BACKGROUND
Stiff person syndrome (SPS) is a rare neurological disorder of unknown etiology that is characterized by progressive rigidity and stiffness of the axial musculature [1-3]. It was first recognized by Moersch and Wolman in 1956 [4].

Clinically, patients with SPS present with rigidity and concurrent waking and waning of muscle spasms. Patients typically experience exacerbating musculoskeletal pain that limits activities of daily living. The axial and 1-2% of muscular atrophy are usually involved, and patients present with an exaggerated upright lumbar lordotic posture. Involvement of the thoracic musculature may cause chest wall restriction and difficulty with breathing. Small muscles of the face can be affected, and spasms of the masseter can cause jaw pain and sluggish. Involvement of the neck muscles may present as neck pain and headaches. Other less commonly affected areas include the proximal musculature in the limbs (stiff limb syndrome), anxious, fear or loud noise can aggravate stiffness or spasms.

PATHOPHYSIOLOGY
SPS is associated with conditions such as vitiligo, pernicious anemia and diabetes mellitus (1,2,6). Observation of oligoclonal and polyclonal IgG in the CSF of SPS provided insight into the autoimmune etiology(3). These autoimmune antibodies in the cerebrospinal fluid are diagnostic of SPS (12) and pathophysiologically important in the ability to impair GABA terminals. The dominant antigen recognized by these antibodies is the GABA-synthesizing enzyme GAD (glutamic acid decarboxylase) (14). Antibodies against epitope -65 of GAD is highly specific for SPS (1,3,8). Circulating antibodies are found in 60% of the patients (8) with SPS, antibodies against epitope -65 of GAD is highly specific for SPS (1,3,8). Observation of oligoclonal and polyclonal IgG in the CSF of SPS provided insight into the autoimmune etiology(3). These autoimmune antibodies in the cerebrospinal fluid are diagnostic of SPS (12) and pathophysiologically important in the ability to impair GABA terminals. The dominant antigen recognized by these antibodies is the GABA-synthesizing enzyme GAD (glutamic acid decarboxylase) (14). Antibodies against epitope -65 of GAD is highly specific for SPS (1,3,8). Circulating antibodies are found in 60% of the patients (8) with SPS, antibodies against epitope -65 of GAD is highly specific for SPS (1,3,8).

Differential Diagnosis; SPS is a diagnosis of exclusion, other differential diagnosis include multiple sclerosis, paraneoplastic myelitis, strychnine poisoning.

Assessment/results: The patient experienced dramatic improvements in his neck stiffness and bruxism after the initial Botox injections. Patient was seen on routine follow up and continued to receive Botox injections into bilateral masseter and neck paraspinal muscles, for maintenance of the beneficial results.

METHODS

Botulinum toxin-A, commercially available as BOTOX (onabotulinumtoxinA 100 & 200 units) vials, was ordered. Standard storage techniques and safety protocols were followed per manufacturer's recommendation. A total of 200 units was used in each session including the above listed agents. Glucocorticoids(12) can also be used as a part of treatment, and were given orally or intrathecally. Other agents used with limited success include tizanidine, valproate, vigabatrin and propofol. Plasmapheresis (13) can be considered in life threatening cases. A multimodality approach includes physical therapy, occupational therapy and behavioral therapy, which can be used adjunctively to improve quality of life.

POINTE OF INJECTION
Figure A and B, showing points of injection with a total of 200 Units was used in each session including injection into the masseter and posterior neck muscles.

CONCLUSION
SPS is a rare neurologic disorder with probable autoimmune etiology which is characterized by generalized rigidity and muscle spasms. The symptoms are relieved with the use of botulinum toxin(A) which act as GABA neuromodulators. The autoimmune etiology hypothesis of SPS has been supported by the relief of the symptoms when patients are provided immunotherapy. Baclofen is another logical therapy for SPS because of its agonistic action on GABA-B receptors. However, for this specific patient, standard medications and the baclofen pump use failed to provide relief specifically for neck stiffness and bruxism. The use of botulinum toxin-A as a localized treatment to the masseter and paraspinal muscles provided effective relief of the rigidity, muscle spasms and pain the patient was experiencing in the facial and neck areas.

REFERENCES