Disputes & Debates: Editors' Choice

Steven Galetta, MD, FAAN, Section Editor

Editors' note: Video NeuroImages: Paraneoplastic spinal myoclonus associated with Caspr2 antibodies

In their case report, Hines et al. reported spinal myoclonus as a novel neuromuscular manifestation of Caspr2 paraneoplastic disease. Caspr2 antibodies have been previously identified in conjunction with limbic encephalitis and neuromyotonia, but not in cases of spinal myoclonus. To make the diagnosis of spinal myoclonus, as Drs. Tipton, van Gerpen, and Chen appropriately acknowledge, there must be simultaneous and stereotypical activation from at least 4 muscle groups with contiguous spinal cord innervation. Given the novelty of the findings, alternative diagnoses—such as stiff-person syndrome (SPS) and the triple flexion response—must be definitively excluded. Hines et al. verify that the 2 most common SPS antibodies, GAD65 and amphiphysin, were absent from the serum, and that EMG confirmed 5 muscle groups were involved in this stereotypical response. In addition, treatment of the underlying malignancy led to resolution of motor symptoms, further implicating a pathogenic paraneoplastic process. Although the final electrographic diagnosis remains contested, this case highlights the clinical diversity of paraneoplastic disorders and the importance of accurate characterization of clinical and EMG findings in neurologic disease.

James E. Siegler III, MD, and Steven Galetta, MD
Neurology® 2019;92:302. doi:10.1212/WNL.0000000000006869

Reader response: Video NeuroImages: Paraneoplastic spinal myoclonus associated with Caspr2 antibodies

Philip W. Tipton (Jacksonville, FL), Jay A. van Gerpen (Jacksonville, FL), and Robert Chen (Toronto, Canada)
Neurology® 2019;92:302–303. doi:10.1212/WNL.0000000000006870

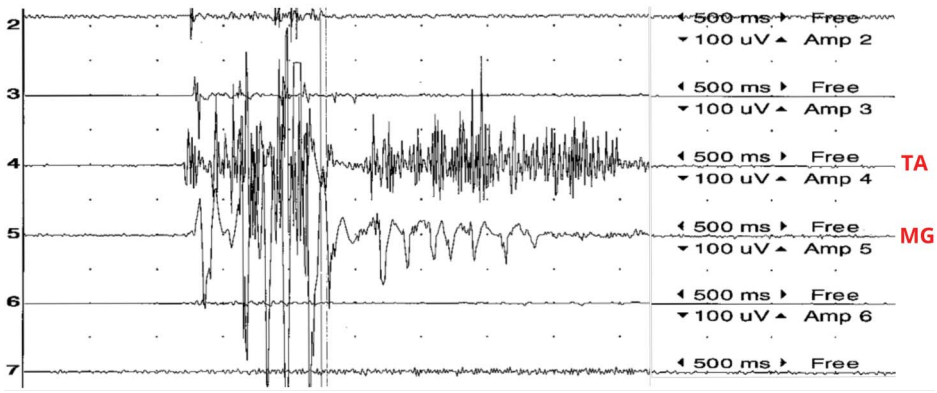
Hines et al.[1] presented spinal myoclonus secondary to Caspr2 antibodies, including a video of intermittent right-leg jerks and an EMG tracing of activity in the rectus abdominis, purporting to show a unique EMG pattern.

We disagree with these conclusions. The clinical phenomenon is most likely a triple flexion response, an exteroceptive reflex indicative of spinal cord corticospinal tract hyperexcitability. This may occur spontaneously or be elicited by various stimuli, and is caused by multiple etiologies, including stiff-person syndrome (SPS).[2] This well-described phenomenon in 2 muscles is illustrated by a surface EMG (sEMG) example (figure), which is what the figure presented by Hines et al.[1] also depicts.

The accurate diagnosis of spinal myoclonus is predicated upon obtaining simultaneous sEMG recordings from at least 4 muscles to ascertain whether the pattern of muscle activation is stereotypical. Thus, definitive conclusions regarding the proper classification of the hyperkinesia in the patient presented by Hines et al. cannot be drawn, though we suspect the patient had an SPS variant (i.e., stiff-limb syndrome, a known paraneoplastic accompaniment).

Author disclosures are available upon request (journal@neurology.org).

Figure Triple flexion response after plantar stimulation in a patient with myelopathy secondary to multiple sclerosis



MG = medial gastrocnemius; TA = tibialis anterior.

1. Hines H, Murray NM, Ahmad S, Jaradeh S, Gold CA. Video NeuroImages: Paraneoplastic spinal myoclonus associated with Caspr2 antibodies. Neurology 2018;90:660–661.
2. Espay AJ, Chen R. Rigidity and spasms from autoimmune encephalomyelopathies: stiff-person syndrome. Muscle Nerve 2006;34:677–690.

Author response: Video NeuroImages: Paraneoplastic spinal myoclonus associated with Caspr2 antibodies

Harrison Hines (San Francisco), Nick M. Murray (Woodside), Sarah Ahmad (Mountain View), Safwan Jaradeh (Palo Alto), and Carl A. Gold (Stanford)
Neurology® 2019;92:303. doi:10.1212/WNL.0000000000006871

We thank Tipton et al. for their comments on our case study describing a man with spinal myoclonus associated with Caspr2 antibodies.[1] We agree that determining the etiology of hyperkinesia of the lower limb in the setting of malignancy is difficult; however, we maintain our conclusions based on additional data we were unable to include in the case report due to word and format limitations.

Surface EMG studies performed in 5 muscle groups all demonstrated hyperkinesia during leg jerks that could be traced back to a spinal origin (a phenomenon that is not consistent with a triple flexion response). Serum studies were negative for GAD and amphiphysin antibodies, and the patient exhibited none of the clinical symptoms of stiff-person syndrome, such as gait abnormalities. Further work is necessary to better characterize the clinical variations associated with Caspr2 antibodies.

1. Hines H, Murray NM, Ahmad S, Jaradeh S, Gold CA. Video NeuroImages: Paraneoplastic spinal myoclonus associated with Caspr2 antibodies. Neurology 2018;90:660–661.

Author disclosures are available upon request (journal@neurology.org).

Reader response: Video NeuroImages: Paraneoplastic spinal myoclonus associated with Caspr2 antibodies

Philip W. Tipton (Jacksonville, FL), Jay A. van Gerpen (Jacksonville, FL), and Robert Chen (Toronto, Canada)
Neurology® 2019;92:304. doi:10.1212/WNL.0000000000006872

We appreciate the response by Hines et al.[1] to our skepticism that their video NeuroImage was accurately interpreted, either clinically or neurophysiologically. However, we do not find support for their conclusion that their case represents a unique form of spinal myoclonus.

The gravamen of our critique is that their patient's clinical phenomenon is an intermittent, spontaneous triple-flexion response. This exteroceptive reflex of spinal cord origin may be induced by processes that uncouple the lateral corticospinal tract from interneuron inhibition. To our knowledge, it has never been classified as a form of spinal myoclonus.[2] Thus, we do not doubt the other 5 muscles they recorded from could be "traced back to spinal origin." Moreover, their presented waveform, allegedly representing a "unique form of spinal myoclonus," has the same morphology as our example of a typical exteroceptive response.

As surface EMG increasingly bolsters the accuracy of diagnosing hyperkinesias, it is imperative that the literature reflect correct surface EMG methods and interpretations.

1. Hines H, Murray NM, Ahmad S, Jaradeh S, Gold CA. Video NeuroImages: Paraneoplastic spinal myoclonus associated with Caspr2 antibodies. Neurology 2018;90:660–661.
2. Espay AJ, Chen R. Rigidity and spasms from autoimmune encephalomyelopathies: stiff-person syndrome. Muscle Nerve 2006;34:677–690.

CORRECTION

Brain death, the determination of brain death, and member guidance for brain death accommodation requests

AAN position statement

Neurology® 2019;92:304. doi:10.1212/WNL.0000000000007117

In the Special Article "Brain death, the determination of brain death, and member guidance for brain death accommodation requests: AAN position statement," by J.A. Russell et al.,[1] there were errors in the byline and the inclusion of a coinvestigator list in the version published online on January 2, 2019. The byline should not have included the wording "on behalf of the Brain Death Working Group." In addition, the inclusion of a coinvestigators list was in error and the link was removed from the Footnote. The linked document was also removed. These corrections published with the final print and subsequent posting of the article on January 29, 2019. The authors regret the errors.

Reference

1. Russell JA, Epstein LG, Greer DM, Kirschen M, Rubin MA, Lewis A. Brain death, the determination of brain death, and member guidance for brain death accommodation requests: AAN position statement. Neurology 2019;92:228–232.

Author disclosures are available upon request (journal@neurology.org).

Brain death, the determination of brain death, and member guidance for brain death accommodation requests: AAN position statement

Neurology 2019;92;304
DOI 10.1212/WNL.0000000000007117