Stiff-Limb Syndrome: A Variant of Stiff-Person Syndrome

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INTRODUCTION

Stiff-person syndrome is a spectrum of rare autoimmune neurological disorders resulting in muscle rigidity and episodic muscle spasms that can be widespread or localized such as the Stiff-limb syndrome variant. These debilitating disorders are usually acquired, progressive, fluctuating, and occur in young women aged 30-50. Here we present an atypical case of a 79-year-old female with 3 days of progressive lower extremity weakness and cramps resulting in falls.

METHODS

On physical exam she was found to have hyperreflexia, increased gastrocnemius tone, and spasticity with plantarflexion. Markers of inflammation, infection, toxicity, paraneoplastic syndrome, autoimmunity, and nutritional deficiency were negative in serum, CSF, and on imaging. Furthermore, antibodies were negative for acetylcholine receptor, anti-signal recognition (SRP), ganglioside, HMG-CoA reductase, Lyme, muscle-specific kinase (MuSK), Sjorgens, voltage gated calcium channels, and voltage gated potassium channels.

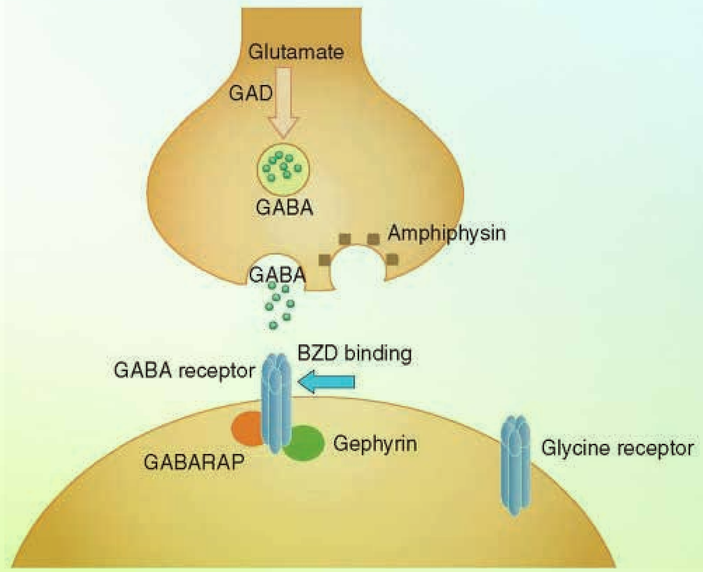

Image from https://neupsykey.com/stiff-person-syndrome-and-peripheral-nerve-and-muscle-hyperexcitability/

RESULTS

The lone positive laboratory marker was antibodies to the Glutamic Acid Decarboxylase (GAD) enzyme at 124 (normal 0-5). The patient was subsequently started on a benzodiazepine with improvement in her symptoms.

CONCLUSION

Stiff-person syndrome and its variants such are Stiff-limb syndrome are suspected to be caused by auto-antibodies to the GAD enzyme. GAD is responsible for converting the principal excitatory neurotransmitter glutamate into the principal inhibitory neurotransmitter gamma-aminobutyric acid (GABA). With increased accumulation of glutamate, clinical findings will include hyperreflexia and increased muscle tone. Benzodiazepines are GABA agonists which can assist in enhancing the GABA neurotransmitter at the GABA receptor resulting in muscle relaxation and therapeutic relief for the patient.

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