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Case Report: Stiff Person Syndrome (SPS)

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Abstract:- Stiff Person Syndrome (SPS), a rare autoimmune disorder, which is not widely studied is illustrated in this case report. The patient, a 52-year-old female with Stiff Person Syndrome, presents with spasticity, muscle aches, and weakness. Elevated levels of anti-glutamic acid decarboxylase (GAD) antibodies were detected, confirming the diagnosis of Stiff Person Syndrome. The patient is being treated with Diazepam, Baclofen, IVIG, and Gabapentin. This cocktail of medication has alleviated her pain and stiffness. She is currently responding well to the administered treatment. Early diagnosis and interdisciplinary treatment are necessary for Stiff Person Syndrome to improve patient outcomes and raise quality of life. The relevance of examining SPS as a possible diagnosis in patients who appear with increasing muscle stiffness and spasms is highlighted by this case report, which also underlines the need for more studies to fully comprehend this complicated condition.

Keywords:- *Stiff Person Syndrome, Autoimmune, Muscular Rigidity, Anti GAD Antibodies, IVIG, Gabapentin.*

I. INTRODUCTION

SPS is a rare syndrome of progressive rigidity of axial and proximal limb muscles (alpha and gamma motor neurons) with extreme lumbar lordosis and muscle hypotrophy. On a background of continuous muscle contraction, intense muscle spasms are superimposed. Gait - slow with stiff legs. Clinical criteria - diagnosis: Limb and axial stiffness (thoracolumbar and abdominal, clinical and electrophysiological confirmation of *co-contraction of agonist and antagonists' muscles, episodic spasms superimposed on chronic stiffness.* SPS is associated with auto immune disorders such as type 1 diabetes, myasthenia gravis, pernicious anemia and vitiligo. Anti-bodies of GAD and other antigens are present. SPS results from impairment of descending suprasegmental pathways leading to inhibition of GABA synthesis.

Paraneoplastic SPS has been associated with various cancers including breast cancer.

II. CASE

- Age: 52 years
- Gender: Female
- Chief Complaints: spasticity, muscle aches and weakness

➢ *History of Presenting Illness:*

Patient is 52 years old female with past medical history of alopecia, tarlov cyst and stiff person syndrome presented to neurology clinic with complaint of spasticity, muscle aches and weakness since past few weeks. She reported that her symptoms initially started as occasional muscle spasms and stiffness in her lower back. However, over time, the symptoms worsened and spread to involve her upper body, leading to significant impairment in daily activities.

➢ *Medical History:*

Patient suffered from alopecia and Tarlov cyst disease but had not experienced any similar symptoms in the past. She denied any recent infections, trauma, or exposure to toxins. She had no family history of neurological disorders.

➢ *Clinical Findings:*

On physical examination, patient exhibited severe stiffness and muscle spasms.

General/constitutional: well; cooperative;

- Eyes: conjunctivae: normal; normal; pupils: equal, round and reactive; normal; Optic disc: normal; sclera: white; cornea: normal; normal; lens: normal; Extraocular muscles: intact; retina: red reflex present; visual acuity: normal;
- Neck: appearance: normal; palpation: supple; thyroid: smooth and non-tender; Range of Motion: Normal;
- Respiratory: auscultation: clear;
- Cardio/vascular: auscultation: rhythm - regular. no murmurs, rubs or gallops;
- GI: abdomen: *auscultation*: normal; palpation: non-tender;
- Extremities: peripheral edema: none

Neuro – mental status – alert, head and neck – flexion strength and tone – 5+, extension strength and tone – 5+, language intact, Cranial nerves – intact, *R arm strength and tone* – deltoid 5+, biceps 5+, triceps 5+, wrist flexion 5+, wrist extension 5+, finger flexion 5+, finger extension 5+, finger abduction – FDI and ADM 5+, thumb abduction 5+, L arm *strength and tone* – deltoid 5+, biceps 5+, triceps 5+, wrist flexion 5+, wrist extension 5+, finger flexion 5+, finger extension 5+, finger abduction – FDI and ADM 5+, thumb abduction 5+, *R leg strength and tone* – hip flexion 5+, hip extension 5+, hip abduction 5+, hip adduction 5+, knee flexion 5+, knee extension 5+, ankle dorsiflexion 5+, ankle plantar

flexion 5+, ankle eversion 5+, ankle inversion 5+, toe flexion and extension 5+, *L leg strength and tone* – hip flexion 5+, hip extension 5+, hip abduction 5+, hip adduction 5+, knee flexion 5+, knee extension 5+, ankle dorsiflexion 5+, ankle plantar flexion 5+, ankle eversion 5+, ankle inversion 5+, toe flexion and extension 5+, sensation of pain (sharp): *head & neck*: intact; *trunk*: intact; *L arm*: intact; *R arm*: intact; *L leg*: intact; *R leg*: intact; sensation of touch (dull): *head & neck*: intact; *trunk*: intact; *L arm*: intact; *R arm*: intact; *L leg*: intact; *R leg*: intact; proprioception & vibration: *Proprioception*: Normal; *Vibration Sense*: Normal; deep tendon reflexes: *bicep(s)*: 2+; *triceps(s)*: 2+; *patellar*: 2+; *Achilles*: 2+; *plantar/Babinski*: L down going toes, R down going toes; coordination: *heel-to-shin*: intact; *finger-to-nose*: intact; *Rapid Alternating Movement(s)*: intact; *Rhomberg*: negative; gait and station: intact; Spasticity in arms, shoulder, and legs.

➢ *DIAGNOSIS:*

Given the clinical presentation, further investigations were carried out to establish a diagnosis. The following tests were performed:

- *Blood tests*: Routine blood investigations-complete blood count, liver and kidney function tests, thyroid function tests, vitamin B12 levels, and autoimmune markers, were all within normal limits.
- *Electromyography (EMG)*: continuous motor unit activity and increased unstructured muscle activity during both active and relaxed states, indicative of hyperexcitability of the motor neurons.
- *Magnetic Resonance Imaging (MRI)*: Brain and spinal cord MRI were performed, and no significant findings were observed.
- *Antibody testing*: Elevated levels of anti - GAD antibodies.

According to clinical presentation, EMG findings, and high levels of anti-GAD antibodies, a diagnosis of Stiff Person Syndrome (SPS) was established. (G25.82)

➢ *TREATMENT & FOLLOW UPS:*

Patient was started on diazepam 10mg, pregabalin 75 mg, Botox 200 units (IM injection in 31 spots over paraspinal trapezius and arms, legs for spasticity), gabapentin 300 mg capsule and dantrolene 25 mg. Physical therapy and counseling were also recommended to help manage patient's symptoms and improve her quality of life.

The patient was scheduled for regular follow-up visits to monitor the response to treatment and adjust medication dosages as needed. Long-term management involved a multidisciplinary approach, including neurology, physical therapy, and psychological support, to address the patient's physical and emotional well-being.

III. DISCUSSION

Stiff Person Syndrome (SPS), a rare neurological disorder indicated by progressive muscle stiffness and spasms, mainly affecting axial muscles. It is believed to be an autoimmune-mediated disorder, where the body's immune system mistakenly attacks and impairs the function of inhibitory neurons in the spinal cord and brainstem.

The diagnosis of SPS is predominantly clinical, supported by characteristic EMG findings and presence of anti-GAD antibodies. Treatment aims to control symptoms, manage muscle stiffness, and improve the patient's quality of life. Medications, physical therapy, and psychological support play crucial roles in the long-term management of SPS.

This case report highlights the importance of recognizing the clinical features of Stiff Person Syndrome and conducting appropriate investigations to confirm the diagnosis. Early diagnosis and multidisciplinary management are vital in providing optimal care and improving outcomes for patients with SPS.

IV. CONCLUSION

A rare neurological condition known as Stiff Person Syndrome (SPS) is characterized by increased muscle stiffness and spasms. A 52-year-old female patient with a characteristic SPS clinical presentation, including significant muscle stiffness and spasms affecting the paraspinal muscles, abdomen, and lower extremities, was described in this case report. The discovery of increased anti-glutamic acid decarboxylase (GAD) antibodies and results of hyperexcitability of motor neurons during electromyography corroborated the diagnosis.

A multi-modal strategy is used to manage SPS, including symptomatic immunomodulatory drugs, muscle relaxant therapy, physical therapy, and psychosocial support. To reduce symptoms, enhance functional capacities, and enhance the patient's quality of life, early diagnosis and treatments are essential.

This case report highlights the significance of identifying the clinical symptoms of SPS, carrying out necessary investigations, creating a diagnosis and treatment plan.

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