JFK Johnson Rehabilitation Institute

A Stiff Finding: Stiff Person Syndrome; A Rare Case Report

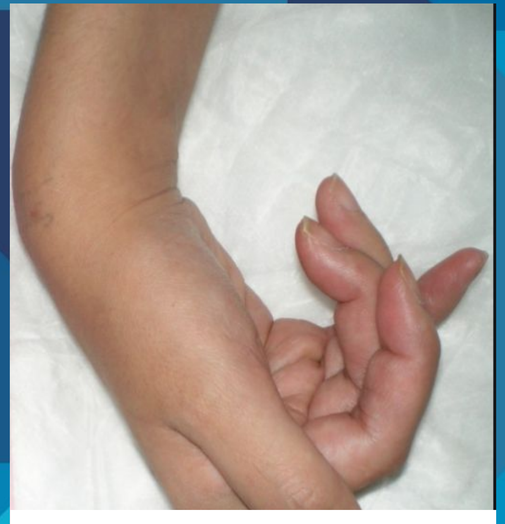
Gabrielle Abissi, MD, David Lee, MD, Beverly Hon, MD, Sara Cuccurullo, MD
Department of Physical Medicine and Rehabilitation, JFK Johnson Rehabilitation Institute, Robert Wood Johnson Medical School - Rutgers University, Edison, NJ

Hackensack Meridian Health JFK Johnson Rehabilitation Institute

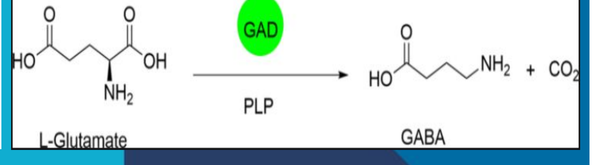
RUTGERS Robert Wood Johnson Medical School

Case Description

A 33-year-old female without any significant history presented with suspected viral encephalitis. She was in her normal state of health and ate Chinese food the day prior. The patient subsequently developed gastrointestinal symptoms, muscle rigidity and stiffness in the right upper and lower extremities. Work-up ensued to identify the cause of her symptoms. She received botulinum toxin injections with some improvement in spasticity. She was maintained on oral baclofen with some further pain relief. The work-up for her neurological condition continued, which included MRI, LP, vEEG and EMG, but the diagnosis was still inconclusive. Ultimately, after all other diagnoses were ruled out, she was diagnosed with Stiff Person Syndrome (SPS) because her glutamic acid decarboxylase (GAD) antibody level was elevated. These antibodies are elevated in certain autoimmune neurological conditions and are specifically linked to SPS.



Initial physical exam presentation: flexed at the metacarpals with MAS of 1+



Discussion

SPS is a rare neurologic disorder of unknown etiology. It is a paraneoplastic neurological syndrome associated with painful spasms and muscle rigidity. In this case, the patient presented with waxing and waning muscle spasticity that required botulinum toxin injections, IVIG, steroids and baclofen for improvement. Glutamic acid decarboxylase (GAD) is an important enzyme that converts glutamate to GABA which works to provide a relaxing effect on the body. In SPS, there are antibodies against this enzyme, thereby causing an excess of glutamate. With glutamate being in excess, there is unopposed contraction of the muscle leading to spasticity.

Conclusion

Although a rare clinical diagnosis, SPS should be on the differential, especially after other neurological conditions are ruled out.
It is important to remember that spasticity is a key feature of this condition. There are many different treatment interventions for spasticity including botulinum toxin injections which work pre-synaptically. Another option is baclofen, which works on the GABA-B receptor. Physiatrists should be aware of the multiple treatment options available for this patient population.

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