An Extremely Rare Case of Acquired Neuromyotonia

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Case Description

The patient presented with worsening symptoms of dysarthria, cognitive impairment, weakness and worsening seizure-like violent tremors of his right arm neck and trunk lasting 30 seconds and separated 5-10 minutes apart. His work-up found autoimmune encephalitis with N-type calcium-channel antibodies. EMG revealed Neuromyotonia. He was treated with IVIG and corticosteroids with some improvement. He missed his last IVIG dose. Two weeks later, there was an acute re-emergence and progression of prior symptoms. There was also new findings of flair hyper-intensities indicating worsening inflammatory response. MRI was consistent with cerebritis. Patient then received plasmapheresis and began IVIG with improvement of symptoms and was discharge to the acute rehab unit. When he received his first sessions of rehab therapy his symptoms got noticeably worse. He was then give another dose of steroids and transferred back the neurology unit. He will also begin treatment for the prostate cancer soon.

References

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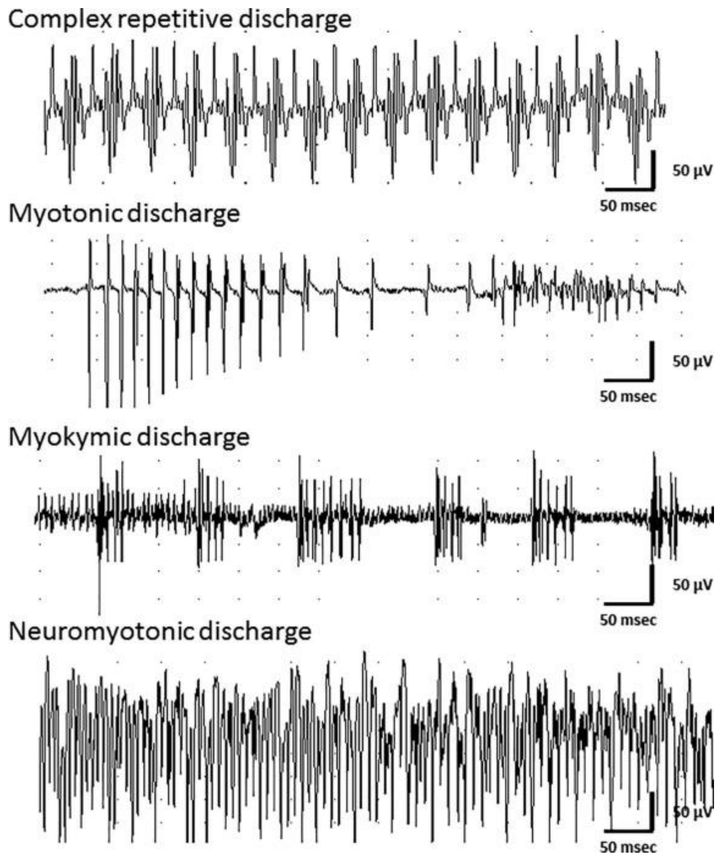

*Images do not reflect the condition of the patient and are for illustration purposes only.

Clinical entity	Aetiology	Clinical findings	Electrodiagnostic findings
Acquired disorders			
Isaacs syndrome	VGKC antibodies	Onset predominantly in the mid-40s Continuous muscle twitching and myokymia, muscle hypertrophy, weight loss, hyperhidrosis Preserved muscle strength and tendon reflexes	Normal sensory and motor NCS, except for after-discharges Neuromyotonic and myokymic discharges, doublets or triplets or multiplets, fasciculation potentials, fibrillation potentials, and cramp discharges, occurring spontaneously or activated by voluntary muscle contraction on needle EMG
Morvan syndrome	VGKC antibodies	Similar to Isaacs syndrome plus CNS: encephalopathy, headaches, drowsiness, and hallucinations	Similar to Isaacs syndrome
Cramp fasciculation syndrome	Uncertain	Muscle cramps, exercise intolerance, and muscle twitching	After-discharges on repetitive nerve stimulation and fasciculation potentials on needle EMG
Other	Toxins: lead, silver and gold		Myotonic discharges on needle EMG

Discussion

This is a report of an extremely rare case of neuromyotonia. Neuromyotonia is a disease with peripheral nerve hyperexcitability that causes spontaneous repetitive motor unit action potentials. The prevalence is unknown but 100–200 total cases have been reported. The cause is unknown, and it can be hereditary (chromosome 12), acquired/autoimmune (anti-voltage-gated potassium channel) or paraneoplastic (mostly thymoma, but also colon and prostate). Peripheral and central symptoms can occur including episodes of involuntary contractions, dysarthia, mood/cognition changes, hallucinations, hyperhidrosis, and myokymia. EMG findings include a sensory-motor demyelinating polyneuropathy, with fasciculations, myokymia, doublets, multiplets, and high frequency discharges, and after-discharges. Cerebrospinal fluid analysis may show oligoclonal bands. The best course of treatment is unknown, but anticonvulsants and plasma exchange have shown benefit. Physical therapy is often a treatment to decrease stiffness and prevent atrophy.

Conclusion

This is a rare and severe case of acquired, autoimmune vs. paraneoplastic neuromyotonia that seemed to be worsened by physical therapy. There are few documented cases, and no cases were found on literature search with such severe episodes of contraction. It is unclear if it truly falls under the parameters of Marvan Syndrome or Isaacs Syndrome, and is an unusual case.