Epstein-Barr associated acute cerebellar ataxia in an adult
Adam N Hofer, MD, MPH; Joan Addington-White, MD
University of Wisconsin-Hospitals & Clinics

Introduction

Acute cerebellar ataxia (ACA) is a rare, often self-limiting, inflammatory disease that can be triggered by both infectious and non-infectious processes. ACA is one of several known central nervous system (CNS) complications of an Epstein-Barr viral (EBV) infection. Here, we present a case of ACA as the primary manifestation of a primary EBV infection without the classical features of infectious mononucleosis.

Case Description

A 70-year-old active and generally healthy woman presented to her primary care physician with a 2-week history of progressive gait disturbance. Her symptoms began following a self-limiting diarrheal illness while traveling to New York City. Her unsteadiness worsened to the point where she had several falls and required assistance for ambulation. She had no other localizing infectious symptoms and denied changes in mental status. She had a history of hypertension and osteoporosis. There had been no recent medication changes. She had no family history of ataxia or other neurological disorders.

On examination, she was afibrile (36.3°C) and hemodynamically stable. Cardiopulmonary and abdominal examinations were normal. There was no cervical lymphadenopathy or tonsillar exudates. She had no dysarthria or nystagmus. She had a wide-based, unsteady gait and was unable to walk without assistance. She also had mild dysdiadochokinesia on finger-to-nose testing. Romberg test was negative.

Complete blood count was notable for a leukocytosis (11.5 K/µL), lymphocytosis (8500/µL), and mild thrombocytosis (430 K/µL). A blood smear demonstrated reactive lymphocytes. A basic metabolic panel was normal. C-reactive protein was mildly elevated (2 mg/dL), ESR was normal. B12 and TSH were normal. An MRI of the brain was notable for ventricular dilatation and a subtle pial contrast enhancement. The influenza virus antibody titer was negative. CSF analysis revealed an increased protein concentration (800 mg/l) and 6,750 leucocytes/mm³.

Discussion

We report a case of acute cerebellar ataxia (ACA) as the primary manifestation of acute Epstein-Barr virus (EBV) infection in a 70-year-old female. In retrospect, the patient in our case reported mild fatigue and a sore throat at the onset of her symptoms. However, while rare, ACA can present without the systemic manifestations of infectious mononucleosis (IM) such as fever, pharyngitis, lymphadenopathy, and fatigue.1 ACA is one of several known neurological complications of EBV (Table 3), which can be seen in up to 7.3% of patients with EBV.2

ACA is rare (incidence 1 in 100,000) and most commonly affects children. In adults, ACA has a male predominance and typically affects patients less than 30 years of age.3,4 However, as in our case, ACA has been documented in patients in their 70s.5 The pathogenesis is incompletely understood, however, two mechanisms have been speculated: (1) a delayed post-infectious autoimmune process, (2) direct viral infection of the cerebellum.1,6,7 The detection of viral DNA in CSF in our case would support the latter.

The differential diagnosis for ACA in an adult is broad, and includes infectious, neoplastic, paraneoplastic, inflammatory, and toxic etiologies (Table 4). In this case, acute EBV infection was concluded to be the causative agent based on positive EBV viral capsid antigen (VCA) IgM in the serum. Repeat EBV serology approximately 10 weeks after diagnosis was notable for negative EBV VCA IgM and positive EBV VCA IgG, consistent with resolving acute infection (Table 2).

MRI is the most commonly performed imaging modality in patients with acute ataxia and may demonstrate bilateral or unilateral diffuse cerebellar abnormalities (Figure 2); however, brain MRI is unrevealing in the majority of patients with ACA and abnormal imaging lacks both specificity and prognostic value. Our patient was noted to have ventriculomegaly, thus initially raising concern for normal pressure hydrocephalus (NPH). However, hydrocephalus has been reported in up to 42% of patients in one case series.6

ACA secondary to EBV infection is usually a benign, self-limiting inflammatory condition that can most often be treated with supportive therapy. Complete recovery is most often seen within 3–13 weeks.8 However, obstructive hydrocephalus and chronic cerebellar atrophy have been reported.9 Treatment has commonly included corticosteroids and/or anti-viral therapy; however, there is currently insufficient evidence to support the use of either in the treatment of ACA.10 Our patient was treated with supportive therapy and made a complete recovery after approximately ten weeks.

Conclusions

- Acute Cerebellar Ataxia (ACA) is a rare, usually self-limiting inflammatory syndrome characterized by cerebellar dysfunction (e.g., gait disturbance, dysmetria).
- EBV may cause ACA in adults and ataxia may be the sole manifestation of a primary EBV infection.
- Conventional MRI is often normal during an acute infection though may demonstrate cerebellar swelling or hydrocephalus.
- Treatment is supportive; there is currently insufficient evidence to support the routine use of corticosteroids or anti-viral medications.

References