NEUROMUSCULAR DISEASE ASSOCIATED WITH PAINFUL LEGS MOVING TOES SYNDROME: PRESENTATION OF TWO CASES

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SUMMARY :

We present two patients with neuromuscular disease developing painful legs moving toes syndrome in the course of their disease. The first case was a diabetic with polyradiculopathy and lumbar spinal stenosis. The second case had paraneoplastic sensory neuropathy. A non-alternating contraction pattern was observed by electromyography in the first patient, with spontaneous motor unit discharges grouped into low amplitude, synchronous bursts in the antagonistic muscles. Although a number of therapies was tried, the response was unsatisfactory. Carbamazepine provided slight relief in both patients.

Key Words: *Movement Disorders; Pain, Intractable; Electromyography; Carbamazepine.*

INTRODUCTION

The syndrome of "painful legs moving toes" is a rare disorder characterized by spontaneous causalgic pain in the lower limbs of the middle aged and elderly individuals and associated with peculiar involuntary movements of the toes and feet (1). Similar symptoms have been observed following a variety of lesions affecting the spinal cord (2), posterior nerve roots, the spinal ganglia and the peripheral nerves (1). Injuries to bony or soft tissues of the feet may also precipitate the disease (3, 4). Hypertrophic mononeuropathy can cause the syndrome (5). Some cases without a definite etiologic cause have been reported (4). Symptoms can occur in the upper limbs called as painful arm or hands and moving fingers (6, 7). We report two patients with neuromuscular disease developing painful legs moving toes syndrome; one associated with lumbosacral polyradiculopathy and the other with paraneoplastic sensory neuropathy.

CASE REPORTS

Case 1: A 74-year-old female patient with a past history of hypertension and diabetes was seen in the neurology outpatient department because of burning pain in her legs and involuntary movements of her feet and toes. She had been complaining of backpain and paresthesias in her legs and feet for six years. Leg pain and involuntary movements appeared a year before. The symptoms alleviated at night when she was tired and were aggravated by walking or standing. The condition gradually worsened over months and she began to develop dyskinesia in her lower limbs. On neurologic examination, stereotyped, rapid irregular involuntary movements of her feet and toes characterized by flexion, extension, and abduction were observed. There were slight (MRC

5-/5) bilateral weakness of ankle dorsiflexion and bilateral L4, L5 and S1 dermatomal hypoesthesia. Both ankle and patellar reflexes were absent. Blood cell count and biochemistry profile were normal. Lumbosacral computerized tomography and magnetic resonance imaging demonstrated degenerative changes in the vertebral bodies. Diffuse annular bulging and bilateral narrowing of the intervertebral foraminas at the L2-S1 levels were observed. Nerve conduction studies using surface stimulating and recording electrodes revealed low amplitude compound muscle action potentials in both peroneal and posterior tibial nerves, with normal sensory nerve conduction velocities (NCV) (Tables 1 and 2). Needle EMG was diagnostic of bilateral L4-S1 polyradiculopathy (Table 3). A simultaneous two channel EMG recording was performed on two muscle pairs in the right lower extremity using a Dantec Cantata electromyograph. Surface electrodes were used while studying the tibialis anterior (TA) and medial gastrocnemius (MG) muscle pair. Extensor digitorum brevis (EDB) was studied by a surface electrode, while recording was made by a concentric needle electrode from the flexor hallucis brevis muscle (FHB). In the TA-MG pair, abnormal feet movements caused spontaneous repetitive motor unit discharges grouped into bursts of 0.5-3 s duration, with an irregular firing rate of 0.3-2 Hz, occurring synchronously in antagonistic muscles. In the EDB-FHB pair single unit discharges were more prominent with grouped bursts of 100-700 ms in duration and a firing frequency of 1-8 Hz. The amplitude of bursts was low, ranging from 200-1,200 μV(Fig. 1). Amitriptyline, haloperidol, baclofen, clonazepam, L-dopa, biperiden, and transcutaneous electrical nerve stimulation (TENS) provided no beneficial effect. Carbamazepine relieved the pain to a certain degree.

Case 2: A 55-year-old female patient with a diagnosis of adenocarcinoma of the ovary since 1993 was brought to the neurology outpatient department. History revealed that she had been previously given cyclophosphamide-cisplatinum-epirubicine combination chemotherapy several times and put on hexamethylmelamine afterwards. For three years, she had been suffering from burning pain, paresthesias and cramps in her legs and feet. Involuntary movements in her feet started two years ago. On neurological examination,

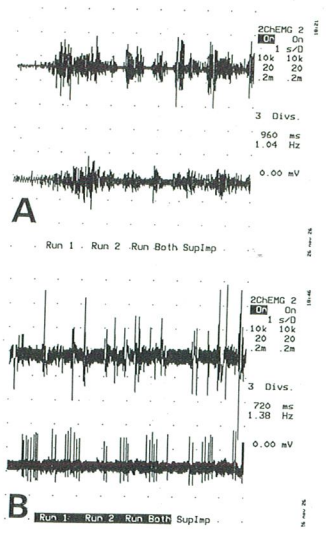

Fig - 1 : Two channel EMG recording of (A)TA(upper tracing)-MG(lower tracing), (B)EDB(upper tracing)-FHB(lower tracing) muscle pairs. In (A) MUP bursts are of long duration and slow frequency. In (B) bursts have shorter duration and faster frequency. Note low MUP amplitudes and arrhythmic synchronous firing of the motor units, typical of a peripheral disorder.

bilateral stocking type distal hypoesthesia, impaired vibration and position sense in the toes were found. Deep tendon reflexes were unobtainable in the lower extremities. Involuntary, stereotyped continual movements characterized by flexion, extension and abduction of the feet and toes were observed particularly on leg extension. She was unable to stand and walk unaided. Nerve conduction study was diagnostic of a diffuse sensory peripheral neuropathy (Tables 1 and 2). She could not tolerate needle EMG. A sural nerve biopsy was refused. Amitriptyline, carbamazepine and narcotic analgesics slightly relieved the pain, but had no effect on involuntary movements.

DISCUSSION

Both of our patients showed a fairly uniform pattern of presentation with constant troubling pain in the legs, burning in quality, associated with involuntary movements of both toes and feet. In the second case with ovarian cancer, symptoms seemed to be more disabling and involuntary movements more prominent on leg extension. The prominent sensory ataxia made her suffering worse. There was no upper extremity involvement in either case. No involuntary movements in the body or face were present. Restless legs syndrome is ruled out by the severe pain, unrelated to the

Nerve	Amplitude(mV)		CV(m/s)		DL(ms)	
	Case 1	Case 2	Case1	Case 2	Case 1	Case 2
Median (APB)	5.5	6.9/8.0*	63.3	52.0/66.6	3.0	3.0/3.2
F latency (<32) ¶					23.6	27.6/28.0
Normal	>4.0		>49.7		<3.8	
Ulnar (ADQ)		15.6		64.2		2.8
F latency (<32)						27.2
Normal	>5.0		>49.9		<3.3	
Peroneal (EDB)	1.4/2.5	7.7/6.4	44.1/46.8	47.5/41.9	4.8/4.0	4.4/4.6
F latency (<52)					NP/NP	49.6/49.2
Normal	>3.5		>40.9		<5.8	
Tibial (AH)	1.2/1.1	4.4/8.7	41.4/34.3	40.3/39.7	6.0/4.2	4.6/5.4
F latency (<52)					NP/56.4	53.2/53.6
Normal	>3.5		>39.6		<6.0	

ADQ: Abductor digiti quinti, AH: Abductor hallucis, APB: Abductor pollicis brevis, CV: Conduction velocity, DL: Distal latency, EDB: Extensor digitorum brevis. NP: No potential
* Right/Left, ¶ Normal values are indicated in parentheses.

Table 1 : Motor nerve conduction studies.

Nerve	Amplitude(μV)		CV(m/s)	
	Case 1	Case 2	Case 1	Case 2
Median	11.2	6.0/9.4*	42.8	41.6/43.4
Normal	>10		>39.4	
Ulnar		6.2/4.8		38.5/43.0
Normal	>7		>37.3	
Sural	21.2/10.8	8.2/NP	51.4/52.2	31.2
Normal	>5		>33.8	

NP: No potential * Right/Left

Table 2 : Sensory nerve conduction studies.

Activity	Right	Left
Spontaneous		
Fib/PSW	TA, MG, PL	TA, MG, L4, L5
Fas	PL	
Absent	L3-S1	VL, PL, L3, S1
VMUP		
Giant	PL	
Normal	TA, MG	TA, MG, VL, PL

Fas: Fasciculation potentials, Fib: Fibrillation potentials, L4,...S1: Lumbar and sacral paraspinals, MG: Medial gastrocnemius, PL: Peroneus longus, PSW: Positive sharp waves, TA: Tibialis anterior, VL: Vastus lateralis, VMUP: Voluntary motor unit action potentials

Table 3 : Case 1: Needle electromyography.

sleep-wake cycle and not relieved by leg movements. These involuntary movements described as pseudoathetosis by Verhagen et al. (7) are different from nocturnal myoclonus.

Our first case had bilateral lumbosacral radiculopathy most likely due to lumbar spinal stenosis as demonstrated in the radiological studies. She also had diabetes, a common cause of painful sensory neuropathy and lumbosacral radiculopathy (8, 9). Pain sensation in "Painful legs and moving toes" arise mainly from damaged group II and III fibers (7). This brings into mind the coexistence of diabetic neuropathy in our patient. However, sensory NCV were normal, which certainly cannot rule out a small fiber neuropathy, as electrodiagnostic testing particularly with surface electrodes, mainly assesses conduction in large myelinated sensory fibers (10). Therefore, contribution of diabetes to her symptoms cannot be excluded. The second case had paraneoplastic sensory neuropathy, with symptoms getting worse despite adequate treatment of the cancer. The prominent sensory ataxia suggests dorsal root ganglionitis and a sural nerve biopsy would be desirable to differentiate this entity from an axonopathy. Therefore, a sensory neuropathy due to cancer chemotherapy can not definitely be ruled out, but seems highly unlikely because combination chemotherapy had been stopped a long time ago. Although a unilateral syndrome after herpes zoster infection of the right L5 dorsal root ganglion has been previously reported (11), this is the first "Painful legs moving toes" case caused by a paraneoplastic sensory ganglionopathy.

Schoenen et al. (12) described 2 patterns of EMG activity in this syndrome. Some patients show short low amplitude bursts of 4 to 6 Hz, occurring in erratic fashion synchronously in the antagonistic muscles, suggesting a peripheral disorder. These involuntary movements sometimes cannot be suppressed by nerve block, due to the distal location of the lesion. A second group displays higher amplitude and longer duration bursts, alternating in antagonistic muscles with a slow 1.5-3 Hz frequency, attributed to central nervous system lesions. Local anesthesia is effective in these patients. EMG study in our first case showed low frequency, arrhythmic motor unit bursts of long duration, particularly in the TA-MG pair, reminiscent of a central disorder. However, synchronous firing in the antagonistic muscles and low amplitude of the discharging units were atypical. EDB-FHB pair showed shorter synchronous bursts of higher frequency more typical of a peripheral disorder. It was clear from this EMG study that our patient had a peripherally located disorder. Whether the lesion was located in the peripheral axons or at the root level remains moot, since we did not perform anesthetic block of the peripheral nerve.

Despite evidence for the peripheral location of lesion in both of our patients, pure peripheral disorders cannot adequately explain the excruciating pain and continuous involuntary movements. Some hold that afferent discharges from the periphery are organized at the spinal or higher levels to give rise to pain (1). Spinal motor neurons are excited, which initiates involuntary movements of the feet and toes (13). Thermocoagulation of nucleus ventralis intermedius abolished movements and reduced the pain in a patient (7).

Different therapeutic regimens have been tried in the past, many without success. ß-blockers, anticonvulsants, neuroleptics, tricyclics give no relief (12). One patient, developing the syndrome during neuroleptic therapy responded to clonazepam and baclofen combination (14). Our patients obtained some but not complete relief of pain by carbamazepine. However, involuntary movements were not affected. Progabide, a GABA receptor agonist was useful in one case; symptoms improved and gradually disappeared in six months (13). TENS with vibratory stimulation applied to the plantar surface of the foot produced effective analgesia and resulted in complete pain relief in one patient. This treatment stimulates large diameter afferents and modulate pain pathways, inhibiting involuntary movements as well (15). Our first patient did not obtain significant benefit form TENS alone. This rare syndrome can be observed originating from peripheral afferent lesions in the course of neuromuscular disorders, where symptomatic treatment poses a difficult problem.

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