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VOLTAGE-GATED POTASSIUM CHANNEL (VGKC) ANTIBODY ASSOCIATED NEUROMYOTONIA: A CASE REPORT

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ABSTRACT

Neuromyotonia is associate immune mediate disorder characterized by muscle stiffness caused by exaggerated muscle cell activity with elevated voltage gated potassium channels. A manifestation usually includes muscle twitching during rest, muscle cramps, delayed muscle relaxation, increased sweating with motor weakness. The clinical significance of this VGKC antibody test can only be determined in conjunction with the patient's clinical history and related laboratory testing. I present a case of 27 year old woman admitted with the complaints of pain in extremities, whole body weakness and need for assistance to do her ADLs. VGKC antibodies were positive and rests of other investigations were in normal range. The patient was subsequently placed on anticonvulsant and steroids therapy.

KEYWORDS

VGKC and Neuromyotonia.

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INTRODUCTION

Autoantibodies directed against VGKC are primarily seen in patients with neuromyotonia, Morvans syndrome, autoimmune encephalitis or idiopathic epilepsy. These antibodies are often directed against the potassium channel-associated proteins CASPR2 (contact in associated protein-2) and LGI1 (leucine rich glioma inactivated protein-1). Antibodies against CASPR2 are more commonly seen in neuromyotonia and Morvans syndrome, while antibodies to LGI1 are more commonly seen in autoimmune encephalitis. Neuromyotonia is a form

of peripheral nerve hyper excitability that causes increased muscular activity resulting from repetitive motor unit action potentials of peripheral origin. Morvans syndrome resembles neuromyotonia where symptoms are associated with pain and severe insomnia. Immunotherapy is more effective against auto antibodies. The clinical significance of this test can only be determined in conjunction with the patient's clinical history and related laboratory testing

CASE PRESENTATION

A 27- year-old woman has been admitted with the history of pain in hands, legs and impaired use of both arm and leg since two months. There was no history of substance use or family history of psychiatric, movement, or neurological disorder.

Clinical evaluation revealed Alien Limb Phenomenon involving her both upper and lower extremities. No psychotic or manic symptoms were noted. She required assistance with all activities of daily living and hoisting for transfers. Extra pyramidal features were ongoing.

Brain MRI showed no focal edema or mass effect seen and no features of idiopathic intracranial hypertension. MRI cervical spine revealed less cervical lordosis with mild reversal, suggestive of mild spasm and mild posterior disc buldge at C4/5 and C5/6 levels indenting the thecal sac. No major herniation or neural compression seen. Tarlov's cyst is seen at S2 level measuring about 10mm.Nerve conduction study normal and Electromyogram (EMG) showed neural myotonia.

A battery of investigations were performed including routine hemogram, hepatic, renal function tests, thyroid function tests, and anti-VGKC antibodies. The remaining investigations were also normal except anti-VGKC antibodies (LGI I and CASPR 2 antibody) that turned out to be positive (normal range: 0–100 pmol/L). T.Tegrital 100mg tds and T.Mirtaz 7.5mg HS were started and also treated with Inj. Solumedrol 1gm IV in 100ml NS over 40minutes .No clinical improvement was noted after sequential treatment with pulse steroids (3-day course of 1000mg/day of Solumedrol)

NEUROMYOTONIA

Neuromyotonia is a diverse disorder also known as Isaacs Syndrome, as a result of hyper muscle activity patient have the symptoms of myotonia, myokymia, hyperhidrosis, fasciculation, exercise intolerance, fatigue and other related symptoms.

CAUSES

Acquired

Acquired neuromyotonia is an autoimmune disease in which the immune system malfunctions so that it damages parts of one's own body. Approximately four-hundredth of people have an effect on antibodies to voltage-gated metallic element channels (VGKC's) that affect the points at that the signals from the fiber meet the muscle fiber (neuromuscular junction).

Paraneoplastic

Paraneoplastic are rare disorders that are triggered by an altered immune system response to a neoplasm

Hereditary

SYMPTOMS

- Muscle cramps
- Stiffness
- Myotonia
- Walking difficulties
- Hyperhidrosis (excessive sweating)
- Myokymia (quivering of a muscle)
- Fasciculations (muscle twitching) are most prominent in the calves, legs, trunk, and sometimes the face and neck.
- Fatigue
- Exercise intolerance
- Myoclonic

TYPES

- Chronic
- Monophasic (symptoms that resolve within several years of onset; post infection, post allergic)
- Relapsing Remitting

DIAGNOSIS

- Neurological examination
- Electromyography

- Nerve Conduction Studies
- CT and MRI scan of Brain and spinal cord Voltage-gated potassium channel antibodies
- TSH, ANA ESR, EEG
- Blood investigations

TREATMENT

The goal of treatment is symptomatic as there's no permanent cure nevertheless.

Anticonvulsants

Phenytoin and carbamazepine for significant relief from the stiffness, muscle spasms, and pain associated with neuromyotonia.

Plasma exchange and IVIG treatment might causes an interference with the voltage-dependent potassium channels, one amongst the underlying problems with hyper-excitability in response neuromyotonia.

Botox injections also provide short-term relief.

Immunosuppressants

Prednisone may provide long term relief

PROGNOSIS

Neuromyotonia disorders are now amenable to treatment and their prognoses are good. Most of the patients respond well to the treatment with immunotherapy and steroids thus it provides significant relief of symptoms.

CONCLUSION

This case study review helps us to understand the spectrum of the symptom associated with the peripheral nerve hyper excitability syndrome and disorders of the central nervous system, including Morvan syndrome, epilepsy and limbic encephalitis (LE). It is important that providers recognize the signs and symptoms of this kind of cases and should know to distinguish from other diseases which have similar clinical features. There is currently no consensus as to the efficacy of prophylactic measures in preventing VGKC associated neuromyotonia.

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CONFLICT OF INTEREST

We declare that we have no conflict of interest.

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