Unmasking of a Relapsing Encephalomyelitis after SARS-CoV-2 Infection and COVID-19 Vaccination

Shuvro Roy, Paula Barrera, Carlos Pardo-Villamizar, Scott D. Newsome
Johns Hopkins School of Medicine
Baltimore, MD, USA

Introduction

- Prior case studies suggest that severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) and its vaccines may **unmask central nervous system neuroinflammatory conditions.**

- We present a case of **relapsing steroid-responsive encephalomyelitis** after SARS-CoV-2 infection and subsequent COVID-19 vaccination.

Case Description

- **1/2021:** COVID infection not requiring hospitalization
- **3/2021**: Start of progressive lower extremity weakness, erectile dysfunction, paresthesias, and falls.
- **4/2021:** Received COVID vaccines
- **5/2021:** Initial presentation. MRIs demonstrated **innumerable enhancing lesions involving the subcortical white matter, basal ganglia, thalami, brainstem, cerebellum, and the entire spinal cord parenchyma**
- **5/2021:** Cerebrospinal fluid testing revealed a lymphocytic pleocytosis (10 WBC, 88% lymphocytes), normal total protein (61mg/dL), normal total glucose (64mg/dL), and transient matched serum and CSF oligoclonal bands.
- Other tests were unremarkable for infections, malignancies, primary demyelinating conditions
- **5/2021:** Treated with IV steroids, continued on oral prednisone
- **10/2021:** Recurrence of symptoms and lesions with weaning of oral prednisone, requiring a second course of IV treatment.
- **12/2021:** After weaning of oral prednisone, the patient had another relapse requiring IV steroids
- **1/2022:** Transitioned to mycophenolate mofetil, and transitioned off of steroids slowly, without any radiologic or clinical recurrence.

Findings

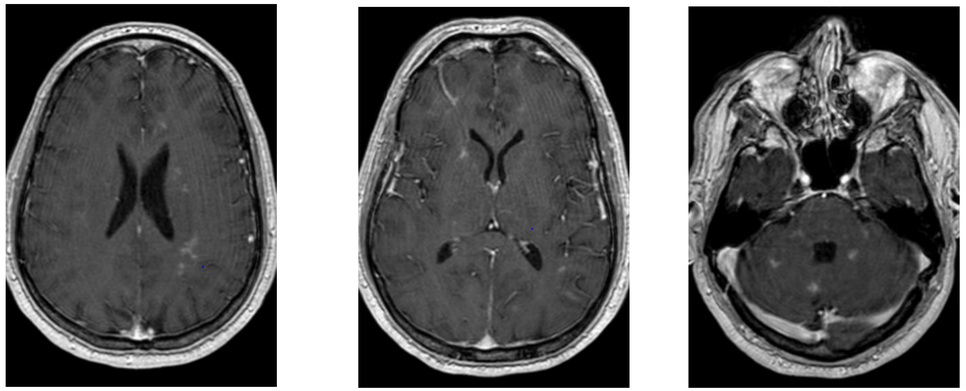

Fig 1. Contrast-enhanced brain MRI demonstrating punctate enhancing lesions scattered throughout the brain parenchyma including subcortical white matter, basal ganglia, and brainstem

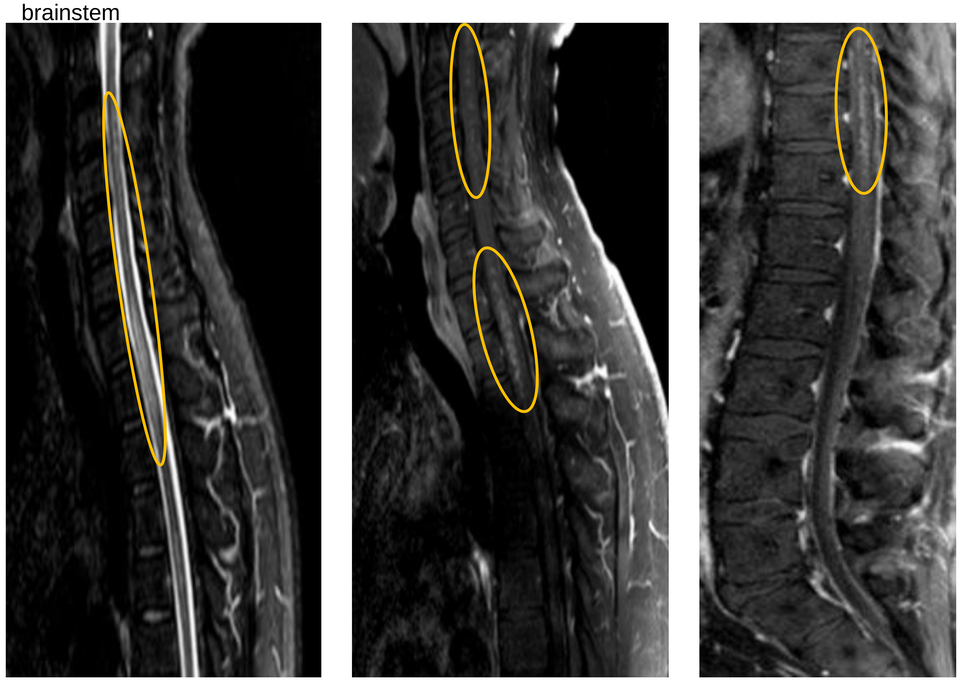

Fig 2. Contrast-enhanced brain MRI demonstrating punctate enhancing lesions throughout the entirety of the spinal cord

Discussion

- The patient's presentation of a relapsing encephalomyelitis that involved the brain parenchyma nd spinal cord raised a broad differential: **NMOSD, MOGAD, histiocytosis or Lyme disease, rheumatologic conditions such as Behcet's disease, Sjogren's syndrome, autoimmune/paraneoplastic neurologic syndromes like Bickerstaff's brainstem encephalitis, as well as CNS vasculitidies**
- The above conditions should be ruled out through serum testing, cerebrospinal fluid testing, and evaluation for smoldering malignancy with PET-CT scan.
- **In the case of recurrent relapses with steroid dependency, patients should be considered for a steroid-sparing immuno-modulating therapy.**
- Apart from respiratory symptoms, SARS-CoV-2 infection can be associated with a variety of neurological manifestations.
- A literature review identified 21 additional cases of CNS neuroinflammatory disease after either SARS-CoV-2 infection or vaccination (11 transverse myelitis, 6 optic neuritis, 3 encephalomyelitis, and one CLIPPERS.)
- Vaccines containing SARS-CoV-2 antigens may enhance autoimmunity. This case highlights that the resulting inflammation may be insidious and extensive, though treatable.
- As COVID-19 constitutes a life-threatening infection, **the benefits of vaccination outweigh the smaller risk of unmasking an immune-related condition.**

References

Tobin WO, Guo Y, Krecke KN, Parisi JE, Lucchinetti CF, Pittock SJ, Mandrekar J, Dubey D, Debruyne J, Keegan BM. Diagnostic criteria for chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids (CLIPPERS). Brain. 2017 Sep 1;140(9):2415-2425. doi: 10.1093/brain/awx200. PMID: 29050399.

Johns Hopkins logo: Johns Hopkins Neurology: Half a Century of Innovation | Johns Hopkins Neurology and Neurosurgery (hopkinsmedicine.org)