Author's response to reviews

Title: Stiff Person Syndrome in the Right Clinical Setting: a case report

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To the Editor:

Thank you for your comments and questions on our recent submission, manuscript ID 6921. Here are the responses. A highlighted version of the manuscript is attached.

The GAD Antibody Assay was sent to the Mayo Clinic in Rochester, MN, where the normal reference range was noted to be # 0.02 nmol/L. It is important to point out that the GAD antibody level is not useful as a marker of disease severity, activity or as a prognostic indicator. However, it is helpful from a diagnostic standpoint, as in this case. The GAD antibody is highly correlated with autoimmune conditions such as diabetes and thyroid conditions. In this case, a thyroid-stimulating hormone was in the normal range, fasting glucose was less than 100 mg/dL and there was no family history of autoimmune disorders. Hemoglobin A1C testing was not performed, as random blood sugars were less than 200 mg/dL during the hospitalization.

The authors agree that the manifestation of stiffness occurring in an arm, as opposed to the legs or thoraco-lumbar spine, accompanied by weakness is a peculiar presentation for SPS. However, we feel that the multidisciplinary approach taken to form this diagnosis calling upon colleagues in neurology to psychiatry helped us consider many other possibilities in the process. Our observations of this as an odd presentation of a clinical condition prompted us to seek publication of this case in the first place. Additionally, we obtained some recent clinical follow-up information on the patient and discovered that her symptoms are currently quite well-controlled on a regimen of diazepam 7.5 mg
orally twice daily and baclofen 20 mg orally every six hours.

Upon an extensive chart review of the medication administration records (as opposed to the physician progress notes), it appears that the patient was never administered diazepam while in the hospital. So, the reported failure of diazepam to relieve arm stiffness/dystonia was in error and has been removed from the case report.

The EMG testing was performed in the following right upper extremity muscles: dorsal interosseous, pronator teres, pectoral radialis, biceps, triceps, deltoid, and opponens pollicis. In all muscles tested, frequent involuntary runs of motor units (continuous motor unit activity) were identified. With limb repositioning, resting activity was able to be studied and revealed absent fibrillations or positive waves. EMG testing of all muscles resulted in normal motor unit morphology and normal recruitment. There was no evidence of myokymic or neuromyotonic discharges.

Although many patients with SPS may not be able to tolerate physical therapy, it was fundamental to the recovery in this case. The authors, again, acknowledge that weakness is not a typical part of Stiff Person Syndrome.

We thank you for your continued consideration of our case report.

Sincerely,

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