A Case of Stiff Person Syndrome Treated Using Intrathecal Baclofen

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Date of submission: 7th August 2023 Date of acceptance: 29th September 2023 Date of publication: 15th October 2023

Abstract

Introduction: Stiff Person Syndrome is a rare autoimmune neurological disorder with chronic muscle spasms leading to chronic pain and debilitation.

Case Report: We describe a case of a 57-year-old man with the history of Diabetes mellitus type 1 diagnosed 15 years back with high-titer positive anti-GAD antibodies and poorly controlled blood sugar levels under highly dosed insulin treatment.

Conclusion: Intrathecal baclofen showed a marked improvement for the relief of spasticity with the choice of a baclofen pump for similar patients.

Key words: Stiff Person Syndrome, spasm, anti GAD antibodies, intrathecal, baclofen

Introduction

Stiff Person Syndrome (SPS)formally known as Stiff Man Syndrome is a rare central nervous system disease characterized by progressive rigidity of the truncal muscles, superimposed spasms, and an exquisite sensitivity to external stimuli[1]. SPS was first described in 1956 as a new clinical entity by Moersch and Woltman in a series of 14 patients. The course of SPS may vary among several patients. Some present with slow progressive disease while others show acute exacerbations and may change from one phenotype to another. Circulating anti-glutamic acid decarboxylase(anti GAD) antibodies are thought to create a y- aminobutyric acid (GABA) -scarce environment within the body[2]. These antibodies ,although orders of magnitude greater in SPS, are also associated with type 1 Diabetes mellitus[3]. We describe a case of a 57-year-old man with the history of Diabetes mellitus type 1 diagnosed 15 years back with high-titer positive anti-GAD antibodies and poorly controlled blood sugar levels under highly dosed insulin treatment.

Access this article online

Website: https://www.nepjol.info/index.php/NJN

DOI: https://doi.org/10.3126/njn.v20i3.57438

HOW TO CITE

Paudel P, Gurung P, Lama MK, Dabadi S, Shrestha R, Gautam R, Pant B. A Case of Stiff Person Syndrome Treated Using Intrathecal Baclofen. NJNS. 2023;20(3):59-61.

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ISSN: 1813-1948 (Print), 1813-1956 (Online)

Case Description

A 57- year -old male with past history of diabetes, anemia and atrophic gastritis was admitted to our institute with history of chronic muscle spasms and pain in the left lower limb in the last 4 months. The pain manifesting initially in the left knee later progressed to the upper thigh. The onset of illness was 10 years back initially presenting with mild spasms over the ankles, then progressing to the trunk, then generalizing making him be bedridden. Performed anti-GAD antibody serum test showed highly positive titers (696 nmol/L, normal ≤ 0.02nmol/L). In the past, the patient had been treated with muscle relaxants and diazepam bringing partial relief. The patient displayed poorly controlled sugar levels with insulin requirements up to 66 IU daily. In addition, the patient had gastrointestinal symptoms at times. Endoscopy done at the previous center, revealed atrophic gastritis and erosive esophagitis LA grade A. The treatment had been completed with marked improvements. Mild anemia had been observed in the past with hemoglobin levels rising after iron treatment.

Magnetic resonance imaging (MRI) study covering the whole spine including bilateral hip joints, showed a loss of cervical lordosis and multilevel disc dedications at C4-C5 level. A mild central to left paramedic disc protrusion indenting on the anterior thecae sac and causing mild stenosis of the left C4-C5 lateral recess was found, as well as L3-L4 disc desiccation with no definite neural compression. Brain MRI previously performed at another

center, revealed bilateral periventricular and deep white matter ischemic changes. A nerve conduction test done on bilateral lower limbs showed with features suggestive of motor neuropathy of the bilateral tibial nerves.

The patient was admitted and managed conservatively with antispasmodics, analgesics. Following, the patient showed marked improvement in pain and spasms with applied intrathecal baclofen. Therefore, the option for baclofen pump insertion was discussed with the patient.

Discussion

SPS as a mainly clinical diagnosis with chronic muscle spasms and pain, leading to decreased activity and debilitation. Up to 80% of SPS patients have an accompanying endocrinopathy, making other autoimmune disorders frequently related with SPS[4, 5]. Pernicious anemia, autoimmune thyroid disease, and insulin-dependent diabetic mellitus have comorbid frequencies of 5%, 10%, and 35–60%, respectively[2, 6]. The physiology of SPS is associated with GABAergic neurotransmission. Gamma Aminobutyric Acid(GABA) is a primary neurotransmitter found in the brain and produced by insulin producing beta cells of the pancreas. GABA performs by reducing neuronal excitability by inhibiting nerve transmission. Anti-Glutamic Acid Decarboxylase antibody (ANTI-GAD) is directed against Glutamate Decarboxylase (GAD) which synthesizes GABA.

High level of circulating Anti-GAD in SPS hence inhibits GABA and causes progressive muscle spasms [7].

Pathophysiologic similarities exist between SPS and type 1 diabetes, supporting the fact that in up to 80% of individuals with type 1 diabetes anti-GAD can be found at the time of diagnosis, although typically lower in titers than in SPS[4, 8–11].

Here, we present a case of SPS who significantly bettered. From bed-ridden presentation, while admitted to an improvement of support guided walking at discharge after conservative management. Baclofen is a muscle relaxant used in multiple conditions for muscle spasms caused by multiple sclerosis, spinal cord injuries and other neurological diseases. The intrathecal system for baclofen delivers to a specific target at the spinal cord. Candidates for intrathecal baclofen are patients with intractable spasticity and chronic pain uncontrollable by drug therapy only. Advantage of the intrathecal system is the direct administration to the cerebrospinal fluid(CSF), bringing the possibility to use less concentrated dosages of the drug and long-term titration. Treatment goal is to improve quality of life by decreasing spasticity with for improved range of motion and reduced contractures.

Similar study conducted in India showed SPS correlation with anti-GAD[12]. Other studies conducted in Sweden and USA showed that immunoglobulin therapy as effective modality of treatment for SPS[13, 14].

Conclusion

SPS is a rare neurological disorder, an autoimmune disease with high levels of ANTI-GAD antibodies associated with other endocrinopathies leading to progressive spasms and chronic pain with leading to decreased quality of life. Intrathecal Baclofen showed a marked improvement for the relief of spasticity with the choice of a Baclofen pump for similar patients.

Ethics and Consent

Ethical approval of case report is not needed in accordance with the local ethical guideline. Written informed consent was obtained from the patient to include the clinical details.

Conflicts of Interest

None of the authors have potential conflicts of interest to be disclosed.

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