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Lauren Ridgway

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Slurred, Blurred, and a Hard-to-Find Word: Acute Progressive Neurologic Changes in a Pediatric Patient

Lauren Ridgway^{1,2}, Amelia Sneve^{1,2}, Andrew Nguyen^{1,2}, Aleisha Nabower^{1,2}

¹Department of Pediatrics, College of Medicine, University of Nebraska Medical Center

²Children's Hospital and Medical Center, Omaha, NE

Background: Acute disseminated encephalomyelitis (ADEM) is typically a viral sequelae, with a 1-3% mortality rate. ADEM has been described as a consequence of acute SARS-CoV-2 infection. This case report describes a child diagnosed with ADEM in the setting of acute SARS-CoV-2 infection with the clinical course complicated by co-infection with Human herpesvirus 6 (HHV-6). HHV-6 positivity in the CSF is often considered an incidental finding, but it has been shown to cause encephalopathy, seizures, and demyelinating disease. This case is unique in that the dual infection with SARS-CoV-2 and HHV-6 may have contributed to the development of ADEM, poor clinical outcome, and resistance to treatment in this case.

Case Presentation: A 9 y.o. male with ADHD presented with 12 hours of acute onset left-sided weakness, blurred vision with left-sided vision loss, expressive aphasia, and ataxia. Head CT was negative at an outside ED prior to transfer. On exam, he was responsive to commands and agitated with waxing and waning consciousness. Exam significant for decreased left peripheral and blurry vision, expressive aphasia, slurred speech, and decreased left grip strength, although full strength testing was limited. Patellar reflexes 3+ but symmetric, Babinski down, and no clonus. Evaluation was pertinent for positive respiratory panel for SARS-CoV-2. CSF was without pleocytosis, elevated protein, and positive for HHV6. Serum studies positive for GAD65 Ab. MRI brain and spine with T2 hyperintensities showing extensive punctate white matter lesions throughout the cerebral hemispheres, midbrain, and cerebellum, enhancing signal abnormality throughout the spinal cord, and dominant lesions in the frontal lobes, to be correlated with vasculitis or demyelinating disorders. Normal MRA/MRV head. The patient received high dose IV steroids followed by IVIG due to lack of response. He was started on IV Ganciclovir, Remdesivir, Cefepime, and Vancomycin. He developed status epilepticus (SE) resistant to multiple anti-epileptic drugs and diabetes insipidus (DI), progressing to cerebral edema and herniation, neurogenic shock, and brain death.

Discussion: The combination of encephalopathy, recent viral illness, acute neurological changes, and multi-white matter lesions in the brain and spine supported a diagnosis of ADEM due to SARS-CoV-2 versus HHV-6. One study reported that seizures occurred in 11% of children with ADEM. SE is a serious complication of ADEM and likely contributed to the development of cerebral edema, resulting DI, and the patient's poor outcome. It is uncertain at this time if co-viral illnesses played a role in this patient's rapid decline.