Corpalgia Pseudoathetosica: Another Face for an Old Syndrome?

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Abstract

Background:
Restless legs syndrome (RLS) is a chronic common condition, often debilitating. Variants of RLS include restless arms and restless genital syndrome. Restless legs may occur in patients with chronic pain.

Aims of the Study:
To report three female patients with generalized involuntary movements superimposed on restless legs in the background of generalized pain

Methods:
Repeated clinical examinations and ancillary testing

Results:
Three female patients are described. The first patient had Charcot Marie Tooth polyneuropathy, painful legs –moving toes syndrome, RLS, dysautonomia, neurogenic bladder, chronic generalized pain and choreoathetosis of late onset. The second patient had severe fibromyalgia, chronic tension headache, temporomandibular joint syndrome and irritable bowel syndrome in addition to athetoid irregular multifocal movements. The third patient had involuntary athetoid movements of the limbs, familial RLS, fibromyalgia and attention deficit disorder-inattentive type.

Conclusion:
Corpalgia pseudoathetosica-CPA- (pseudoathetosis associated with generalized chronic body pain) represents an expanded clinical version of restless legs syndrome in patients with underlying chronic pain that needs to be distinguished from typical akathisia. Movements may constitute a self-therapeutic or compensatory physiologic pre-conscious response to pain, in virtue of the known analgesic properties of motor cortex stimulation.

Introduction

Restless legs syndrome (RLS) is a common neurological disorder affecting up to 24% of the population in a primary care setting(1). Women are mostly affected. A familial form is recognized as well as a variant associated with peripheral neuropathy, i.e., CMT disease(2). The typical syndrome manifests as evening bilateral or unilateral leg restlessness while at rest that may persist into the night delaying sleep onset. A tonic-dystonic painful variant is exceptionally found in clinical practice, resembling stiff limb syndrome(3). Restless arms may occur in patients with RLS and rarely in isolation(4). Overlapping syndromes are restless genital syndrome in patients with continuous erotic hyper-arousal not relieved by orgasm, a very distressing occurrence, “painful legs and moving toes syndrome” and its variant “painful arms and moving fingers”(5,6,7). The cause of RLS is still a matter of debate. Peripheral hypoxia seems to be a significant element favoring its appearance(8). If RLS antecedes Parkinson’s disease (PD) or not it is not yet established, however, patients with PD may suffer from troublesome RLS that can be ameliorated by deep brain stimulation (DBS)(9). Pain is frequently reported by patients with RLS, in particular in the tonic-dystonic form. In fact, the typical patient describes the entire experience of restless legs as unpleasant, and pain of articular origin, as in rheumatoid arthritis for example, may trigger restless legs in up to 30 % of the cases(10). Furthermore, it has been established that RLS associates with fibromyalgia (FMA) and irritable bowel syndrome (IBS), both chronic painful conditions(11, 12).

Case Report

Case 1:
A 78 year old female was seen in consultation because of painful hand cramps induced by movement, chronic numbness of the legs and bilateral foot drop due to Charcot Marie Tooth disease type II. She reported muscle twitching and tremors at night, intractable back and leg pain, difficulty walking and periodic unilateral facial erythema in response to warm weather. Previous history was extensive including osteoarthritis, hypertension and macular degeneration. She had childhood Sydenham’s chorea and Tourette-like syndrome during adolescence. Subsequently she had no involuntary movements until very late in life. She also had severe lumbar spinal stenosis, neurogenic bladder, chronic generalized neuropathic pain, nocturnal myoclonus and periodic Harlequin syndrome on basis of neuropathy related...
Case 2:
A 62 year old female was referred originally to the neurologist because of chronic tension-type headache. She has history of chronic severe muscle pain diagnosed as fibromyalgia. She had hypothyroidism, osteoarthritis, temporomandibular joint dysfunction, chronic depression, chronic obstructive lung disease, and irritable bowel syndrome. She had remote history of closed head injury. She was medicated with citalopram, levothyroxine, albuterol, gabapentin and hydrocodone. She had no family history of neurological disorders. She took pregabalin along with gabapentin, tizanidine, celecoxib and tolterodine tartrate. On neurological examination she exhibited irregular athetoid movements of the limbs of variable duration and persistence over time. She had pin point pupils, areflexia, foot dorsiflexion weakness bilaterally, ataxia of gait, leg dysmetria on heel to shin testing and diminished pin prick and touch perception on the legs. She had no position sense at the toes and had purple discoloration of the feet. She had irregular respiration with labored breathing, abnormal gait, leg dysmetria, areflexia, foot dorsiflexion weakness bilaterally, ataxia of gait, leg dysmetria on heel to shin testing and diminished pin prick and touch perception on the legs. On examination she had brisk reflexes, very tender muscles on palpation. Her EMG and NCV testing were normal. Her sedimentation rate, creatine kinase (CK) and antinuclear antibody (ANA) titer were normal. Her skin nerve biopsy showed normal findings. Her neurological examination she exhibited periodic reinnervation and neurogenic muscle loss. The patient was kept on the same medications that helped her symptoms to a degree and alleviated her hand muscle cramps.

Case 3:
A 57 year old was seen in consultation because of severe restless legs, sleep disturbance (probably secondary to periodic leg movements of sleep), chronic muscle pain and stiffness. She had two sisters with RLS. She had been diagnosed previously with fibromyalgia, attention deficit disorder without hyperactivity (ADD) and bipolar affective illness-depressive type. The patient gave no history of rheumatic disorders. She was taking therapeutic doses of venlafaxine, mirtazapine and ropirinole. On neurological examination she exhibited periodic movements of the body as she was generally uncomfortable and her limb muscles were tender at palpation. Her EMG and NCV testing were normal. She was prescribed gabapentin in addition to ropirinole, for alleviation of RLS and fibromyalgia.

Discussion

Pseudoathetosis (PA) is distinguished from true athetosis because it tends to be inconsistent or discontinuous, less stereotypical and perhaps of lesser severity, in patient with no identifiable brain lesion. Pseudoathetosis in association to peripheral neuropathy, either congenital (CMT disease as in Case One of this series) or acquired, for instance in cases of sensory Guillain Barre Syndrome and auto-immune neuropathy (multifocal sensory and motor neuropathy), may result from loss of proprioception (2, 13). If obvious neuropathy is clinically evident, presence of sensory ataxia supports an underlying proprioceptive deficit (14). The presence of neuropathy may not be obvious in patients with small fiber sensory neuropathy having no proprioception deficit, unless skin nerve biopsy demonstrates loss of dermal nerves, or if somatosensory evoked responses (SSERs) document slow signal conduction of peripheral stimuli. None of the three patient herein discussed had skin nerve biopsy or evoked responses studies. Meanwhile, these patients had FMA. Fibromyalgia is normally understood as a musculoskeletal or rheumatic disorder, but under new light, it may be classified as a form of small fiber painful sensory neuropathy. Serra, et al recently reported a sophisticated microneurography study in 30 females patients with FMA exhibiting slowing of high frequency electric signal transmission, increased spontaneous activity and sensitization to mechanical stimulation in peripheral C nociceptors(15).

Conversely, true athetosis cases are identified by presence of autoimmunity (autoimmune chorea with or
without neuropathy), history to exposure to toxins, presence of encephalopathy or of CNS lesions (16). Familial and sporadic cases of paroxysmal kinesigenic and non-kinesigenic dyskinesia are normally not accompanied by significant pain while central nervous system (CNS) abnormalities may be identified by MRI studies or by EEG (17). Pseudoathetosis must be also distinguished from neuropathic focal dystonia infrequently observed in patients with entrapment neuropathy (18).

Alternatively, PA may be interpreted as a reflex or compensatory movement disorder developing within the context of chronic pain or peripheral paresthesia, in where athetoid contorisions implement a conscious or semi-conscious attempt to relief pain. Movements may suppress pain paradoxically in painful conditions. One patient with post-traumatic mid-facial pain and Meige’s syndrome obtained relief by retrograde propulsion of the head (“retrocollis”), while a second patient also from this author practice, with familial barometric pressure induced migraine, was able to suppress her headache by performing a repetitive motor pre-dormital ritual of rotating her right foot (“jactatio extra-capitis”) (19,20).

On the other hand, the relationship of RLS-pseudoathetosis and pain is unclear. We know from clinical experience that patients with movement disorders experience sensory symptoms preceding movements. Patients with blepharospasm and Tourette syndrome often report “scratchy eyes” preceding the urge to close their eyes forcefully and individuals with complex regional pain syndrome develop on occasion involuntary movements often mislabeled as “psychogenic dystonia” when in fact they have “dystonia causalgia syndrome” (21). In RLS, sensory symptoms precede the movements as illustrated by Pelletier, et al, in a group of ten patients (22).

Other than a proprioception deficit and cortical somatosensory deafferentation, central disinhibition of the motor cortex has been proposed as a precondition for the development of involuntary movements in painful disorders of peripheral origin (23,24). The pathogenesis of this sensory-motor complex interaction is uncertain but direct motor cortex stimulation plays a role in pain mitigation in the pain–deafferentation animal model (25). The therapeutic effect of motor cortex stimulation is mediated by the activation of the cingulate cortex as detected by c-Fos expression, a recognized pain marker (26). Viisanen, et al, reported that pain modulation by the motor cortex is implemented by activation of dopamine (D2) receptors at striatal level (27). Of parallel significance striatal dopaminergic dysfunction has been demonstrated in patients with RLS (28). The latter report suggests that eventual dopaminergic neuronal excitotoxicity originating in persistent compensatory motor hyperactivity mediates the chronic evolution, and the eventual development of dopamine-agonist “augmentation phenomenon” observed in RLS. Finally, akathisia, a clinical condition defined as “an inability to sit” because of a sensation of inner restlessness and often precipitated by chronic use of antipsychotics, needs to be differentiated from CPA. However, patients with akathisia normally have no history of chronic pain or history of RLS.

I submit that CPA represents another face of RLS that can be also labeled less formally as “painful restless body syndrome”. Treatment with medications both useful for RLS, insomnia and pain (gabapentin or pregabalin as specific safer examples) are indicated on these patients, at least on a trial basis. Disorders of ominous prognosis like Huntington’s chorea must be ruled out.

References

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