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***Case Report***

# CASE REPORT OF MORVAN SYNDROME

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## ABSTRACT
Morvan's fibrillary chorea or Morvan's syndrome is characterized by neuromyotonia, pain, weight loss and various severe neuropsychiatric manifestations such as hallucinations, delirium and insomnia. The occurrence of Morvan syndrome is quite rare, as there are only 14 reported cases in English literature. The natural history of Morvan's is highly variable. We describe an 18-year-old male patient with neuromyotonia, pain in the chest, abnormal jerky movements of the left thigh, autonomic symptoms, motor weakness, delirium and cognitive impairment. The electromyography findings were suggestive of neuromyotonia. It showed spontaneous activity in the form of fasciculations in all the muscles tested except tibialis anterior of the right thigh. Although metabolic profile, as well as VGKC antibodies results of this patient, were unremarkable. The MRI findings of brain and spine, as well as CSF analysis, were also unyielding. We have tried a high dose of steroids for initial management in this patient, but his condition did not improve; however, he subsequently responded to a course of immunoglobins. In Morvan syndrome, plasma exchange therapy was also proven to be effective. From our case presentation, though we have mentioned variable clinical manifestations, the mechanism behind these presentations is still not fully understood. However, further research on combining different imaging modalities is required for this rare syndrome due to its variable clinical presentation.

## INTRODUCTION
Morvan syndrome is a rare, autoimmune disease named after nineteenth-century french physician Augustin Marie Morvan. He described a patient with muscle twitching associated with muscle pain, excessive sweating and insomnia. The occurrence of Morvan syndrome is quite rare. The natural history of Morvan's is also highly variable, predominantly found in males patient. Neuromyotonia found in Morvan syndrome is a rare peripheral nerve hyper-excitability disorder producing spontaneous muscular activity as a result of repetitive action potentials. It is also associated with various autoimmune diseases. In this case presentation, we describe a patient with all the features of Morvan syndrome without any detectable levels of VGKC antibodies.

## CASE
A previously healthy 18-year-old male presented with Pain on the left side of the chest since two months, insidious, progressive and radiated to the left thigh. He was taking NSAIDS but required opioids later. He also had episodes of abnormal Jerky movements of the left thigh, severe attacks of headache, agitation, anxiety and crying spells, temporarily relieved by IV benzodiazepine. After admission, he also developed motor weakness (was not able to bear weight)and sensory loss of touch and pain sensations in the left leg. There were no episodes of tongue bite, bladder or bowel incontinence. There were also episodes of Breathlessness, tachycardia, high fever, a decline in cognition and talking incoherently. The EMG sampling was done in bilateral tibial anterior, vastus lateralis and other muscles of the thigh. The findings showed increased insertional muscle activity as well as spontaneous activity in the form of fasciculations, doublets and triplets in all the muscles tested except tibialis anterior muscle. Therefore, the findings were suggestive of neuromyotonia. There was no family history for similar complaints; however, one of his cousins is suffering from autoimmune IBD.
The metabolic profile for potassium, calcium, magnesium, phosphorus was normal. Antibody scan for Glutamate receptor NMDA, AMPA1, AMPA2, CASPR, LGi1 (VGKC associated), GABA1,2 was negative. Chest x-ray and chest CT were normal (done for Ruling out thymoma). CSF analysis, brain

MRI, EEG, were unyielding. High dose steroids were started, but his condition did not improve, he subsequently responded to a 5-day course of IVIg. Antiepileptics (phenytoin, valproate, levetiracetam) SSRI (paroxetine) and Benzodiazepines were also given for neuropathic Pain.

## DISCUSSION

Morvan syndrome is a rare disorder characterized by peripheral nerve hyperexcitability, autonomic dysfunction, and encephalopathy. It is also known as Morvanfibrillary chorea (MVC) (Abou-Zeid *et al.*, 2012). Morvan syndrome is mostly seen in males. It is believed to be an autoimmune disease because of its association with thymoma as well as with voltage-gated potassium channel (VGKC) antibodies. VGKC antibodies may play a critical pathogenic role in peripheral as well as the CNS manifestation; however, these antibodies are not always detected in patients with Morvan syndrome (Newsom-Davis *et al.*, 2003).

Morvan syndrome is mainly a clinical diagnosis. The cause of characteristics features of Morvan syndrome are by central, autonomic and peripheral nervous system hyperactivity (Haug *et al*., 1989). The CNS features of Morvan syndrome include encephalopathy, severe insomnia, vivid complex hallucinations, delirium, spatial disorientation, confusion, amnesia and agitation while the autonomic nervous system (ANS) features, due to autonomic instability are excessive sweating, fever, palmoplantar erythema, drooling, severe constipation, excessive lacrimation, arrhythmias and hypertension (Buckley *et al.*, 2001*)*. Moreover, the peripheral nervous system (PNS) features consists of peripheral nerve hyperexcitability, neuropathic Pain, areflexia and a stocking-type sensory loss. The manifestation of continuous muscle fibre activity in various muscle fibres seems as neuromyotonia (myokymia) (Iwaski *et al*., 1990). However, it is significantly less likely for an individual patient to manifest all the above clinical features as they generally have varied clinical features.

For the diagnosis of Morvan syndrome, a high index of clinical suspicion was required when a patient presented with a combination of various clinical features. Most of the investigations that performed in Morvan syndrome patients, such as CSF analysis, brain MRI, EEG, PET scan, are typically unyielding. EMG (electromyography) studies can confirm symptoms of myokymia. However, the detection of VGKC-complex antibodies is quite diagnostic but not always detected in all cases.

Clinicians have tried several different treatment modalities with variable clinical response. These include antiepileptic agents such as carbamazepine, valproate, phenobarbital, phenytoin. Moreover, immunomodulators and surgical procedures such as thymectomy were also proven effective in the number of cases. In this patient, high dose steroids were started, but his condition did not improve, however; subsequently, five days course of immunoglobin were proven effective. Moreover, plasma exchange appears to be the most effective treatment available for Morvan syndrome as seen in several cases, besides immunosuppression (Josephs *et al*., 2004). However, due to unclear reasons, clinical response to plasma exchange and immunosuppression is also variable.

## CONCLUSION

Although a rare presentation, we should consider the diagnosis of Morvan syndrome in a patient with the above clinical features but a high index of suspicion is required. There are various diagnostic modalities, such as imaging studies, CSF analysis, VGKC antibodies. The prognosis of Morvan syndrome is quite variable, some remit spontaneously, others require extensive treatment while some are fatal. In our patient, clinical improvement was seen after IV immunoglobin. However, best results are achieved with an inter-professional team of specially trained nurses and clinicians.

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