Response to Letter to the Editor: Spinomedullary Weston Hurst Syndrome After COVID-19 and Influenza Co-Infection

Spencer K. Hutto, MD1 and Nagagopal Venna, MD, MRCP2

Keywords
Weston hurst disease, acute hemorrhagic leukoencephalitis, COVID-19, influenza, case report

We thank Dr Finsterer for his interest in our report and the Editor for providing us with a forum to discuss some of the key concepts of the paper at greater depth.

Given the novelty of neuroinflammatory complications associated with COVID-19, we agree that alternative etiologies should be thoroughly sought and refuted. For this very reason, we were careful to highlight in Tables 1 and 2 the extensive laboratory testing of serum and CSF specimens that occurred in this case, including all of the potential alternative diagnoses suggested in the letter.1 In the initial phase of the diagnostic work-up, we were equally concerned about the possibility for sarcoidosis and malignancy (lymphoma included) as well as non-inflammatory myelopathies (including vascular myelopathies, such spinal cord infarction and dural arteriovenous fistulas), which sparked our decision to obtain whole body PET CT and MRA of spinal blood vessels, both of which were unrevealing of abnormalities to suggest an alternative diagnosis. CLIPPERS most commonly manifests radiographically as punctate or curvilinear areas of enhancement centered predominantly in the pons and cerebellum; the large lesion in the medulla of our patient with confluent longitudinal extension into the spinal cord would be very atypical for this disorder (as illustrated in Figure 1).1,2 With such extensive testing conducted, we believe that alternative etiologies were thoroughly investigated and appropriately excluded as outlined in the original report.

The patient’s quadriplegia was accompanied by other exam findings of upper motor neuron dysfunction, including spasticity and hyperreflexia, clinically in keeping with a lesion of the central nervous system that was subsequently identified on MRI as displayed in Figure 1.1 The episode of altered consciousness was presumed to be a seizure and managed with levetiracetam. EEG monitoring subsequent to the event only revealed generalized slowing without epileptiform abnormalities or seizures.

In the pre-COVID era, most cases of post-infectious CNS inflammatory complications have traditionally been held to occur within 1 month of the antecedent infection.3 It is important to note however that SARS-CoV-2 is a novel virus, and therefore a different disease than those upon which many of the original estimates of post-infectious ADEM were described. Interestingly, many studies have proven the capacity for nucleic acids of this virus to linger in its host, which may represent a prolonged antigenic trigger for neuroinflammation.4-6 Other larger studies on post-COVID CNS inflammatory diseases share our interest in this particular issue of delayed latency to neurologic onset and have routinely documented cases occurring 30-60 days following viral infection.7-10 At this stage of scientific discovery, however, we emphasize that neuroinflammatory diseases occurring in proximity to COVID-19 infection remain an interesting association and have not been definitively proven to be causally-linked.

We were not previously aware of the case report of post-COVID GBS with the CSF cytokine and chemokine profile presented by Dr Finsterer in his letter. The authors of this paper do cite in their report specifically that this immunological profile was discovered in the context of GBS, a peripheral nervous system complication distinctly different from the central nervous system complication of our patient.11 On the basis of a single case report, it is challenging to advocate for the necessity of conducting this testing in other patients with a different disease. With that noted, it is certainly a fascinating topic that deserves additional attention in the research sphere to determine its future clinical utility.

1Division of Hospital Neurology, Department of Neurology, Emory University School of Medicine, Atlanta, GA, USA
2Division of Neuroimmunology and Neuroinfectious Diseases, Department of Neurology, Massachusetts General Hospital, Boston, MA, USA

Corresponding Author:
Spencer K. Hutto, MD, Division of Hospital Neurology, Department of Neurology, Emory University School of Medicine, 12 Executive Park Dr NE, Atlanta, GA 30329, USA. Email: shutto@emory.edu
Declaration of Conflicting Interests
The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding
The author(s) received no financial support for the research, authorship, and/or publication of this article.

ORCID iD
Spencer K. Hutto  https://orcid.org/0000-0002-5644-9638

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