e-ISSN: 0976-822X, p-ISSN:2961-6042

Available online on http://www.ijcpr.com/

International Journal of Current Pharmaceutical Review and Research 2026; 18(2); 782-786

Original Research Article

The Unstoppable Spasm: A Rare Case of Stiff Person Syndrome

Chetan Patil[1], Nahush Patil[2], Ashish Shirodkar[3], Jitendra Kodilkar[4], Jeetendra Singh[5]

[1]MD General Medicine, Associate Professor, Department of Medicine, Dr Vasantrao Pawar Medical College, Hospital and Research Centre, Nashik, MH, India

[2]DM Neurology, Associate Professor, Department of Medicine, Dr Vasantrao Pawar Medical College, Hospital and Research Centre, Nashik, MH, India

[3]MD General Medicine Resident, Department of Medicine, Dr Vasantrao Pawar Medical College, Hospital and Research Centre, Nashik, MH, India

[4]Professor and Head - Department of Medicine, Dr Vasantrao Pawar Medical College, Hospital and Research Centre, Nashik, MH, India

[5]Professor and Head, Dept of Pharmacology, Maharashtra Postgraduate Institute of Medical Education and Research, Maharashtra University of Health Sciences, Nashik, MH, India

Received: 12-12-2025 / Revised: 13-01-2026 / Accepted: 15-02-2026

Corresponding Author: Dr. Jitendra Kodilkar

Conflict of interest: Nil

Abstract:

Background: Stiff-person spectrum disorders (SPSD) are rare autoimmune neurological conditions characterized by progressive muscle stiffness, painful spasms, and exaggerated startle responses, often leading to severe functional impairment. Diagnosis is frequently delayed due to clinical overlap with other neuromuscular disorders.

Case presentation: We report a 45-year-old male with an acute history of progressive axial stiffness, lower-limb rigidity, and episodic painful spasms causing gait disturbance and recurrent falls. Neurological examination revealed marked paraspinal and proximal limb rigidity with preserved sensation and reflexes. Routine laboratory investigations were unremarkable. Serum anti–glutamic acid decarboxylase (GAD65) antibodies were strongly positive. Needle electromyography demonstrated continuous motor unit activity at rest, confirming SPSD. Brain and spinal imaging excluded structural pathology.

Management and outcome: The patient was initiated on benzodiazepines and baclofen, which provided partial symptomatic relief. Given the autoimmune profile and progressive disability, a short course of steroids (dexamethasone) was administered, resulting in a significant reduction in spasm frequency and improved gait at 3-month follow-up.

Conclusion: This case highlights the importance of considering SPSD in adults presenting with progressive axial rigidity and spasms. Early recognition, supported by Electromyography (EMG) and GAD65 antibody testing, facilitates timely initiation of symptomatic and immunomodulatory therapies, which can markedly improve mobility and quality of life.

Keywords: Stiff Person Syndrome, Anti-GAD Antibodies, Electromyography, GABAergic Agents, Baclofen, Lorazepam, muscle stiffness.

Key Messages:

- Stiff Person Syndrome is a rare immune-mediated neurological disorder often misdiagnosed due to overlapping psychiatric and neurological features.
- Anti-GAD antibody testing and EMG are pivotal for accurate and timely diagnosis.

Early GABAergic therapy with physiotherapy ensures substantial recovery, even in resource-limited settings.

DOI: 10.25258/ijcpr.18.2.128

Introduction

Stiff Person Syndrome (SPS) is a rare, immune-mediated disorder of the central nervous system characterized by progressive muscle rigidity and stimulus-triggered painful spasms, most often affecting the axial and proximal limb muscles. First described in 1956 by Moersch and Woltman in 14 patients, it was initially termed "stiff-man syndrome" and is now more appropriately referred to as Stiff Person Syndrome or Moersch–Woltman Syndrome [1].

The classic phenotype, accounting for 70–80% of cases, is strongly associated with antibodies against glutamic acid decarboxylase (anti-GAD), an enzyme essential for γ-aminobutyric acid (GABA) synthesis [2]. SPS typically develops insidiously and progresses to severe disability, with mortality

reported in advanced stages. It is frequently accompanied by autoimmune disorders such as type 1 diabetes mellitus, autoimmune thyroiditis, pernicious anemia, celiac disease, and vitiligo [1]. Variants include stiff limb syndrome, jerky SPS, cerebellar and dystonic forms, and SPS associated with epilepsy [1]. A paraneoplastic subtype, representing 5–10% of cases, is linked to malignancies such as breast, colon, thyroid, or lung cancers, as well as lymphomas, and may precede tumor diagnosis [2]. Progressive encephalomyelitis with rigidity and myoclonus (PERM), part of the SPS spectrum, is characterized by diffuse rigidity, myoclonus, and autonomic instability [1].

The pathogenesis involves B-cell–mediated autoimmunity against inhibitory GABAergic pathways. Anti-GAD65 antibodies are detected in up to 80% of patients, while antibodies to glycine receptor (GlyR), GABA(A) receptor–associated protein (GABARAP), and dipeptidyl- peptidase-like protein-6 (DPPX) are also implicated [1,3,4]. Paraneoplastic cases are more often associated with amphiphysin and, less commonly, gephyrin antibodies [1, 2].

Diagnosis is primarily clinical, supported by antibody assays and electromyographic studies5. However, overlapping features with psychiatric, neuromuscular, and functional disorders often delay recognition. Early diagnosis and timely treatment with benzodiazepines, baclofen, or immunotherapy are critical for improving outcomes [1]. Case reports remain essential for raising clinical awareness, refining differential diagnoses, and guiding management. Here, we present a rare case of SPS with distinctive features, highlighting diagnostic challenges and the importance of early intervention.

Objective:

To report a rare case of anti-GAD–positive Stiff Person Syndrome, emphasizing clinical features, diagnosis, management, and outcomes.

Methods

Patient Information: A 45-year-old Indian male presented to the emergency department with progressive generalized muscle stiffness and impaired mobility involving all four limbs. He reported recurrent, stimulus-sensitive spasms—initially affecting the paraspinal and facial muscles and later spreading to the extremities—often precipitated by emotional stress. These episodes were associated with facial weakness, difficulty rising from a supine position, and inability to ambulate.

He had no history of similar complaints, comorbidities, substance use, long-term medications, fever, weight loss, sensory disturbances, loss of consciousness, or autonomic dysfunction. Family history was unremarkable.

Clinical Findings: The patient was alert, oriented, and cooperative. Examination revealed generalized spasticity causing severe functional limitation and lumbar hyper lordosis. Gait could not be assessed.

Neurological evaluation showed increased tone in the paraspinal and facial muscles with cervical rigidity. Deep tendon reflexes were brisk (grade 3) with spread, while superficial reflexes were preserved. Cranial nerves were intact except for mild dysarthria without dysphagia. Muscle bulk and strength were preserved, with no involuntary movements. Sensory and coordination assessments were limited due to marked rigidity.

Timeline

- **Day 0:** Onset of generalized stiffness and spasms with inability to ambulate.
- **Day 1:** Hospital admission; physiotherapy assessment—maximal assistance required for bed mobility, unable to stand or walk.
- **Hospital course (20 days):** Neurology-led management with gradual improvement.
- **Post-discharge:** Weekly follow-ups for 1 month, then biweekly; progressive recovery of ambulation

Diagnostic Assessment

- **Laboratory tests:** Serum electrolytes, renal and liver function, glucose, ESR, CRP, CK, CPK-MB, TSH, and WBC counts—all normal.
- **Neuroimaging:**
    - CT spine – mild degenerative changes without cord compression.
    - CT brain – mild cerebral atrophy with right bitemporal hypodense area suggestive of old infarct (likely incidental).
- **Electromyography (EMG):** Continuous spontaneous motor unit activity across examined muscles.
- **Serology:** Anti-GAD antibodies positive, confirming Stiff Person Syndrome (SPS).

Differential diagnoses including seizure disorder and tetanus were excluded. Further antibody testing was limited by financial constraints.

Therapeutic Intervention

- **Initial therapy:** Diazepam (5 mg/day) provided minimal benefit and was substituted with lorazepam.
- **Definitive therapy:** Lorazepam, titrated to 5 mg four times daily, achieved sustained spasm control.

Baclofen (10 mg three times daily) was added, resulting in marked improvement.

- **Other measures:**
    - Levetiracetam (500 mg twice daily) initiated empirically, discontinued after SPS confirmation.
    - Intravenous dexamethasone (8 mg twice daily) given initially, then withdrawn.
    - Daily physiotherapy for mobility training and spasticity management.

By discharge, the patient could roll in bed, sit independently, achieve full limb range of motion, and ambulate with assistance.

Follow-Up and Outcomes: The patient was discharged on oral baclofen and a tapering course of methylprednisolone. At follow-up, he demonstrated substantial functional recovery—progressing from bed-bound status to independent standing and assisted ambulation within weeks. No residual disability was noted. He was advised to continue physiotherapy and symptomatic use of benzodiazepines & baclofen. Follow-up visits were maintained weekly for 1 month, then biweekly.

Results

Table 1: Summary of Clinical Features, Diagnostic Findings, Management, and Outcomes

Clinical Features	Diagnostic Findings	Management	Outcomes
Progressive axial and limb rigidity	Positive anti-GAD65 antibodies	Lorazepam (5 mg four times daily)	At discharge: Independent rolling/sitting, assisted ambulation
Painful, stimulus-sensitive spasms	EMG: Continuous motor unit activity at rest	Baclofen (10 mg three times daily)	At 3-month follow-up: Independent standing, minimal residual disability
Lumbar hyperlordosis	Normal laboratory tests (electrolytes, renal, liver, ESR, CRP, CK, CPK-MB, TSH, WBC)	Intravenous dexamethasone (8 mg twice daily, then tapered)	Full limb range of motion achieved
Gait disturbance and recurrent falls	CT spine: Mild degenerative changes without cord compression	Daily physiotherapy for mobility and spasticity management	Progressive functional recovery
Mild dysarthria	CT brain: Mild cerebral atrophy, incidental right bitemporal hypodense area (old infarct)	Levetiracetam (500 mg twice daily, discontinued after SPS confirmation)	No residual disability noted

The patient showed progressive functional improvement during hospitalization. At admission, he was bed-bound and unable to roll or sit independently. By discharge, he could roll and sit independently, ambulate with assistance, and demonstrated full limb range of motion. At 3- month follow-up, he was able to stand independently with assisted ambulation and had minimal residual disability.

Discussion

Stiff Person Syndrome (SPS) is a rare, immune-mediated neurological disorder characterized by progressive axial and proximal muscle rigidity, painful stimulus-sensitive spasms, and functional impairment. It is most commonly associated with antibodies against glutamic acid decarboxylase (anti-GAD), which disrupt γ-aminobutyric acid (GABA)-mediated inhibitory pathways, leading to motor neuron hyperexcitability [1]. Our case demonstrates the classical anti- GAD–positive phenotype, with paraspinal and facial rigidity, spasms triggered by emotional stress, and significant mobility limitation.

SPS remains difficult to diagnose due to its rarity and overlapping features with psychiatric, functional, and neuromuscular disorders. In this patient, continuous spontaneous motor unit activity on EMG combined with anti-GAD antibody positivity confirmed the diagnosis. Other antibodies—such as anti-GlyR, anti-GABARAP, and anti-amphiphysin—have been reported in SPS variants but were not assessed due to resource limitations[6]. Alternative diagnoses, including seizure disorder, tetanus, and structural spinal pathology, were systematically excluded based on clinical evaluation and investigations.

Table 2: Differential Diagnosis of Stiff Person Syndrome

Condition	Key Features	Diagnostic Tests	SPS Distinguishing Features
Tetanus	Trismus, opisthotonos, recent injury or wound	Clinical history, toxin testing	Anti-GAD positivity, continuous EMG activity, stimulus-sensitive spasms
Seizure Disorder	Episodic loss of consciousness, convulsive movements	EEG, response to anticonvulsants	No loss of consciousness, positive anti-GAD antibodies, EMG findings
Parkinsonism	Tremor, bradykinesia, rigidity without spasms	Dopamine response, brain imaging (e.g., DaTscan)	Stimulus-sensitive spasms, anti-GAD positivity, no tremor
Multiple Sclerosis	Sensory deficits, relapsing-remitting course, optic neuritis	MRI brain/spine, cerebrospinal fluid analysis	No sensory deficits, positive anti-GAD antibodies, EMG continuous activity
Psychogenic Movement Disorder	Inconsistent symptoms, distractibility, variable presentation	Normal EMG, psychological evaluation	Consistent rigidity/spasms, anti-GAD positivity, EMG findings
Spinal Cord Pathology	Sensory loss, weakness, bowel/bladder dysfunction	MRI spine, electrophysiological studies	No sensory loss, normal spine MRI, anti-GAD positivity

The autoimmune hypothesis of SPS is supported by the presence of anti-GAD antibodies, which are frequently associated with other autoimmune conditions such as type 1 diabetes mellitus. Anti-GAD antibodies reduce central GABA levels, causing motor neuron hyperexcitability that manifests as rigidity and stimulus-sensitive spasms. In a retrospective cohort, Tsiortou et al. [7] highlighted that Anti-GlyR antibodies are linked to SPS spectrum disorders, including progressive encephalomyelitis with rigidity and myoclonus (PERM) [7].

Management of SPS involves both symptomatic relief and immunomodulation. GABAergic agents—including benzodiazepines, baclofen, gabapentin, and pregabalin—enhance inhibitory neurotransmission and provide rapid symptom control. In our patient, diazepam provided minimal relief, whereas lorazepam combined with baclofen led to marked improvement in mobility. Immunomodulatory therapies, such as corticosteroids, intravenous immunoglobulin (IVIG), plasmapheresis, and rituximab, are reserved for refractory cases and target the underlying autoimmune process [8,9]. Intensive physiotherapy was essential for restoring mobility and preventing secondary musculoskeletal complications.

As reported by Bose et al. [10], SPS carries a variable prognosis. Delayed recognition often results in severe functional impairment, psychological burden, and misdiagnosis as a psychiatric or functional disorder [10]. Early clinical suspicion and prompt initiation of therapy, as demonstrated in this case, were crucial for favorable outcomes. The patient progressed from bed-bound status to independent standing and assisted ambulation within weeks, highlighting the potential for recovery even in resource-limited settings.

This case emphasizes the importance of clinical vigilance in patients presenting with unexplained rigidity and spasms. Multidisciplinary management—neurology, physiotherapy, and pharmacotherapy—optimizes functional recovery. Anti-GAD antibody testing and EMG remain central to diagnosis, while additional evaluation to exclude paraneoplastic causes should be considered when clinically indicated [6,7]. Even in resource-constrained environments, timely recognition and intervention can produce substantial functional improvement and reduce long- term disability.

This case illustrates the classical anti-GAD–positive SPS phenotype and underscores the need for high clinical suspicion, thorough differential diagnosis, and multidisciplinary management. Symptomatic GABAergic therapy, combined with immunomodulation and physiotherapy, can achieve significant recovery even when advanced antibody testing or IVIG is not immediately available.

Conclusion

Stiff Person Syndrome (SPS) is a rare, immune-mediated neurological disorder that poses significant diagnostic and therapeutic challenges. This case underscores the importance of early recognition, as delayed identification often leads to severe disability, psychological distress, and misdiagnosis as psychiatric or functional illness. Anti-GAD antibody testing and EMG remain central to diagnostic confirmation, while systematic exclusion of alternative and paraneoplastic causes is essential. Symptomatic treatment with GABAergic agents, combined with immunomodulatory therapy and physiotherapy, can yield substantial functional recovery. Importantly, even in resource-limited settings, timely multidisciplinary management can markedly improve outcomes and reduce long-term

morbidity. Sustained clinical vigilance is vital to ensure prompt diagnosis and optimize patient care.

Informed Consent: Written informed consent was obtained from the patient prior to all evaluations and interventions.

Data Availability: No data were used to support the current study.

References

1. Muranova A, Shanina E. Stiff Person Syndrome. [Updated 2023 Jul 10]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2025 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK573078/
2. Rakocevic G, Floeter MK. Autoimmune stiff person syndrome and related myelopathies: understanding of electrophysiological and immunological processes. Muscle & nerve. 2012 May;45(5):623-34.
3. Alexopoulos H, Dalakas MC. Immunology of stiff person syndrome and other GAD-associated neurological disorders. Expert review of clinical immunology. 2013 Nov 1;9(11):1043-53.
4. Ciccotto G, Blaya M, Kelley RE. Stiff person syndrome. Neurologic clinics. 2013 Feb 1;31(1):319-28.
5. El-Abassi R, Soliman MY, Villemarette-Pittman N, England JD. SPS: understanding the complexity. Journal of the neurological sciences. 2019 Sep 15; 404:137-49.
6. Sarva H, Deik A, Ullah A, Severt WL. Clinical spectrum of stiff person syndrome: a review of recent reports. Tremor Other Hyperkinet Mov (N Y). 2016 Mar 4; 6:340.
7. Tsiortou P, Alexopoulos H, Dalakas MC. GAD antibody-spectrum disorders: progress in clinical phenotypes, immunopathogenesis and therapeutic interventions. Therapeutic advances in neurological disorders. 2021 Mar; 14:17562864211003486.
8. Ortiz JF, Ghani MR, Cox ÁM, Tambo W, Bashir F, Wirth M, Moya G. Stiff-person syndrome: a treatment update and new directions. Cureus. 2020 Dec 9;12(12).
9. Dalakas MC, Fujii M, Li M, Lutfi B, Kyhos J, McElroy B. High-dose intravenous immune globulin for stiff-person syndrome. New England Journal of Medicine. 2001 Dec 27;345(26):1870-6.
10. Bose S, Jacob S. Stiff-person syndrome. Pract Neurol. 2025;25(1):6–17. doi:10.1136/pn-2023-003974. Available from: http://pn.bmj.com/content/25/1/6.abstract