[1]

FMD

As seen in the patient, the myoclonus is characterized by acute onset with inconsistent frequency and response to positional stimulation. However, FMD usually involves the neck and face, is variable in jerk characteristic (intensity, duration, and distribution), has rapid progression with spontaneous remissions, does not occur during sleep, and has multiple somatization and history of emotional or psychiatric disturbances. Another characteristic of FMD is a reduction with distractibility which we were not able to test since the jerk was not elicited during the visit.[2]

Sporadic essential myoclonus

Patient's abrupt onset, isolated myoclonus without progression and lack of family history for movement disorder is consistent with sporadic essential myoclonus. It is a heterogeneous entity regarding distribution, exacerbating factors, and findings on neurological examination; however, there are some salient features of essential myoclonus such as oscillatory (back-and-forth movement along a joint axis), segmental (rhythmic or semi-rhythmic contractions of muscle groups supplied by contiguous segments of brainstem or spinal cord), and multifocal (involving more than one muscle groups, usually involving proximal limb muscles) myoclonus which were absent in our patient.[3]

Primary myoclonus of aging

Primary myoclonus of aging is consistent with patient's acute onset myoclonus without dementia; however, patients are usually older than 65 years old, and jerks are asymmetric that occur at different muscle groups, not usually associated with position or sleep onset.

Myoclonic seizure

Myoclonic seizure can present as acute onset of isolated jerk that occur during sleep onset without loss of consciousness; but it is usually associated with epilepsy syndrome diagnosed early in life.

Sleep myoclonus

It is also known as hypnic jerk that occurs during sleep or sleep onset without any other symptoms or progression. However, it is rare in adults, usually involves smaller muscles (fingers, toes, facial muscles), and does not occur when the patient is awake.

Paraneoplastic syndrome

Paraneoplastic syndrome is consistent with patient's acute onset myoclonic jerk; however, it usually manifests with opsoclonus and symptoms suggesting malignancy such as weight loss, chills, night sweats, or cough (most commonly associated with small cell lung cancer).

Creutzfeldt-Jakob disease

Creutzfeldt-Jakob disease is consistent with patient's acute onset myoclonic jerk; however, there were no changes in orientation, memory, personality, mood, or startle myoclonus.

DISCUSSION

PSM is a rare disorder that is characterized by hyperkinetic movements that consist of flexion or extension in the axial muscles (including the abdomen, trunk, hip, or neck region) with the frequency of 1–6 Hz. It has been hypothesized to arise from a spinal generator that transmits activity rostrally or caudally in the spinal cord through the long propriospinal pathways.[4] Mean age of onset has been recorded to be 43 years old, in which 55% of the studied subjects were male. The myoclonic events are acute on onset usually painless and arrhythmic. They have been reported to occur at the time when the patient is attempting to fall asleep, drowsy, or waking up. Lying down can exacerbate movements.[1]

PSM can be idiopathic or secondary in which the latter was suggested to be linked to spinal cord pathology. Cases of PSM resulting from spinal cord pathology include but are not limited to ischemic myelopathy, cervical tumor, neuromyelitis optica, syringomyelia, disc herniation, and back trauma.[5] Other PSM cases were reported in isolated cases of Herpes zoster, Lyme infection, hepatitis C, gluten sensitivity, Fragile X permutation, myasthenia gravis, breast cancer, B12 deficiency, and cannabis abuse.[1] More recently, psychogenic forms of PSM or FMD are increasingly recognized. A large cohort study showed at least 30% of patients with previously diagnosed idiopathic spinal myoclonus which had Bereitschaftspotential (BP), indicating that the etiology was psychogenic.[6] Besides the presence of BP on electromyography (EMG), FMD can be differentiated from idiopathic PSM by previous somatizations, spontaneous remissions, distractibility and suppressibility of symptoms, involvement of facial movements, or vocalizations.[1]

Pathologic or Imaging Discussion

A detailed history elicited by the patient as well as a polymyography is crucial for making a diagnosis. The most common polymyography finding in Van der Salm *et al.*'s study is a slow conduction velocity of 1–15 m/s.[1] The slow conduction velocity suggested the involvement of the propriospinal tract which was reported in 51% of the published cases. However, this typical electrophysiological pattern for PSM can also be found in patients with psychogenic axial jerks. Therefore, the absence of BP in an electroencephalogram (EEG)-EMG, which measures activity in the motor cortex leading up to voluntary muscle movement, can aid in distinguishing idiopathic PSM from a psychogenic cause. It should be noted that limited institutions have the capability to conduct simultaneous EEG-EMG recordings.[7]

Imaging with computed tomography (CT) or magnetic resonance imaging (MRI) is essential to exclude spinal cord lesions or myelopathy, resulting in axial jerks. It is, especially, important when BP is absent or not recordable to avoid misdiagnosis, which can be due to high jerk frequency or too much movement artifact. Red flags for PSM that would suggest underlying structural lesion include concurrent optic neuritis, urinary urgency, ataxia, abnormal reflexes, and thoracic sensory changes. These signs would warrant investigation for possible myelopathy. One study looked into 10 idiopathic PSM cases in which MRI-diffusion tensor imaging (DTI) was performed - 3 of the 10 cases revealed microstructural abnormalities that did not match the presumed spinal level determined by the onset of jerks in the EMG.[8] The study suggested that spinal cord microabnormalities detected on DTI could be consequential rather than causative results of the jerks.

Management

For patients with secondary PSM, treating the underlying etiology most often will resolve the myoclonus. Clonazepam is considered first-line therapy for PSM patients with spinal myoclonus. It is, especially, helpful when taken before sleep to prevent myoclonus at sleep onset. A study shows that levetiracetam is synergistic when used in combination with clonazepam in reducing myoclonic events.[9] Clonazepam is also helpful in treating patients with essential myoclonus.[2] Other medications with sporadic effectiveness include SSRI and valproate. Carbamazepine, prednisone, and baclofen were reported as ineffective. In selected articles, successes with unconventional therapies such as autogenic biofeedback training and transcutaneous electrical nerve stimulation have been reported as well.[10] Finally, in FMDs, botulinum toxin, electrical nerve stimulation therapy, and cognitive behavioral therapy have been effective.[11]

In our patient with isolated truncal myoclonic jerk, we ordered complete blood count, comprehensive metabolic panel, blood toxicology screen, liver function tests, renal function tests, thyroid function test, CT-head/spine/chest, and EEG with and without EMG to narrow diagnosis. Trial with clonazepam 1 mg PO, pelvic inflammatory disease will be initiated for symptomatic therapy.

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