Teaching NeuroImages: Severe spasms resembling status dystonicus as an unusual presentation of stiff-person syndrome

Daniel G. Di Luca, MD, Sagari Bette, MD, Carlos Singer, MD, and Corneliu Luca, MD, PhD

Neurology® 2019;92:e748. doi:10.1212/WNL.0000000000006925

Correspondence

Dr. Di Luca
Daniel.garbindiluc@
jhsmiami.org

MORE ONLINE

Video

→Teaching slides

links.lww.com/WNL/A810

A 20-year-old man presented with frequent episodes of generalized rigidity. He had an 18-month history of painful muscle spasms, diaphoresis, and falls. These episodes were associated with anxiety related to pain and the bizarre, unpredictable nature of the symptoms. The patient was admitted with severe tonic spasm episodes lasting for up to 5 minutes, which were initially thought secondary to status dystonicus (video).

CSF analysis revealed an elevated GAD-65 titer (116) and a diagnosis of stiff-person syndrome was made.^{1,2} The episodes were refractory to pharmacologic therapy, requiring use of paralyzing agents and IV immunoglobulin with marked improvement in his condition.^{1,2}

Author contributions

D.G. Di Luca and S. Bette: review of the literature and drafting the case report. C. Luca: patient consent and video footage. C. Luca and C. Singer: critical review of the manuscript and final approval of the version to be published.

Study funding

No targeted funding reported.

Disclosure

The authors report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.

References

1. Espay AJ, Chen R. Rigidity and spasms from autoimmune encephalomyelopathies: stiff-person syndrome. *Muscle Nerve* 2006;34:677–690.
2. Baizabal-Carvallo JK, Jankovic J. Stiff-person syndrome: insights into a complex autoimmune disorder. *J Neurol Neurosurg Psychiatry* 2015;86:840–848.

Neurology®

Teaching NeuroImages: Severe spasms resembling status dystonicus as an unusual presentation of stiff-person syndrome

Daniel G. Di Luca, Sagari Bette, Carlos Singer, et al.

Neurology 2019;92:e748

DOI 10.1212/WNL.0000000000006925

This information is current as of February 11, 2019

Updated Information & Services

including high resolution figures, can be found at:
<http://n.neurology.org/content/92/7/e748.full>

References

This article cites 2 articles, 1 of which you can access for free at:
<http://n.neurology.org/content/92/7/e748.full#ref-list-1>

Permissions & Licensing

Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at:
http://www.neurology.org/about/about_the_journal#permissions

Reprints

Information about ordering reprints can be found online:
<http://n.neurology.org/subscribers/advertise>

Neurology® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright © 2019 American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.