**Case Report**

Focal neuromyotonia as a presenting feature of lumbosacral radiculopathy

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**Abstract**

Neuromyotonia is characterized by motor, sensory, and autonomic features along with characteristic electrophysiologic findings, resulting from hyperexcitability of the peripheral nerves. We describe the case of a 36-year-old man, who presented with the disabling symptoms suggestive of focal neuromyotonia involving both the lower limbs. His neurological examination revealed continuous rippling of both the calf muscles with normal power, reflexes, and sensory examination. Electrophysiology revealed spontaneous activity in the form of doublets, triplets, and neuromyotonic discharges along with the neurogenic motor unit potentials in bilateral L5, S1 innervated muscles. Magnetic resonance imaging lumbosacral spine revealed lumbar intervertebral disc protrusion with severe foraminal and spinal canal stenosis. Patient had good response to steroids and carbamazepine. The disabling focal neuromyotonia, occurring as a result of chronic active radiculopathy, brought the patient to medical attention. Patient responded to medical management.

**Key Words**

Electrophysiology, magnetic resonance imaging, neuromyotonia, radiculopathy

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**Introduction**

Neuromyotonia is a rare disorder of autoimmune origin, which is characterized by the hyperexcitability of peripheral motor nerves. Neuromyotonia clinically presents with muscle twitches, muscle cramps, muscle stiffness, increased sweating, muscle hypertrophy, pseudomyotonia, and reduced muscle power.\(^1\) Neuromyotonia was initially termed as the cramp fasciculation syndrome. The first description was provided by Tahmoush et al., and the condition was later described, in detail, by Isaac.\(^2,3\) Neuromyotonia is either hereditary or acquired and either generalized or focal. Apart from the genetic, autoimmune and paraneoplastic causes, various rare other conditions, like toxins, radiation therapy, infections, mechanical irritation during the surgery, can produce neuromyotonia. Infrequently, chronic radiculopathies can produce focal myokymia and neuromyotonia.\(^4\) We describe an unusual case of focal neuromyotonia involving both lower limbs.

**Case Report**

A 36-year-old man presented with back pain and cramp like pain in both lower limbs of 4 month duration. Pain was initially dull aching localized to the lower back, aggravate on walking and partially subside with rest and analgesics. Simultaneously, he developed cramp like pain in both calves along with stiffness, which he initially noticed while walking. Initially, he had to take pauses while walking. Later on, there was severe difficulty in walking. Since, 2 months, patient developed severe cramp like pain even at rest, affecting his sleep. Along with the muscle cramps, he noticed formation of a transient lumps in both calves due to focal muscular contraction. He did not complain of any radicular pain, tingling, numbness or any other sensory complaints. There was no excessive sweating. He noticed twitching in both calves since 2 months. There was no history of any weakness or thinning noticed in lower limbs. He did not have any complaints in upper limbs. Family history was not significant. His general examination was unremarkable. Higher mental functions and cranial nerves were normal. Upper limb examination was normal. Lower limb examination revealed mild thinning in the left leg. There was frequent rippling noted in both calves. Tone, power, and reflexes were normal. There was no sensory loss, but because of pain and cramps he had severe limitation of walking. Spine examination revealed tenderness and spasm in lower paraspinous muscles. There was no deformity. Straight leg-raising testing was positive on both sides. There was no evidence of autonomic involvement.

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Hematological and biochemical tests including blood sugar, kidney function tests, serum electrolytes including calcium, thyroid profile were normal. Nerve conduction study revealed mildly reduced tibial compound muscle action potential amplitude with normal sural sensory nerve action potential. Electromyography (EMG) in bilateral gastronomies lateral head revealed spontaneous activity in the form of doublets, triplets and neuromyotonic discharges (160-200 Hz waning type of discharges with pinging sound). There was reduced recruitment, incomplete interference pattern along with polyphasic motor unit potentials (MUP) in bilateral L5, S1 innervated muscles (Tibialis anterior and hamstrings). Lumbar paraspinal EMG revealed fibrillation potentials. Bilateral H reflex testing didn't show any recordable waveforms [Figure 1]. EMG in both quadriceps muscles was normal. Magnetic resonance imaging Lumbosacral spine revealed annular tear along with disc protrusion at L3-4, L4-5 with severe foraminal (right > left) and spinal canal stenosis along with compression of the nerve roots. There was contrast enhancement noted in the protruded disc and nerve roots suggestive of active inflammation [Figure 2]. Patient received intravenous methylprednisolone for 3 days, carbamazepine 200 mg twice a day, duloxetine 20 mg once a day and physiotherapy. There was marked relief in pain, stiffness and cramps after 10 days. Even the calf muscle rippling was drastically reduced.

Discussion

The present patient is unusual because, in addition to backache and neurogenic claudication, patient had focal neuromyotonia involving both the calf muscles. However, one thing that was unusual in our case was that, patient had more symptoms because of focal neuromyotonia along with chronic backache. Generalized neuromyotonia is usually, a sporadic autoimmune disease but for focal neuromyotonia several peripheral and central disorders of nervous system, like radiation plexopathies, inflammatory and hereditary neuropathies, multiple sclerosis, brain stem mass lesions and snake bite, were described.[5-7] Pangaria and co-workers reported etiological spectrum in 20 cases of neuromyotonia. Eleven of these 20 patients reported intake of unknown
ayurvedic medicines in the preceding 1 month. Bell’s palsy was associated in four, peripheral neuropathy in two and residual poliomyelitis in two patients.\textsuperscript{[6]}

There are several reports ascribing lumbosacral radiculopathy with calf hypertrophy but not to calf neuromyotonia. Lumbosacral radiculopathy has been shown produce in neurogenic unilateral or bilateral calf muscle hypertrophy. When calf hypertrophy occurs unilaterally, it is usually the result of an S1 radiculopathy. Unilateral calf hypertrophy has also been reported with L5 radiculopathy, polyneuropathy, and poliomyelitis. Bilateral calf hypertrophy has been reported to be caused by polyneuropathy or anterior horn cell disease. Electrophysiology in these patients, frequently, reveals electromyographic findings suggestive of neuromyotonia. The muscle hypertrophy is considered secondary to persistent overactivity present in the affected muscle groups. Proposed mechanism responsible for overactivity of the affected muscle included ephaptic transmission among nerve fibers at the root compression or type I fiber hypertrophy in response to the complex repetitive discharges.\textsuperscript{[8-10]}

The electromyographic changes of neuromyotonia originate from peripheral nerves. Neuromyotonia, on electromyographic evaluation, is characterized by repetitive, single MUPS that fire spontaneously at a high frequency, 150-250 Hz, with characteristically decrementing amplitudes and frequencies. The single motor unit activities usually start and stop suddenly and can last for up to several seconds. EMG may also reveal electrical myokymia, which consists of rhythmic bursts of waveforms representing single motor units firing as doublets, triplets, or multiplalets. The electromyographic changes of neuromyotonia, possibly, arise from spontaneous depolarization or ephaptic transmission along segments of a demyelinated nerve secondary to altered membrane excitability.\textsuperscript{[7,11,12]} Our patient had typical findings of neuromyotonia limited to the muscles innervated by the L5, S1 radicles along with spontaneous activity in lumbar paraspinous muscles.

Several sodium channel-blocking agents, like phenytoin, carbamazepine, and mexiletine, which act as membrane-stabilizing agents, are often helpful in patients with neuromyotonia. These drugs are reduces muscle stiffness. Several immune-modulatory therapies like corticosteroids, azathioprine, plasmapheresis, and intravenous immunoglobulin, were used in patients with generalized neuromyotonia, with variable success.\textsuperscript{[13,14]} We also used methyl-prednisolone along with membrane stabilizing drugs along with symptomatic treatment with marked relief. Methyl prednisolone was given because neuroimaging had revealed inflammatory changes in the protruded disk as well in the compressed root.

**Conclusion**

Rarely focal neuromyotonia can be a presenting feature of radiculopathy. Neuromyotonia might be more troublesome than the typical features of radiculopathy. Immune-modulatory therapy and sodium channel-blocking drugs help in relieving symptoms of neuromyotonia.

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