"Pure Motor Restless Legs Syndrome" Associated with Spinal Pathology Mimicking Myoclonus

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ABSTRACT

Objective: To codify a clinical entity of leg "myoclonus" that responds to dopamine agonists
Design: Case reports and discussion
Patients: We report two cases of elderly patients with spinal pathology who were referred for myoclonus. Both had semi-rhythmic leg movements that partially improved while standing, but denied any urge to move. These movements improved dramatically with pramipexole, a dopamine agonists used for RLS.
Conclusions: We propose that this "myoclonus" is actually the isolated motor component of the same pathology that causes restless legs syndrome.

INTRODUCTION

Restless legs syndrome (RLS) is clinically defined by the presence of four clinical features: 1. an urge to move the limbs with or without sensations, 2. worsening at rest, 3. improvement with activity, 4. worsening in the evening or night. [1] Periodic limb movements of sleep (formerly called nocturnal myoclonus) [2] accompany RLS in most cases. [3] Wakeful movements, variable called dyskinesia while awake or periodic limb movements while awake, are also seen in some cases. [4] These movements are often considered the "motor" component of RLS whereas the urge to move is the "sensory" component. That said, no criteria for partial RLS or probable RLS in subjects partially meeting the four cardinal features for RLS exist, nor has a syndrome of an isolated motor component of RLS without the required sensory component been formally described.

In contrast, myoclonus is a heterogenous group of conditions, basically defined as brief (< 0.25 second) jerks. Various classifications have developed predicated on the anatomy of the movements, the presumed anatomy of physiological origin, and electrophysiologic criteria. However, these are still largely descriptive. We report two cases which we feel represent isolated motor component of RLS, misdiagnosed as myoclonus, in the setting of spinal cord pathology.

METHODS

CASE #1

This is an 80-year-old right-handed male who presented to the Baylor College of Medicine Movement Disorders Clinic with leg jerking and a diagnosis of myoclonus. The patient reports that the leg jerking began indolently approximately four years prior to our evaluation; however, became quite severe four months prior to evaluation. It is worse when he is seated or reclined and partially, though not completely, relieved while standing or walking. The patient has these constantly, every few seconds, and they significantly impair his ability to sleep, sit comfortably, and even ambulate. He absolutely denied any sort of urge to move or premonitory sensation prior to the movements.

The patient had a long history of both cervical and lumbar spinal disease and is status post two cervical and two lumbar surgeries. He continues to suffer from debilitating lumbar and cervical pain, and is also diagnosed with an axonal neuropathy with neuropathic feet pain. He does not correlate the movements with any pain. For the movements, he has tried several medicines; most recently, clonazepam, which helped modestly. He also takes alprazolam, gabapentin, Duragesic patch, hydrocodone, atomoxetine, thyroid replacement, aspirin, omeprazole, and multivitamins. The patient recently had a spinal cord stimulator placed and, a year before that, an implantable pain pump with morphine and clonidine. Neither of these interventions altered the involuntary movements but did modestly improve pain. He has some urinary urgency but also is diagnosed with benign prostatic hypertrophy, and denies any bowel abnormalities. His other medical complaint was that his eye lids close spontaneously which his family felt was an inability to maintain wakefulness.

On examination, he was in some discomfort from lower back pain. His mental status was essentially normal. Cranial nerves were normal except for a tendency for the upper lids to fall, with a slight quiver in the pre-tarsal area of the orbicularis oculi, consistent with apraxia of eyelid opening. Sensation was decreased to light touch and vibration in the legs. Reflexes were absent in the legs and normal in the arms. His gait was both antalgic and mildly wide-based with arm adduction. He was unable to tandem gait but there were no appendicular cerebellar signs. Romberg testing was normal. The involuntary movements were an almost constant, semi-rhythmic, quick, predominantly leg abduction movements at the hip, which were generally symmetric. [Video segment 1] These did lessen when standing and stopped while walking.

Despite the fact that the patient denied any sort of urge to move, we felt that the movements were consistent with the motor features of restless legs syndrome/periodic limb movements. The patient was put on pramipexole 0.25 mg q.h.s. and titrated up to 0.5 mg three times a day. He reported almost complete resolution of the leg jerking, and moderate improvement in pain. There were no adverse events. The patient subsequently tapered off all benzodiazepines and reduced narcotics by 50%.

CASE #2

The patient is an 86-year-old right-handed Caucasian female who presented to the Movement Disorders Clinic with leg jerking, diagnosed as myoclonus. The exact scenario of events is not entirely clear; however, approximately ten years ago, she went to the emergency room with acute thoracic pain. At that time, it was noted that she had spinal compression with ruptured discs at T8 and T10. Per the patient, she subsequently was not able to ambulate at all and was placed in an extended rehabilitation unit. Over the next six months, she gradually regained the ability to ambulate secondary to improvement in strength. Per medical records at that time, she was never completely unable to ambulate, and she had more of an antalgic gait than gait abnormalities due to true weakness. Nonetheless, over the subsequent one to two years, her subjective weakness and pain improved to the point of being essentially normal.

It was one to two years after this incident that she began to have the insidious onset of jerkiness in her legs. These gradually worsened and in the past three to four years have been quite severe, occurring hundreds of times throughout the day with some nocturnal intensification. They improve while standing or walking but are still subjectively present and now interfere with her gait. She specifically denies any urge to move or sensory component, and is unaware of any rhythmicity. Currently she denies back pain or any bowel or bladder difficulty.

The patient had an extensive workup including 24-hour EEG monitoring. Per her and available medical records, she has tried more than 20 medications for the movements. She specifically recalls failing clonazepam, alprazolam, and valproic acid with a presumed diagnosis of "spinal myoclonus".

On physical examination, the patient had mild cognitive impairment, with a mini- mental status of 24/30. Her cranial nerve examination was normal. Strength testing was 5/5 throughout, with normal bulk and tone. Sensation showed mild decreased vibration sense in both feet but was normal to other modalities. Romberg testing was negative. She did have some stooped posture and a slow modestly wide based gait. She was unable to tandem walk. Reflexes were zero in the ankles and 1/4 elsewhere with downgoing toes. The involuntary movement was predominantly adduction / adduction of the legs at the hips, which occurred at a frequency of approximately 1 Hz during the examination period. [Video segment 2] She also had hip flexion associated with truncal flexion interspersed with the adduction / abduction. This would largely dissipate when standing.

Laboratory evaluation demonstrated that she had a low iron-binding percent saturation of 11%, although ferritin level was normal at 123 ng/mL. Other electrolytes were within normal limits.

The patient was placed on pramipexole which was titrated up to 0.5 mg three times a day. She reported complete cessation of the movements without adverse events. Her sleep was much better, and mood was subsequently improved.

DISCUSSION

We propose that these two similar cases (involuntary symmetric leg movements without urge, gradually developing in the setting of spinal pathology) most likely represent the isolated motor component of RLS. The pathophysiology of RLS is not understood but descending spinal dopaminergic tracts have been postulated to be involved in both RLS and periodic limb movements (often considered to be the motor component of the syndrome). [5, 6] Spinal cord injuries have also been reported to be associated with RLS and especially periodic limb movements of sleep. [7-11] However, it should be emphasized that these cases do not meet criteria for RLS, and both were diagnosed as myoclonus. There is no way to physiologically or pathologically prove a diagnosis of pure motor RLS but the dramatic response to pramipexole is very suggestive. If one insists on calling this phenomenology myoclonus, then it should be emphasized that a dopamine agonist completely controlled the movements when numerous medicines typically tried for myoclonus did not help.

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