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Letter to the Editor

Acute truncal ataxia in a healthy adult with varicella zoster virus cerebellitis: A case report and literature review

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ABSTRACT

Acute cerebellitis is a well recognized complication of varicella zoster virus (VZV) infection in children. It has been described in adults in the setting of virus reactivation with a preceding herpes zoster rash, but it is exceedingly rare in adults who are not elderly or immunocompromised, particularly in the absence of a rash. To our knowledge, there has been only one reported case of acute cerebellitis in an immunocompetent adult less than age 65 with virological confirmation of acute VZV infection. We describe a 59-year-old immunocompetent man who presented with acute truncal ataxia without rash and was diagnosed with VZV cerebellitis, supported by anti-VZV IgM and anti-VZV IgG antibodies in the serum and a positive VZV polymerase chain reaction in cerebrospinal fluid. He had robust improvement with intravenous acyclovir treatment and was free of neurologic disability at two month follow-up. This case highlights the importance of virological evaluation in patients with acute ataxia, even in the absence of typical features of infection.

1. Case description

A 59-year-old male with a past medical history significant for tobacco dependence presented with a three day history of progressive gait instability and falls. He endorsed no recent travel, rashes, insect bites, or constitutional symptoms. On exam, the patient was afebrile and hemodynamically stable. Neurological examination demonstrated truncal ataxia characterized by profound postural instability without other cerebellar features. Neurologic examination was otherwise normal; no cutaneous findings were present.

Head computed tomography demonstrated mild ventriculomegaly with features suggestive of normal pressure hydrocephalus (NPH); brain magnetic resonance imaging (MRI) with and without gadolinium showed no additional abnormalities (Fig. 1). MRI of the cervical and thoracic spine revealed mild spondylosis but were otherwise unremarkable. A urine drug screen, heavy metal evaluation and serum ethanol screen were negative. Initially, the acuity of his gait instability was unclear, so a high-volume lumbar puncture (LP) was performed to determine whether the presence of ventriculomegaly was contributing; post-LP assessment revealed no improvement in his gait. Cerebrospinal fluid (CSF) studies demonstrated an opening pressure of 21 cm H2O; total nucleated cells, 104 cells/μL (82% lymphocytes); protein, 57 mg/dL; IgG index, 1.0 (normal < 0.85); and 6 supernumerary oligoclonal bands. Paraneoplastic autoantibody evaluation and cytology were normal. Microbiological evaluation revealed a positive CSF VZV polymerase chain reaction (PCR) and positive serum VZV IgM and IgG, with an IgG index of > 8.0. Additional CSF infectious evaluation included normal testing for cryptococcus, Tropheryma whipplei, mycobacteria, Lyme disease, syphilis, arboviruses, herpes simplex viruses, and John Cunningham virus.

Our patient was diagnosed with VZV cerebellitis and started on a ten-day course of intravenous acyclovir, 10 mg/kg every eight hours. He demonstrated marked improvement after three days of treatment. At his 2-month follow-up he was able to walk unassisted without any evidence of ataxia; he remained free from any rashes.

2. Discussion

We describe an immunocompetent, previously healthy adult who presented with acute truncal ataxia without a rash with acute virological evidence of VZV in the serum and CSF. The differential diagnosis of acute cerebellar ataxia is broad and includes vascular, neoplastic, immune-mediated, toxic-metabolic, demyelinating, and infectious causes. In the absence of imaging abnormalities to narrow the differential, a lumbar puncture should be performed to evaluate for infectious and immune-mediated causes. Infectious causes of cerebellar ataxia are important to recognize and differentiate from autoimmune/paraneoplastic cerebellar ataxia, as treatment and prognosis are vastly different. Careful screening for an infectious etiology should be considered in patients with acute cerebellar ataxia, even without a typical rash or signs of infection. Virological testing including VZV PCR in the CSF and anti-VZV IgM and IgG antibodies should be considered in the setting of acute cerebellar ataxia.

Acute cerebellar ataxia is the most common neurologic complication of VZV infection in healthy children; however, in adults it is a rare manifestation of VZV reactivation, and when present is typically associated with gait ataxia and a predominant tremor [1,3]. A thorough literature review revealed only 5 additional cases of virological confirmed VZV cerebellitis without associated rash, one of which was in a patient less than 65 years of age and immunocompetent [8–12]. Immunosuppression is a major risk factor for VZV reactivation in adults with around 50% of herpes zoster cases occurring in immunosuppressed subjects [2,6]. Other neurological manifestations of VZV reactivation are diverse and include meningoencephalitis, vasculopathy, myelitis, segmental zoster paresis, and polyneuritis cranialis [4,5,10].

The pathophysiological mechanisms of ataxia with VZV reactivation remains unclear, with speculations of an inflammatory syndrome caused by direct infection or postinfectious autoimmune mechanisms

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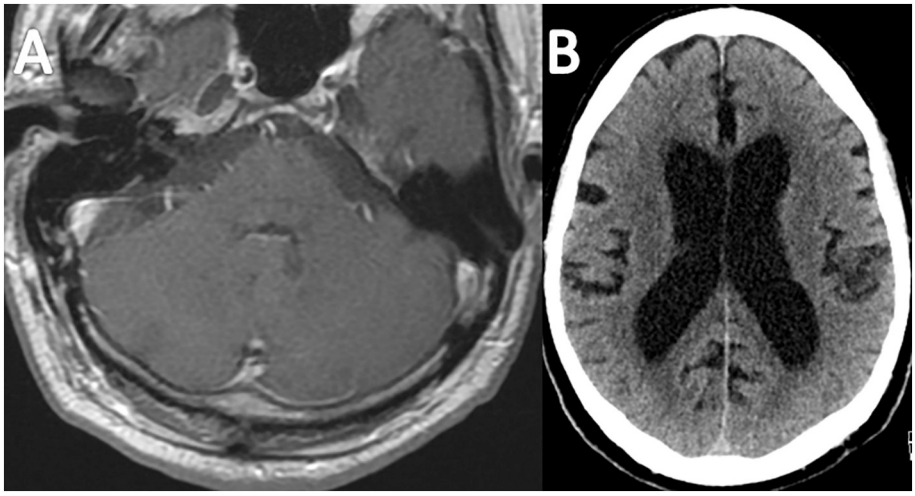

Fig. 1. A) T1-axial MRI brain with gadolinium demonstrating no abnormal enhancement in the cerebellum or pons; and B) Axial head CT depicting mild ventriculomegaly at the time of presentation.

[7]. Additionally, the most efficacious treatment also remains unclear; however, experts typically recommend treating inciting VZV infections with IV acyclovir [3]. While VZV cerebellitis is often self-limited and patients typically recover within weeks without long term sequaelae, treatment with IV acyclovir likely hastens recovery and reduces morbidity.

This case of VZV cerebellitis highlights the importance of considering virological evaluation in patients with acute cerebellar ataxia, even in the absence of typical features of infection.

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Conflict of interest

The authors declare no conflicts of interest.

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