Proprioceptive Stimuli as a New Type of Trigger for Epilepsy in Stiff Person Syndrome

Author(s):
Paula Panos Basterra, MD1; Florentino Nombela, MD, PhD1; Pablo Iriarte, MD1; Celia Romero Del Rincon, MD1; Jaime Alonso Maroto, MD1; Sonia Quintas, MD1

Corresponding Author:
Paula Panos Basterra, panospaula@gmail.com

Affiliation Information for All Authors: 1. Neurology Department, Hospital Universitario de la Princesa

Equal Author Contribution:

Contributions:
Paula Panos Basterra: Drafting/revision of the manuscript for content, including medical writing for content
Florentino Nombela: Drafting/revision of the manuscript for content, including medical writing for content; Major role in the acquisition of data
Pablo Iriarte: Major role in the acquisition of data
Celia Romero Del Rincon: Major role in the acquisition of data
Jaime Alonso Maroto: Major role in the acquisition of data
Sonia Quintas: Drafting/revision of the manuscript for content, including medical writing for content; Study concept or design

Neurology® Published Ahead of Print articles have been peer reviewed and accepted for publication. This manuscript will be published in its final form after copyediting, page composition, and review of proofs. Errors that could affect the content may be corrected during these processes. Videos, if applicable, will be available when the article is published in its final form.
A healthy 74-year-old woman was diagnosed with Stiff-Person (SP) syndrome (encephalopathy and stiff leg) with amphiphysin autoantibodies and T1N1M0 breast cancer (HER2+,PR-,ER-). She received immunotherapy (2 high-dose steroid cycles and 7 plasmapheresis sessions), followed by right mastectomy, axillary chain lymphadenectomy and adjuvant chemotherapy (letrozole, trastuzumab, cyclophosphamide), with poor clinical response (ECOG 4). After discharge, she presented to the emergency room with seizures induced by knee flexion of the stiff leg (video 1). EEG showed epileptic seizures initiating in the left temporal lobe (image 1). Reduced presynaptic GABAergic inhibition could be the underlying mechanism of the core signs of amphiphysin-SP, which might explain the presence of epilepsy in these patients(1). However, no reflex proprioceptive seizures had been previously reported in SP.

WNL-2022-201118_vid1 --http://links.lww.com/WNL/C311

References

Figure 1. Electroencephalogram Shows Epileptiform Discharges Initiating in Left Temporal Region

[For comp: no legend]

Video 1.
Passive knee flexion of the left leg produced clonic movements that started in the contralateral limb, and quickly evolved to bilateral tonic-clonic activity. No other triggers were found. The patient remained aware during the whole seizure. After 3mg of intravenous diazepam, epileptic activity ceased.
Proprioceptive Stimuli as a New Type of Trigger for Epilepsy in Stiff Person Syndrome
Paula Panos Basterra, Florentino Nombela, Pablo Iriarte, et al.
*Neurology* published online August 31, 2022
DOI 10.1212/WNL.0000000000201196

This information is current as of August 31, 2022