

# A rare presentation of stiff-person syndrome with a nonspecific focal myositis: A case report

Arij Ezzouhour Yahyaoui<sup>1</sup>, Sameh Sayhi<sup>2</sup>, Nour Elhouda Guediche<sup>2</sup>, Zakaria Saied<sup>3</sup>, Bilel Arfaoui<sup>4</sup>,  
Faïda Ajili<sup>1,2</sup>, Ines Bedoui<sup>5</sup>, Samia Ben Sassi<sup>3</sup>, Nadia Ben Abdelhafidh<sup>1</sup>

<sup>1</sup>Tunisia Autoimmune Diseases Research Unit UR17DN02, Military Hospital of Tunis, Tunis, Tunisia

<sup>2</sup>Internal Medicine, Military Hospital of Tunis, Tunis, Tunisia

<sup>3</sup>Neurology Department, Mongi Ben Hmida National Institute of Neurology, Tunis, Tunisia

<sup>4</sup>Internal Medicine, Military Hospital of Bizerte, Tunis, Tunisia

<sup>5</sup>Neurology Department, Military Hospital of Tunis, Tunis, Tunisia

## ABSTRACT

**Introduction.** The stiff-person syndrome (SPS) is a rare disease whose incidence is estimated at approximately 1 in a million individuals in the general population. Diagnosis relies on a combination of clinical, immunological, and electromyographic items. We present the case of a patient diagnosed with the idiopathic SPS, initially misdiagnosed as focal paravertebral myositis.

**Case presentation.** A 36-year-old patient was referred to us for the investigation of subacute dorsal myalgias. The patient reported a severe torticollis seven months ago, which resolved spontaneously after one month, and some episodes of back stiffness. On examination, the patient was afebrile, had hyperlordosis with a paravertebral contracture and tenderness. Neurological and cognitive examinations were normal. C-reactive Protein was at 137 mg/l. The rest of laboratory investigations, including creatine phosphokinase (CPK), were within normal range. Spinal MRI revealed T2 hyperintensity in the semispinalis muscle, erector spinae muscle, trapezius muscle, as well as the intervertebral muscles. Therefore, focal paravertebral myositis was suspected. The electromyogram (EMG) revealed the presence of a continuous motor unit activity in agonist and antagonist muscles, suggestive of stiff-person syndrome. Antinuclear antibodies, anti-glutamic acid decarboxylase 65 and onconeural antibodies were negative. Analysis of the cerebrospinal fluid, including anti-glutamic acid decarboxylase 65 test, was normal. We noticed that the paravertebral contracture became less noticeable when the patient was commenced on diazepam. Diagnosis of SPS was established according to Dalakas criteria. Investigations for an underlying neoplasm, were normal. Associated autoimmune disorders have been ruled out. The MRI description was rather explained by the continuous contraction of the affected muscles. Treatment included diazepam, baclofen, intravenous immunoglobulin and corticosteroids. The patient showed important signs of improvement.

**Conclusion.** SPS is a rare condition whose diagnosis can be delayed. Recognizing and managing SPS as early as possible is crucial. It is based on clinical reasoning, imaging, biological features and EMG.

**Keywords:** stiff-person syndrome, anti-glutamic acid decarboxylase 65, myositis

## INTRODUCTION

Stiff-person syndrome (SPS), previously known as Moersch-Woltman Syndrome or stiff man syndrome, is a rare neurological disorder characterized by increased muscular activity due to impaired inhibitory GABAergic neurotransmission [1,2]. Despite its rarity, SPS presents significant diagnostic

and therapeutic challenges, requiring a comprehensive understanding of its clinical spectrum and immunological subtypes. This article reports the case of a patient diagnosed with the idiopathic SPS and explores the various facets of SPS, including its clinical manifestations, immunological associations, diagnostic criteria, differential diagnosis, and evolu-

*Corresponding author:*

Arij Ezzouhour Yahyaoui

E-mail: erij.yh@yahoo.com

*Article History:*

Received: 20 September 2024

Accepted: 29 September 2024

ing treatment paradigms. By shedding light on these aspects, we aim to highlight that internists should be able to identify SPS despite an unusual and sometimes confusing neurological presentation. An etiological investigation is necessary, particularly the research of an underlying neoplasm.

## CASE REPORT

A 36-year-old patient was referred to us for the investigation of dorsal myositis, manifested by subacute dorsal paravertebral myalgias, which exacerbate with the slightest movement. Visual analogue scale was at 9/10. The patient reported a severe torticollis seven months ago, which resolved spontaneously after one month, and some episodes of back stiffness. On examination, the patient was afebrile, had hyperlordosis with a paravertebral contracture, as seen in Figure 1. The deep tendon reflexes were symmetrical. He had no motor or sensory deficit. No coordination disorder was noticed. Cognitive examination was normal. He had paravertebral tenderness with no skin lesion. Creatine phosphokinase, lactate dehydrogenase and transaminases were within normal range. C-reactive Protein was at 137 mg/l. Spinal MRI revealed T2 hyperintensity in the semispinalis muscle, erector spinae muscle, trapezius muscle, as well as the intervertebral muscles

Therefore, the patient was referred to our department for suspicion of a focal myositis. The electromyogram (EMG) revealed the presence of a continuous motor unit activity in agonist and antagonist muscles, suggestive of stiff-person syndrome. Diagnosis of SPS was established according to Dalakas criteria [3,4].

Blood culture, tuberculosis skin test, antinuclear antibodies, anti-glutamic acid decarboxylase 65 and onconeural antibodies were negative. Analysis of the cerebrospinal fluid, including anti-glutamic acid decarboxylase 65 test, was normal. Investigations for an underlying neoplasm were normal. Associated autoimmune disorders, especially type 1 diabetes, autoimmune thyroiditis and coeliac disease have been ruled out echocardiogram and thoracic-abdominal-pelvic scanner were normal.

The MRI description was rather explained by the continuous contraction of the affected muscles.

The patient was commenced on diazepam and baclofen. Diazepam was initially administered intravenous and the dose was progressively increased from 10 to 30 mg per day. Then, it was switched into oral administration. Intravenous immunoglobulin was commenced on the third day at 400 mg/kg daily for 5 days. On the 7<sup>th</sup> day of treatment, Visual analogue scale progressed to 4/10 and on examination, paravertebral contracture decreased remarkably.



**FIGURE 1.** Hyperlordosis with paravertebral contraction

The patient was discharged with a prescription of 10 mg of diazepam given thrice a day, baclofen 10 mg twice a day and 20 mg of prednisone daily. The symptoms responded well and the patient showed important signs of improvement, paravertebral contracture disappeared and the patient was able to get back to his normal activities.

## DISCUSSION

Moersch-Woltman Syndrome (MWS), also known as stiff man syndrome, was first described by Moersch and Woltman in 1956 [1,2]. It is a rare, progressive condition characterized by heightened muscle activity resulting from reduced inhibitory control within the brain and spinal cord.

The estimated prevalence of the condition is 1–2 cases per million, with an incidence of approximate-

ly 1 case per million annually. Symptoms most commonly begin to appear between the ages of 20 and 50. The syndrome shows a clear preference for women, occurring two to three times more often in females than in males [3,5-7]. However, it's noteworthy that some individuals diagnosed with stiff man syndrome present in pediatric age groups or are adult males, indicating variability in the clinical presentation of the condition [8].

Moersch and Woltman, along with findings from 13 additional cases, documented a 49-year-old male who exhibited progressive stiffness in the neck, shoulders, and upper back, accompanied by episodic painful muscle spasms and walking difficulties. Since then, numerous cases with similar clinical features have been reported [1,2].

The term "stiff man syndrome" has been recently updated to the more gender-neutral "stiff-person syndrome" (SPS). This change was first suggested by Asher in 1958 [9]. The shift in terminology gained support after Blum and Jankovic [10] noted that nearly 20 out of 84 reported cases between 1967 and 1991 involved female patients.

The onset of classic SPS is often gradual. The clinical presentation is varied, including progressive muscle stiffness, primarily affecting the axial muscles, and intermittent muscle spasms, which can be triggered by stimuli such as loud sounds, bright lights, or emotional stress. Patients frequently experience ongoing muscle discomfort and may develop abnormal postures, most notably lumbar hyperlordosis. The widespread rigidity associated with SPS can cause breathing difficulties and reduced exercise capacity. Furthermore, limited chest and abdominal expansion may result in early satiety. Muscle spasms are sometimes preceded by jerky contractions (myoclonus) and typically subside gradually [5,11].

SPS is also marked by psychiatric symptoms like depression and anxiety, alongside other neurological signs such as horizontal and vertical supranuclear gaze palsy, nystagmus, exaggerated reflexes, and episodes of paroxysmal dysautonomic crises [5,12-15]. Severe dysautonomic were reported, including cricopharyngeal muscle spasm and hypoxemic respiratory failure [5].

The frequent occurrence of gastrointestinal symptoms and transit abnormalities among individuals with SPS indicates that gut motility is susceptible to similar disorders as the skeletal neuromuscular system in this condition, the mechanism remains unclear [16-18]. The association between SPS and biliary dyskinesia has been reported in the literature, few articles available on this topic. Consequently, elucidating the correlation between these two conditions poses a challenge due to the scarcity of research findings [7].

Stiff-person syndrome spectrum disorders (SPSSDs) have broadened to encompass a range of conditions exhibiting signs and symptoms similar to those observed in traditional SPD [19].

In 1999, Brown et al. introduced the "Diagnostic Criteria for Classic Stiff-Person Syndrome," which categorized SPS into two primary subtypes: classic SPS, which occurs without encephalomyelitis, and SPS plus, which includes cases with encephalomyelitis, such as progressive encephalomyelitis with rigidity and myoclonus (PERM), jerking stiff man syndrome, and stiff limb syndrome (SLS) [20].

Currently, it is categorized into classical and variant SPS [5]. Variant SPS includes focal or segmented SPS, SPS with spasms, PERM, SPS with ataxia, epilepsy, etc and paraneoplastic variant [5,21-23].

The possibility of an immunological cause was suggested by the high prevalence of diabetes (reported in up to 35% of some studies) and the presence of other autoimmune conditions, including vitiligo, celiac disease, rheumatologic disorders, and thyrogastric disorders [2,3,24]. Currently, three major immunological subtypes are identified: (1) glutamic acid decarboxylase 65 (GAD65)-positive SPS associated with other autoimmune conditions; (2) anti-amphiphysin-positive SPS associated with tumors; and (3) seronegative idiopathic SPS [25]. It has been reported that the prevalence of anti-GAD65 antibodies can range from 80% to 98% among patients diagnosed with SPS [22,26]. It is linked to classic SPS [19,27]. Additionally, other autoantigens associated with SPSSD include glycine receptors (associated with PERM) [28-30], amphiphysin (associated with cancers) [31], GABAA receptors [32] and its related protein GABAA receptor-associated protein (GABARAP) [33], dipeptidyl-peptidase-like protein-6 (DPPX), and Zic4 (linked to small-cell lung cancer) [34].

Some researchers have raised concerns about the cryptogenic category, proposing that it may encompass autoimmune cases where known antibodies exist at undetectable levels or where antibodies have not yet been identified [35]. Remarkably, unlike autoimmune and paraneoplastic cases, the majority of cryptogenic cases primarily involved males. Notably and similarly to our patient, most cryptogenic reported cases showed symptomatic amelioration [35].

Like our patient, another case was reported where the patient presented a non-specific focal myositis secondary to sustained muscle contraction. The myositis aspect in imaging was rather explained by the continuous contraction of the affected muscles. That case was also cryptogenic [36].

The identification of anti-GAD antibodies prompted a reevaluation of the diagnostic criteria for stiff-person syndrome (SPS), which were first es-

established by Gordon et al. in 1967. In 2009, Dalakas proposed updated diagnostic criteria, stating that a diagnosis of SPS requires the following elements: 1) stiffness in the axial muscles, especially in the abdomen and thoracolumbar paraspinals, resulting in hyperlordosis; 2) painful spasms triggered by tactile or auditory stimuli; 3) electromyographic findings indicating continuous motor unit activity in both agonist and antagonist muscles; 4) the absence of other neurological signs that could indicate a different diagnosis; and 5) positive serological results confirmed through immunocytochemistry, Western blot, or radioimmunoassay [3,37].

The differential diagnosis for stiff-person syndrome (SPS) includes a wide variety of conditions, such as myelopathy, myopathy, Isaac's syndrome, Parkinson's disease, atypical Parkinsonian syndromes, primary lateral sclerosis, ankylosing spondylitis, neuroleptic malignant syndrome, serotonin syndrome, hereditary or tropical spastic paraparesis, spinal interneuronitis with rigidity, dystonia, neuromyotonia, and tetanus [23,24,35].

Paraneoplastic variants represent less than 10% of all patients with SPS [38]. Anti-amphiphysin antibodies are the most common markers of this variant [39]. Literature research indicates that breast cancer is the most commonly associated carcinoma with SPSSDs, followed by lung cancer and lymphoma. Additionally, classic SPS appears to be the most prevalent subtype of SPSSD, followed by stiff limb syndrome and progressive encephalomyelitis with rigidity and myoclonus [8,19,22]. Some reviews illustrated the intricate relationship between cancers, autoantigens, and SPSSDs, indicating that new insights in this area are continually emerging worldwide [8].

The predominant and rational therapeutic strategy involves a combination of GABA-enhancing medications and immunotherapy. This approach is justified by the distinct mechanisms. Dalakas et al. suggested in 2023 step-by-step therapies in SPS Based on disease pathophysiology. This strategy includes GABA-enhancing drugs, Immunotherapies, a combination of both of these therapies and supportive physical therapies. GABA-enhancing drugs are the first-line therapies, as they enhance GABAergic inhibitory neurotransmission, attenuate cortical hy-

perexcitability, or elevate central nervous system (CNS) GABA levels [4,40]. Our patient responded well to diazepam (GABA receptor-binding benzodiazepines) and baclofen (GABA receptor-binding benzodiazepines, centrally acting antispasmodic drug). Immunotherapies should be initiated promptly when first-line therapies utilizing GABA-enhancing medications do not yield complete efficacy. They include intravenous immunoglobulins (IVIg), plasmapheresis and rituximab. Our patient presented a great evolution after IVIg administration.

Some series and case reports supported the use of corticosteroids, azathioprine and mycophenolate mofetil [41-46].

Other studies showed complete remission in patients started on Efgartigimod alfa [42].

As for eculizumab, it showed inefficacy and the treatment was complicated by meningitis in one patient, whereas symptomatic control on eculizumab alone was noted in another patient [45].

Autologous hematopoietic stem cell transplantation (auto-HSCT) may present an alternative promising therapeutic approach, according to some series [47-50].

## CONCLUSION

SPS poses diagnostic and therapeutic challenges, necessitating a comprehensive understanding of its clinical spectrum, immunological subtypes, and evolving treatment paradigms. Further research is warranted to elucidate underlying mechanisms and optimize personalized management strategies for improved patient outcomes.

### *Data availability:*

All data underlying the results are available as part of the article and no additional source data are required.

### *Consent:*

Written informed consent from the patient for publication of his clinical details and clinical images is obtained.

### *Competing interests:*

No competing interests were disclosed.

*Ethics committee approval* was deemed not necessary in our institutions for case reports.

*Financial support:* none declared

## REFERENCES

1. Moersch FP, Woltman HW. Progressive fluctuating muscular rigidity and spasm ("stiff-man" syndrome); report of a case and some observations in 13 other cases. *Proc Staff Meet Mayo Clin.* 1956 Jul;31(15):421-7.
2. Ali F, Rowley M, Jayakrishnan B, Teuber S, Gershwin ME, Mackay IR. Stiff-person syndrome (SPS) and anti-GAD-related CNS degenerations: protean additions to the autoimmune central neuropathies. *J Autoimmun.* 2011 Sep;37(2):79-87. doi: 10.1016/j.jaut.2011.05.005.
3. Dalakas MC. Stiff person syndrome: advances in pathogenesis and therapeutic interventions. *Curr Treat Options Neurol.* 2009 Mar;11(2):102-10. doi: 10.1007/s11940-009-0013-9.
4. Dalakas MC. Therapies in Stiff-Person Syndrome: Advances and Future Prospects Based on Disease Pathophysiology. *Neurol* -

- Neuroimmunol Neuroinflammation*. 2023 May;10(3):e200109. doi: 10.1212/NXI.000000000200109.
5. Baizabal-Carvallo JF, Jankovic J. Stiff-person syndrome: insights into a complex autoimmune disorder. *J Neurol Neurosurg Psychiatry*. 2015 Aug;86(8):840-8. doi: 10.1136/jnnp-2014-309201.
  6. Mancinelli R, Franchitto A, Glaser S, Meng F, Onori P, Demorrows S, et al. GABA induces the differentiation of small into large cholangiocytes by activation of Ca(2+) /CaMK I-dependent adenylyl cyclase 8. *Hepatology*. 2013 Jul;58(1):251-63. doi: 10.1002/hep.26308.
  7. Louis-Jean S, Agrawal N, Chaudhry S, Mazer A. Biliary Dyskinesia in Stiff Person Syndrome: An Association Between Reduced GABA Production and Gastroenteric Dysmotility. *J Community Hosp Intern Med Perspect*. 13(5):94-6. doi: 10.55729/2000-9666.1239.
  8. Peng Y, Yang H, Xue Y hui, Chen Q, Jin H, Liu S, et al. An update on malignant tumor-related stiff person syndrome spectrum disorders: clinical mechanism, treatment, and outcomes. *Front Neurol*. 2023 Oct;14:1209302. doi: 10.3389/fneur.2023.1209302.
  9. Asher R. A Woman with the Stiff-man Syndrome. *Br Med J*. 1958 Feb;1(5065):265-6. doi: 10.1136/bmj.1.5065.265.
  10. Blum P, Jankovic J. Stiff-person syndrome: An autoimmune disease. *Mov Disord*. 1991;6(1):12-20. doi: 10.1002/mds.870060104.
  11. Meinck HM, Ricker K, Hülser PJ, Solimena M. Stiff man syndrome: neurophysiological findings in eight patients. *J Neurol*. 1995 Feb;242(3):134-42. doi: 10.1007/BF00936885.
  12. Ortiz JF, Ghani MR, Morillo Cox Á, Tambo W, Bashir F, Wirth M, et al. Stiff-Person Syndrome: A Treatment Update and New Directions. *Cureus*. 12(12):e11995. doi: 10.7759/cureus.11995.
  13. Oskarsson B, Pelak V, Quan D, Hall D, Foster C, Galetta S. Stiff eyes in stiff-person syndrome. *Neurology*. 2008 Jul;71(5):378-80. doi: 10.1212/01.wnl.0000319725.22925.b4.
  14. Jr E, Jc H. Eye movement abnormalities in stiff person syndrome. *Neurology* [Internet]. 11 August 2005 [cited 5 nov 2023];65(9). Available on: <https://pubmed.ncbi.nlm.nih.gov/16275836/>.
  15. Mitsumoto H, Schwartzman MJ, Estes ML, Chou SM, La Franchise EF, De Camilli P, et al. Sudden death and paroxysmal autonomic dysfunction in stiff-man syndrome. *J Neurol*. 1991 Apr;238(2):91-6. doi: 10.1007/BF00315688.
  16. Koshorek J, Wang Y, Maldonado DP, Reyes-Mantilla MI, Obando D, Balshi A, et al. The many faces of gastrointestinal dysfunction in stiff person syndrome spectrum disorders. *Front Neurol*. 2023 Oct 6;14:1273256. doi: 10.3389/fneur.2023.1273256.
  17. Auteri M, Zizzo MG, Serio R. GABA and GABA receptors in the gastrointestinal tract: from motility to inflammation. *Pharmacol Res*. 2015 Mar 1;93:11-21. doi: 10.1016/j.phrs.2014.12.001.
  18. Aggarwal S, Ahuja V, Paul J. Dysregulation of GABAergic Signalling Contributes in the Pathogenesis of Diarrhea-predominant Irritable Bowel Syndrome. *J Neurogastroenterol Motil*. 2018 Jul;24(3):422. doi: 10.5056/jnm17100.
  19. Newsome SD, Johnson T. Stiff person syndrome spectrum disorders; more than meets the eye. *J Neuroimmunol*. 2022 Aug 15;369:577915. doi: 10.1016/j.jneuroim.2022.577915.
  20. Brown P, Marsden CD. The stiff man and stiff man plus syndromes. *J Neurol*. 1999 Aug 1;246(8):648-52. doi: 10.1007/s004150050425.
  21. Barker RA, Revesz T, Thom M, Marsden CD, Brown P. Review of 23 patients affected by the stiff man syndrome: clinical subdivision into stiff trunk (man) syndrome, stiff limb syndrome, and progressive encephalomyelitis with rigidity. *J Neurol Neurosurg Psychiatry*. 1998 Nov 1;65(5):633-40. doi: 10.1136/jnnp.65.5.633.
  22. McKeon A. Stiff-Man Syndrome and Variants: Clinical Course, Treatments, and Outcomes. *Arch Neurol*. 2012 Feb 1;69(2):230. doi: 10.1001/archneurol.2011.991.
  23. Mas N, Saiz A, Leite MI, Waters P, Baron M, Castaño D, et al. Antiglycine-receptor encephalomyelitis with rigidity. *J Neurol Neurosurg Psychiatry*. 2011 Dec 1;82(12):1399-401. doi: 10.1136/jnnp.2010.229104.
  24. Ehler E, Latta J, Mandysová P, Havlasová J, Mrklovský M. Stiff-person syndrome following tick-borne meningoencephalitis. *Acta Medica (Hradec Kralove)*. 2011;54(4):170-4. doi: 10.14712/18059694.2016.44.
  25. Buechner S, Florio I, Capone L. Stiff Person Syndrome: A Rare Neurological Disorder, Heterogeneous in Clinical Presentation and Not Easy to Treat. *Case Rep Neurol Med*. 2015;2015:278065. doi: 10.1155/2015/278065.
  26. Walikonis JE, Lennon VA. Radioimmunoassay for Glutamic Acid Decarboxylase (GAD65) Autoantibodies as a Diagnostic Aid for Stiff-Man Syndrome and a Correlate of Susceptibility to Type 1 Diabetes Mellitus. *Mayo Clin Proc*. 1998 Dec 1;73(12):1161-6. doi: 10.4065/73.12.1161.
  27. Martinez-Hernandez E, Ariño H, McKeon A, Iizuka T, Titulaer MJ, Simabukuro MM, et al. Clinical and Immunologic Investigations in Patients With Stiff-Person Spectrum Disorder. *JAMA Neurol*. 2016 Jun 1;73(6):714-20. doi: 10.1001/jamaneurol.2016.0133.
  28. Graus F, Saiz A, Dalmau J. GAD antibodies in neurological disorders — insights and challenges. *Nat Rev Neurol*. 2020 Jul;16(7):353-65. doi: 10.1038/s41582-020-0359-x.
  29. Crisp SJ, Balint B, Vincent A. Redefining progressive encephalomyelitis with rigidity and myoclonus after the discovery of antibodies to glycine receptors. *Curr Opin Neurol*. 2017 Jun;30(3):310-6. doi: 10.1097/WCO.0000000000000450.
  30. Hutchinson M, Waters P, McHugh J, Gorman G, O'Riordan S, Connolly S, et al. Progressive Encephalomyelitis, Rigidity, and Myoclonus: A Novel Glycine Receptor Antibody. *Neurology*. 2008 Oct 14;71(16):1291-2. doi: 10.1212/01.wnl.0000327606.50322.f0.
  31. Bataller L, Wade DF, Fuller GN, Rosenfeld MR, Dalmau J. Cerebellar degeneration and autoimmunity to zinc-finger proteins of the cerebellum. *Neurology*. 2002 Dec 24;59(12):1985-7. doi: 10.1212/01.wnl.0000038352.01415.ce.
  32. Petit-Pedrol M, Armangue T, Peng X, Bataller L, Cellucci T, Davis R, et al. Encephalitis with refractory seizures, status epilepticus, and antibodies to the GABAA receptor: a case series, characterisation of the antigen, and analysis of the effects of antibodies. *Lancet Neurol*. 2014 Mar;13(3):276-86. doi: 10.1016/S1474-4422(13)70299-0.
  33. Raju R, Rakocevic G, Chen Z, Hoehn G, Semino-Mora C, Shi W, et al. Autoimmunity to GABAA-receptor-associated protein in stiff-person syndrome. *Brain*. 2006 Dec 1;129(12):3270-6. doi: 10.1093/brain/awl245.
  34. Bernardo F, Rebordão L, Rêgo A, Machado S, Passos J, Costa C, et al. Stiff person spectrum disorders: An illustrative case series of their phenotypic and antibody diversity. *J Neuroimmunol* [Internet]. 15 April 2020 [cited 5 nov 2023];341. Available on: [https://www.jni-journal.com/article/S0165-5728\(19\)30636-8/fulltext](https://www.jni-journal.com/article/S0165-5728(19)30636-8/fulltext).
  35. Sarva H, Deik A, Ullah A, Severt WL. Clinical Spectrum of Stiff Person Syndrome: A Review of Recent Reports. *Tremor Hyperkinetic Mov*. 2016 Mar 4;6(0):340. doi: 10.7916/D85M65GD.
  36. No SW, Im IK, Kim DH. Stiff Person Syndrome With Evidence of Nonspecific Focal Myositis Secondary to Sustained Muscle Contraction: A Case Report. *PM R*. 2018 Dec;10(12):1426-30. doi: 10.1016/j.pmrj.2018.04.007.
  37. Gordon EE, Januszko DM, Kaufman L. A critical survey of stiff-man syndrome. *Am J Med*. 1967 Apr 1;42(4):582-99. doi: 10.1016/0002-9343(67)90057-5.
  38. Murinson BB, Guarnaccia JB. Stiff-person syndrome with amphiphysin antibodies: distinctive features of a rare disease. *Neurology*. 2008 Dec 9;71(24):1955-8. doi: 10.1212/01.wnl.0000327342.58936.e0.
  39. Pittock SJ, Lucchinetti CF, Parisi JE, Benarroch EE, Mokri B, Stephan CL, et al. Amphiphysin autoimmunity: paraneoplastic accompaniments. *Ann Neurol*. 2005 Jul;58(1):96-107. doi: 10.1002/ana.20529.
  40. Saigal R, Goyal L, Yadav R, Agrawal A, Mital P, Patel B. Stiff Person Syndrome. *J Assoc Physicians India*. 2015 Aug;63(8):81-2.
  41. Eisenhut K, Faber J, Engels D, Gerhards R, Lewerenz J, Doppler K, et al. Early Neuroaxonal Damage in Neurologic Disorders Associated With GAD65 Antibodies. *Neurol - Neuroimmunol*

- Neuroinflammation* [Internet]. 1 jan 2024 [cited 19 nov 2023];11(1). Available on: <https://nn.neurology.org/content/11/1/e200176>.
42. Di Stefano V, Alonge P, Rini N, Militello M, Lupica A, Torrente A, Brighina F. Efgartigimod beyond myasthenia gravis: the role of FcRn-targeting therapies in stiff-person syndrome. *J Neurol*. 2024 Jan;271(1):254-262. doi: 10.1007/s00415-023-11970-1. Epub 2023 Sep 8.
  43. Perri M, Pellegrini D, Uribe Roca C, Gonzalez F, Buero A, Chimondeguy D, Bruetman JE. Síndrome de la persona rígida asociado a timoma [Stiff person syndrome associated with thymoma]. *Medicina (B Aires)*. 2023;83(4):626-630. Spanish.
  44. Moustafa A, Alsamman MA, Fernandes D. Rare Association of Autoimmune Limbic Encephalitis and Stiff Person Syndrome. *R I Med J* (2013). 2023 Jul 5;106(6):7-9.
  45. McCombe JA, Klassen BT, Flanagan EP, Teener JW, Zekeridou A, Pittock SJ, McKeon A. Eculizumab for the treatment of glycine receptor antibody associated stiff-person syndrome. *J Neurol*. 2023 Sep;270(9):4555-4557. doi: 10.1007/s00415-023-11777-0.
  46. Rauschenberger V, von Wardenburg N, Schaefer N, Ogino K, Hirata H, Lillesaar C, et al. Glycine Receptor Autoantibodies Impair Receptor Function and Induce Motor Dysfunction. *Ann Neurol*. 2020;88(3):544-61. doi: 10.1002/ana.25832.
  47. Sanders S, Bredeson C, Pringle CE, Martin L, Allan D, Bence-Bruckler I, et al. Autologous Stem Cell Transplantation for Stiff Person Syndrome: Two Cases From the Ottawa Blood and Marrow Transplant Program. *JAMA Neurol*. 2014 Oct 1;71(10):1296-9. doi: 10.1001/jamaneurol.2014.1297.
  48. Kass-Iliyya L, Snowden JA, Thorpe A, Jessop H, Chantry AD, Sarrigiannis PG, et al. Autologous haematopoietic stem cell transplantation for refractory stiff-person syndrome: the UK experience. *J Neurol*. 2021;268(1):265-75. doi: 10.1007/s00415-020-10054-8.
  49. Burt RK, Balabanov R, Han X, Quigley K, Arnautovic I, Helenowski I, et al. Autologous Hematopoietic Stem Cell Transplantation for Stiff-Person Spectrum Disorder: A Clinical Trial. *Neurology*. 2021 Feb 9;96(6):e817-30. doi: 10.1212/WNL.00000000000011338.
  50. Jaime-Pérez JC, Meléndez-Flores JD, Ramos-Dávila EM, González-Treviño M, Gómez-Almaguer D. Hematopoietic stem cell transplantation for uncommon immune-mediated neurological disorders: A literature review. *Cytotherapy*. 2022 Jul 1;24(7):676-85. doi: 10.1016/j.jcyt.2021.12.006.