May 2nd, 12:00 AM

Case Report: Varicella Zoster Virus Encephalitis Presenting with Monochorea

Chetna Thawani
Jefferson Health NJ

Frank Wheeler
Jefferson Health NJ

Case report: Varicella Zoster Virus Encephalitis Presenting with Monochorea

Chetna Thawani, MD, Frank Wheeler, DO

Department of Emergency Medicine, Jefferson New Jersey

Abstract

Meningoencephalitis is a broad range of symptoms as a sequelae of inflammation of the CNS system. It is a diagnosis that is often missed, especially in atypical presentations. We describe a case of HSV encephalitis that presented to the ER afebrile with atypical arm movements, most similar to a monochorea, and discuss other cases in the literature of atypical presentations of this disease.

Case Presentation:

An 83 year old male arrived to the ER by EMS in the late afternoon. EMS reported he was “acting abnormal,” and “recently started on valacyclovir” for a diagnosis at urgent care of shingles. Shortly thereafter, family was also able to provide history. Per them, he was started on valacyclovir yesterday and given one dose yesterday evening. They reported that he had decreased appetite which was unusual for him. When he woke up, he still did not wish to eat, and they felt he was acting different and had started making abnormal hand motions. Throughout the day he was slow to respond to questions, and when he was answering, his sentences were not making complete sense. Initially, they were concerned it was a side effect of the medication. Upon further ROS, they did note that the patient had a fever to 101F that previous night. They also noted that he was continually picking at his rash. The patient had a past medical history of atrial fibrillation, large B cell lymphoma in remission, HTN, HLD, and prostate cancer, and dementia. Per family, at baseline he was aox1-2. On physical exam, his vitals were stable - normotensive and with normal heart rate and respiratory rate - and he was afebrile. (98.1, 72, 18, 125/68, 99%). the patient himself overall appeared well, with a rash and a smile on his face. The rash was primarily on the left side and appeared to be erythematous and pustular with crusting, and scabbing, not unlike a shingles rash. He was answering questions with short answers, but nodding yes and no to questions, Aox1. Otherwise, his heart and lung sounds were benign, and his abdomen was nontender, and he did not have any other focal deficits, no facial asymmetry. He was moving all extremities equally, but he was occasionally moving his arms, particularly his left hand, irregularly in all directions, without clear purpose. Otherwise, he had normal reflexes and no nuchal rigidity. He had normal pulses and normal capillary refill. Initial ED investigation involved typical workup for a change in mental status, including primarily infectious and metabolic testing. Notably he had a WBC at 3.9; he also had normal metabolic panel including normal kidney function. His urinalysis was negative as was his covid / flu testing. His EKG and troponin were within normal limits. His chest xray did not show any signs of pneumonia or any other acute findings. His CT head without contrast showed central atrophy and small vessel ischemic change without any acute infarct or space occupying lesion.

Case Presentation Continued:

As we could not find any true cause at that time for his change in mental status and the abnormal hand movements, we decided to proceed with completing a lumbar puncture. He was at that time empirically started on antibiotics and antivirals for meningitis and admitted to the hospital. Shortly after, lumbar puncture results came back positive for varicella zoster meningitis, cultures were not positive for any other organism. Therapy was narrowed to just acyclovir, and patient was in the hospital for 3 days receiving IV acyclovir, then discharged to a SNF with a PICC line for continued antibiotics administration. Neurology was consulted but patient was unable to tolerate MRI without anesthesia, and family elected for this to be completed outpatient. Of note he was noted to have some erratic behavior throughout hospital stay, and this was considered to be possibly secondary to dementia underlying acute infection.

Discussion:

In our case presentation, we describe a lesser known phenomenon in the clinical presentation of VZV meningoencephalitis. While seizures are not uncommon in the presentation, our patient presentation was not clinically concerning for generalized seizure activity. Initially, when the initial workup was negative, we considered that the patient may just be having worsening of his dementia, and were considering a discussion with family regarding admission for observation versus discharge and follow-up outpatient. Then upon repeat history we learned about the patients fever at home the night before, which prompted us to reconsider meningitis / encephalitis in our differential. Initially, we were unsure of the clinical significance of the patients abnormal arm movements which most resembled a monochorea. While CT scan did not show any clear evidence of encephalitis, MRI may have shown abnormalities in the motor cortex.

References

available on request:
chetna.thawani@jefferson.edu

Discussion:

Upon further literature review, we identified many “atypical” presentations of VZV meningoencephalitis. Platanaki et al identify a case where severe headache was the only symptom prompting the consideration of LP and subsequent diagnosis. A few case studies depicted facial dystonias as symptoms. Another case study identified a case with involuntary lip movements and intractable hiccups as a presenting sign of VZV meningitis.[4] Facial dystonia was described in another case.[5] Another case report depicts a pediatric patient with symptoms of dystonia among other neurological symptoms (generalized tonic clonic seizures, cerebellar ataxia) who ended up having transient bilateral striatal lesions.[6]

Few studies have depicted the hemichoreal motions present in our patient. One noted a hemi-chorea in a patient with VZV endocarditis.[7]

Of note, VZV tends to replicate in arteries. Post varicella arteriopathies, leading to sequale such as ischemic stroke, have been noted in literature, primarily in children. A few case reports showed children with prior VZV infections presenting with hemichorea, noted to have vascular stenosis in the M1 branch of MCA. Some literature supports the idea that VZV induces production of pro-coagulation auto-antibodies, causing some of these vasculopathies.[8] It is possible our patient had a similar vascular involvement causing his monochorea symptoms.

Our patient did present with shingles rash without evidence of Ramsay Hunt syndrome (no facial paralysis). It has been described that shingles rash in the facial nerve poses a higher risk of developing encephalitis. Notably, our patient had a fast transition from rash to encephalitis, both happening within the span of 2 days.

Conclusions:

Our case demonstrates an atypical presentation of VZV encephalitis, presenting with atypical / monochorea-like arm movements. In particular, we noted the importance of obtaining a full history from family up front if possible as this can change decision making earlier on; had we known about the fever earlier we may have considered encephalitis earlier in the case. The importance of keeping a broad differential is important as well; the patient’s family was concerned that the new medication was causing the symptoms, while in reality it was the disease itself.