Moving fingers moving toes: A heterogeneous entity

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INTRODUCTION:

Painful legs and moving toes (PLMT) is a rare disorder first described in 1971. It is characterized by semi-rhythmic spontaneous and continuous contractions of the toe flexors, extensors, abductors and adductors. The same type of disorder was also described in the upper extremities as painful arms and moving fingers (2). In 1993 a similar syndrome "painless legs and moving toes" was reported in which the patient had continuous, involuntary movements of all toes. In that particular case there was no history of preceding trauma or peripheral neuropathy, problems usually seen associated with PLMT. With or without pain, electrophysiological studies show different types of repetitive spontaneous discharges and complex alternating patters in antagonist muscles. We describe two patients who presented with movements of fingers and toes. They do not present with pain or other associated neurological disorder. Electrodiagnostic studies revealed myokymic discharges in doublets, triplets and multiplets. We postulate that different mechanisms lead to these discharges. Myokymic discharges can be seen as a manifestation of neuromyotonia. Autoimmunity plays a role in a subgroup of these patients and raises the possibility that immune-modulating therapy could be beneficial.

METHODS:

Case one:

A 47-year-old woman presented with a ten-year history of gradual progression of abduction and adduction movements of the left index finger. The movement was constant, including sleep. There was no pain. She was able to suppress the movement voluntarily only for few seconds. She did not report any history of trauma, weakness or sensory abnormalities in the affected limb. Three years prior to office presentation she developed involuntary flexion and extension of the second through fourth toes in the right foot. The movements were also continuous, painless and persistent during sleep. Her past medical history did not include any neurological problem, such as peripheral neuropathy.

On physical examination: Normal cranial nerves, full strength in al of the major muscle groups of the upper and lower extremities. Continuous semi rhythmic movements of flexion – extension and abduction - adduction of second through fourth toes in the right foot were noted. There was also adduction – abduction movements of the left index finger. Reflexes were symmetrical and plantar responses extensor. Sensory exam was normal for pin prick, touch, temperature and vibratory sensation. Gait and coordination were intact.

Laboratory tests: Complete blood count and chemistry profiles were within normal limits. Magnetic resonance imaging (MRI) scans of the cervical and lumbosacral spines were normal. Assay for potassium channel antibodies was negative. Nerve conduction velocities (NCV) were normal except for absent bilateral H reflex responses. Electromyography (EMG) showed insertional irritability of the left middle and lower cervical and right lumbosacral paraspinal muscles. EMG of the left first dorsal interosseus and right flexor digitorum brevis muscles revealed semi-rhythmic discharges in doublets, triplets and multiplets. The discharge frequency was 3-4 Hz with intraburst frequency ranging between 40-80 seconds. The amplitudes ranged between 200µV-4mV.

Case two:

A 31 year-old woman presented with a ten-year history of involuntary and continuous movements of her left toes. It began in the second through fourth toes and eventually involved of five ones. She denies sensory abnormalities, pain, tingling or burning sensation. In addition, she complained of a "dimple over my left buttock". Her past medical history was unremarkable for trauma or any significant neurological problem.

On physical examination: Normal cranial nerves. Motor exam revealed normal tone, bulk and power in the major muscles of upper and lower extremities. Reflexes were symmetrical and plantar responses flexor. There was mildly diminished sensation to pin prick over the lateral left leg. A circumscribed 5 cm depression was found over the left buttock. There was no discoloration of the skin or tenderness. However there was mild diminished sensation to pin prick, light touch and cold stimuli around the area.

Laboratory tests: Blood counts and basic metabolic panel were normal. Rheumatologic evaluation was negative. MRI scans of the brain, ankle and foot were normal. MRI and CT myelogram of the lumbosacral spine were unremarkable. Electrophysiological studies showed normal NCV. Electromyography was significant for myokymic discharges in multiplets affecting the left extensor digitorum brevis, left abductor hallucis and left flexor hallucis brevis. The discharges fired at a frequency of 2-3/second and last approximately 200msecs. The intraburst frequency varied between 70-8-/sec. The patient did not respond to a trial of carbamazepine and declined further therapy.

DISCUSSION:

In 1971, Spillane et al. described six patients with painful legs and moving toes (1). The symptoms ranged between 4 and nine years and included severe pain. The movements consisted of slow clawing and straightening, fanning and circular movements of the toes, involving flexion, extension and adduction. Electrophysiological studies were inconclusive. In one patient repetitive multiplet discharges (8 to 9) at a rate of 30/sec were noted. Sural nerve biopsy in one patient with pain and spontaneous clawing movements revealed segmental demyelination and short internodal segments. Lumbar sympathetic chain blockade with local anesthetic relieved the pain in four patients.

Schott suggested that trauma initiated the syndrome (3). The severity of trauma varied from minor injury to surgical intervention. He postulated that pain and focal involuntary movements were caused by peripheral nerve injury in nature, but also suggested that subsequently the central nervous system became involved. He proposed that minor peripheral trauma induced a spreading, irritative central discharge, possibly in the lower spinal cord, that variously involved sensory, motor and autonomic fibers. Later, Montagna et al. described three patients with continuous and involuntary movements of toes with painful legs and clinical evidence of peripheral neuropathy (4). Electrodiagnostic studies revealed bursts of spontaneous discharges of motor units in a frequency of 0.5 Hz. These bursts were noted to alternate in the extensor digitorum brevis and abductor digiti minimi muscles. The authors also noticed disruption of sleep patterns and some resemblance with the movement disorder restless legs syndrome. Consequently, Montana et al. postulated that the symptoms are initiated by a peripheral nerve trauma that subsequently affects higher, "central" levels. In 1993 a patient was described with a painless but otherwise similar involuntary, semi continuous movement of toes for over 33 years (6). EMG showed disproportion between the time of contraction and relaxation phases. Schoenen et al. (7) studied six patients and classified them into two groups: The electrophysiological studies in the first group had simple erratic pattern (SEP) consisting of discharges with low amplitude (100µV to 2mV) and frequency of 4-6 Hz. They occurred in erratic fashion, most often synchronously in antagonist muscles. The second group showed complex alternating pattern (CAP), which consisted of alternating bursts of motor activity. The amplitude of the discharges was higher (1 to 3 mV), and the duration longer compared to SEP group. The authors suggested that different mechanisms were involved in painful legs and moving toes. Those patients with SEP represented peripheral pathology with spontaneous activity, myokymia or fasciculations. Patients with CAP had central nervous system disorder as a cause, with EMG findings similar to dyskinesias.

Our patients are similar to those with painful legs and moving toes although neither patient has pain or a history of peripheral trauma. The absent H-reflexes in case 2 and insertional irritability of lumbar paraspinal muscles on EMG also suggests a peripheral nerve disorder. Both our patients had myokymic discharges with a painless movement disorder.

Neuromyotonia, a condition with excess muscle fiber activity usually presents with diffuse involvement and continuous, irregular "worm-like" movements. The pathogenesis of neuromyotonia is not yet established, although autoimmunity is postulated in many cases. This is also seen with exposure to toxins, radiation, paraneoplastic, hereditary, inflammatory neuropathies. On electrophysiological studies, neuromyotonia is characterized by doublet, triplet and multiplet discharges firing irregularly with high intraburst frequency (40-200/sec). Newsom-Davis et al. suggested that an antibody-mediated autoimmune mechanism, probably affecting the peripheral nerve K+ channels, was involved (9). This leads to interference with the function of K+ channels which normally stabilize the membrane potential and regulates repetitive firing. EMG study in neuromyotonia reveals myokymic discharges, neuromyotonic discharges, fibrillations and fasciculations. Our two patients with painless moving toes and fingers and three patients described by Schoenen had single-unit discharges that can be secondary to induction at a peripheral site. The pathophysiological mechanism in the patients is probably peripheral with some suprasegmental and central influence. Verhagen and Horstink suggested that peripheral sensory dysfunction of group II and III afferent fibers interact with supraspinal pathways leading to the syndrome, although the authors could not rule out segmental discharges of motor neurons (5). Zielasek et al. have shown neuromyotonic discharges in mice with hereditary myelinopathies (12). They postulate different sites of involvement on acquired neuromyotonia, i.e. distal motor nerve, nodal, proximal at neuron level or even in the central nervous system. This may explain heterogeneity of electrophysiological presentation in patients with "moving toes and fingers."

Muscle relaxants, such as baclofen, antiepileptic drugs such as phenytoin, carbamazepine and sodium valproate have been used to treat neuromyotonia effectively. However, our patients did not respond to these therapies favorably. Sympathetic blocks have reduced pain and abnormal movements in some patients.

CONCLUSION:

1- The common features shared in the syndromes of painful legs and moving toes, painless legs and moving toes and our patients are continuous movements of toes and fingers and semi rhythmic discharges in the involved muscles in the EMG studies. These common features could be summarized as continuous focal muscle fiber activity.

2- Continuous movements of toes and fingers in the absence of known pathology associated or not with pain may be a presentation of focal neuromyotonia.

3- Since possible autoimmune mechanism is suggested for neuromyotonia immune-modulating therapy may be useful in "focal neuromyotonia," especially associated with pain

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