

Case Report

Movements Mimicking Myoclonus Associated with Spinal Cord Pathology: Is this a “Pure Motor Restless Legs Syndrome”?

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Abstract

Background: The neuroanatomic substrate of restless legs syndrome (RLS) is poorly understood, and the diagnosis is clinically made based upon subjective sensory symptoms, although a motor component is usually present.

Case Report: We report two cases of elderly patients with spinal pathology who were referred by neurologists 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 agonist used for RLS.

Discussion: We propose that this “myoclonus” is actually the isolated stereotypic motor component of RLS.

Keywords: Restless legs syndrome, myoclonus, dopamine agonists, periodic limb movements, dyskinesia while awake

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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 during sleep (formerly called nocturnal myoclonus)² accompany RLS in most cases.³ Wakeful movements, variably 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 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.

Myoclonus is a heterogeneous group of conditions, basically defined as brief (<0.25-second) jerks. Classifications have developed that are predicated on the anatomy of the movements, the rhythmicity, the presumed neuroanatomy of physiological origin, and electrophysiologic criteria. We report two cases that may represent the isolated motor

component of RLS, misdiagnosed as myoclonus by referring neurologists, in the setting of spinal cord pathology.

Case report

Case #1

This 80-year-old right-handed male presented with leg jerking and a diagnosis of myoclonus. He reports that the leg jerking began indolently approximately 4 years prior to our evaluation; however, it became quite severe within the past 4 months. It worsens when he is seated or reclined and partially improves while standing or walking. The patient has these constantly, every few seconds, and they significantly impair his ability to sleep, sit comfortably, and ambulate. He absolutely denied any sort of urge to move or premonitory sensation. There is no family history of RLS.

The patient had a long history of both cervical and lumbar spinal disease and previously has undergone two cervical and two lumbar surgeries. He continues to suffer from debilitating lumbar and cervical pain but does not correlate the movements with any pain. He has tried several medicines; most recently, clonazepam 2 mg, which helped

Case 1: Before Treatment**Video Segment 1A**

Video Segment 1A. The patient shows semi-rhythmic abduction/adduction movements at the hip that improve upon standing.

slightly. The patient had a spinal cord stimulator and an implantable pain pump with morphine and clonidine. Neither of these altered the involuntary movements but did modestly improve pain.

Examination was normal except for decreased leg sensation to light touch, and vibration in the feet most consistent with either a neuropathy or L5/S1 radiculopathy, and absent leg reflexes. His gait was both antalgic and mildly wide-based. The involuntary movements were an almost constant, semi-rhythmic, quick, predominantly leg

abduction movements at the hip, which were generally symmetric (Video Segment 1A). These did lessen when standing and stopped while walking.

The patient was placed 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 subjectively and on subsequent examination, and moderate improvement in pain. His examination was otherwise unchanged. (Video Segment 1B). There were no adverse events. The patient subsequently tapered off all benzodiazepines and reduced oral narcotics by 50%. He has successfully maintained therapy with >95% improvement in movements but some return of the chronic low back pain for more than 24 months.

Case #2

This 86-year-old right-handed Caucasian female presented with leg jerking, diagnosed as myoclonus. Approximately 10 years ago, she had experienced acute thoracic pain diagnosed as spinal compression with ruptured discs at T8 and T10. Gait was markedly impaired secondary to pain and weakness but she gradually regained the ability to ambulate. One to 2 years after this incident she began having the insidious onset of jerkiness in her legs. These gradually worsened and in the past 3 years has been quite severe, occurring hundreds of times throughout the day with some nocturnal intensification. They partially improve while standing or walking but are still subjectively present and interfere with her gait. She specifically denies any urge to move or sensory component, and is unaware of any rhythmicity. There is no family history of RLS. The patient had an extensive workup and has tried more than 20 medications for the movements.

She had mild cognitive impairment (mini-mental status examination (MMSE) 24/30). Sensation showed mild decreased vibration sense in

Case 1: After Treatment**Video Segment 1B**

Video Segment 1B. The patient from Segment 1 at initial follow-up. Movements are markedly diminished in amplitude and intensity. Subsequent examinations showed no movements.

Case 2**Video Segment 2**

Video Segment 2. The patient shows leg abduction/adduction movements and hip flexion movements that improve while standing. Visualization of the movements in this video is brief but identical movements were seen constantly throughout the evaluation prior to treatment.

both feet. She did have some stooped posture, slow, modestly wide-based gait, and was unable to tandem walk. Reflexes were 0 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 (Video Segment 2). She also had hip flexion associated with truncal flexion interspersed with the adduction/abduction. This would largely dissipate when standing.

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. Post-treatment examination showed no movements. Her sleep was much better, and mood was subsequently improved. She has continued good response for more than a year.

Discussion

These two similar cases (large amplitude, involuntary symmetric leg movements without urge, gradually developing in the setting of spinal pathology) could represent the isolated physiologic 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 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 However, it should be emphasized that these cases do not meet criteria for RLS, and both were diagnosed as myoclonus by other neurologists. “Dyskinesia while awake” is an older term in the RLS literature used to describe wakeful stereotype, usually ankle dorsi/plantar flexion, is also seen in typical RLS but is distinct from this. There is no way to physiologically or pathologically prove a diagnosis of pure motor RLS but the dramatic response to pramipexole is very suggestive. Periodic limb movements (motor component) respond more dramatically to dopaminergics than to

other treatments used for RLS. Myoclonus is of a shorter duration and usually less complex than these movements. 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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