Morvan's Syndrome—A Case Report of a Rare Clinical Scenario

Dinesh Kumar¹, Saniya Kapila²

ABSTRACT

Background: Neuromyotonia is a disorder of peripheral nerve hyperexcitability, causing spontaneous muscular activity. It is an extremely rare disease. Infrequency with which it is encountered and nonspecific, vague symptoms in young people make it a formidable diagnostic challenge.

Case description: We present a case of a 34-year-old male with complaints of twitching in muscles and pain in the thigh and calf region. These complaints occurred on and off since the last 6 months. He has a positive family history of similar symptoms. The patient was positive for serum voltage gated potassium channels (VGKC) antibodies. Electromyography (EMG) studies showed evidence of neuromyotonia. A diagnosis of Morvan's syndrome (neuromyotonia) was made on the basis of symptoms and EMG findings. The treatment of the patient was started on intravenous steroids, antiepileptics, and SSRI.

Conclusion: Diagnosis of neuromyotonia is a challenge. Awareness among people is important to mitigate the symptoms and provide timely management.

Keywords: Agrypnia excitata, Antivoltage-gated potassium channel antibodies, Morvan's syndrome.

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INTRODUCTION

Severe muscle weakness with a stiff jerky body in a young person is an uncommon disorder, where the defect may be in the muscles or the nerves supplying these muscles or a metabolic disorder. Neuromyotonia (or Morvan's syndrome) is a disorder of the peripheral nerve hyperexcitability, causing spontaneous muscular activity owing to repetitive action potentials. Only about 100–200 cases of neuromyotonia have been reported so far.¹ Patients can present with muscle cramps, muscle stiffness, increased muscle tone, walking difficulties, hyperhidrosis, fasciculations, myokymia (quivering of a muscle), and myoclonic jerks.²

CASE DESCRIPTION

A 34-year-old male presented with complaints of twitching in muscles and pain in the thigh and calf region (Fig. 1), insidious in onset, and progressive in nature.

These complaints were present since the last 6 months, for which he had been taking painkillers. He has a family history of abnormal movements in various muscle groups (his both siblings were affected). On examination, muscle power, coordination, and reflexes were normal. There was no other evidence of systemic diseases. On biochemical testing, his metabolic parameters (i.e., serum calcium, magnesium, phosphorus, and potassium levels) were normal, ruling out any deficiency of these. The patient's serum was tested positive for serum VGKC antibodies. The EMG showed short-duration polyphasic motor unit action potential (MUAP) (waxing and waning pattern), which persist during sleep (Fig. 2).³

Nerve-conduction studies showed normal motor and sensory nerve conduction. Hence, on the basis of clinical findings, a positive family history, presence of specific antibodies, and classical EMG findings, a diagnosis of Morvan's syndrome was made. The treatment of the patient was started on intravenous steroids, which led to improvement in the symptoms. Antiepileptics (phenytoin

¹Department of Medicine, Sri Guru Ram Das Institute of Medical Sciences and Research, Amritsar, Punjab, India

²Sri Guru Ram Das Institute of Medical Sciences and Research, Amritsar, Punjab, India

Corresponding Author: Dinesh Kumar, Department of Medicine, Sri Guru Ram Das Institute of Medical Sciences and Research, Amritsar, Punjab, India, Phone: +91 8427158005, e-mail: manishksharma27@gmail.com

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and carbamazepine) and SNRI (duloxetine) were given for a short duration for treating the neuropathic pain.^{4,5}

DISCUSSION

Neuromyotonia is a disorder in which repetitive action potentials are generated in the nerve, leading to prolonged muscle contraction and muscle stiffness. Only about 100–200 cases of the disease have been reported so far. The most common feature is muscle twitching (present in 90% of cases).⁶ Central nervous system changes (i.e., mood changes and sleep disorder) were present in a quarter of cases.⁷ A very classical symptom—agrypnia excitata (which consists of severe total insomnia of long duration associated with mental confusion, hallucinations, motor agitation, and complex motor behavior mimicking dreams, and autonomic activation)—may be present in some cases but was not seen in our patient.⁸ CNS and autonomic symptoms are caused by an impaired corticolimbic control of the subcortical structures regulating the sleep-wake and autonomic functions. Neuromyotonia is a channelopathy that results from an antibody-mediated attack against the peripheral nerve VGKCs.

Diagnosis is made on the basis of clinical features, EMG findings, and nerve conduction studies. Treatment includes

Figs 1A to D: (A) Pseudohypertrophy of lower limb muscles; (B) Wasting of anterior compartment of leg muscles; (C) Distal weakness of muscles of right leg; (D) Distal weakness of muscles of left leg

immunosuppressants, steroids, antiepileptics, and plasmapheresis. Carbamazepine at a dose of 400–600 mg per day leads to disappearance of fasciculations and neuropathic pain.⁴

Differential Diagnosis

The differential diagnoses for neuromyotonia are:

- Stiff man syndrome (discharges appear in sleep)⁹
- Motor neuron disease (neuromyotonic discharges can be seen early in the course).¹⁰

CONCLUSION

Morvan's syndrome is a rare neurological disorder but has a good prognosis if diagnosed correctly and in time.

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Fig. 2: EMG showing short-duration polyphasic MUAP (waxing and waning pattern), which persists during sleep