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Post-Epstein-Barr Virus Acute Cerebellitis in an Adult

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest: None declared

Patient: Male, 23-year-old
Final Diagnosis: Acute cerebellitis
Symptoms: Ataxia • dizziness
Medication: —
Clinical Procedure: Lumbar puncture
Specialty: General and Internal Medicine

Objective: Unusual clinical course
Background: Acute cerebellitis in adults is a rare disease. The etiology is unknown but postulated to be due to primary infection or para-infection. Different presentations have been reported, which complicates the diagnosis process.
Case Report: We report the case of a young man who presented with headache, vomiting, and vertigo. He was found to have ataxia and cerebellar signs, bradycardia magnetic resonance imaging (MRI) of the brain showed acute cerebellitis, and cerebrospinal fluid (CSF) studies showed lymphocytosis. Further investigations showed the presence of Epstein-Barr virus (EBV) immunoglobulin M (IgM) and IgG. His symptoms resolved completely with corticosteroid and antiviral treatments.
Conclusions: Acute cerebellitis can present in various ways. Bradycardia, along with neurological deficits, should raise the suspicion of acute cerebellitis.

MeSH Keywords: Cerebellar Diseases • Diffusion Magnetic Resonance Imaging • Epstein-Barr Virus Infections

Full-text PDF: <https://www.amjcaserep.com/abstract/index/idArt/918567>

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Background

Acute cerebellitis is inflammation of the cerebellum. There is no consensus on the etiology of acute cerebellitis, yet most cases are caused by primary infection, post-infectious syndrome, or after vaccination [1,2]. There have been reported cases that described a broad spectrum of symptoms ranging from mild symptoms, such as headache and ataxia, to more severe symptoms, such as hydrocephalus, cerebellar atrophy, or tonsillar herniation [2]. Acute cerebellitis is found mostly among children, and very few cases have been reported in adults [1]. We describe the case of a 23-year-old man who presented with dizziness and generalized fatigue. Magnetic resonance imaging (MRI) was suggestive of acute cerebellitis.

Case Report

A 23-year-old Nigerian man, who was previously healthy, presented to the hospital with a 4-day history of dizziness. He described his dizziness as vertigo. This was associated with generalized body aches and weakness. He reported a few episodes of vomiting 4 days prior to admission. He reported not having photophobia, headache, sensory deficits, double vision, difficulty swallowing, or change in voice. He denied having had contact with a sick person or recent travel. There was no fever. Family history was unremarkable.

Physical exam and findings

Initially, upon admission, the patient had 1 episode of a fever of 38.2°C (which resolved spontaneously) and was bradycardic (minimum reading of 41 bpm). Physical examination revealed a well-built man who was alert and oriented but not cooperative, with poor eye contact. He was noted to have dysdiadochokinesia and dysmetria. The patient was barely able to walk, with unsteadiness; he had a tendency to fall to the right while walking. He had impaired tandem gait. The scale for the assessment and rating of ataxia (SARA) was 5. Romberg's sign was positive. The rest of the physical examination was unremarkable.

Laboratory investigations showed normal complete blood cell counts. Inflammatory markers were negative. Drug screening was negative. Cardiac enzymes were normal. The electrocardiogram (EKG) and 24-h cardiac monitor showed sinus bradycardia.

A non-contrast head computed tomography was performed and was unremarkable. Lumbar puncture was performed and revealed leukocytes of 125/μL (84% lymphocytes), red blood cells of 252/μL, with a high protein of 0.99 gm/L and glucose of 3.3 mmol/L. CSF cultures, cryptococcal antigen, and syphilis, TB acid-fast bacilli culture, smear, and TB PCR were all negative.

HIV testing was negative. Viral polymerase chain reaction (PCR) data were normal. However, blood serology showed the presence of EBV IgG and IgM.

Blood cultures were negative. Mycoplasma PCR was negative. Viral PCR for herpes simplex type 1 and 2, Rotavirus, enterovirus, and parvovirus B19 were negative. Screening for autoimmune diseases (i.e., an antinuclear antibody) was negative. *Coxiella burnetii* and dengue virus were not tested.

Management

The patient was started on ceftriaxone (2 g every 12 h) and acyclovir (10 mg/kg/dose every 8 h) based on the local guidelines. The brain was imaged by MRI. MR T1-weighted sequence with gadolinium demonstrated multiple focal patchy cerebellar and leptomeningeal enhancement. In addition, subtle swelling and effacement of cerebellum folia were detected. The findings were suggestive of acute cerebellitis (Figures 1, 2).

Therefore, the patient was immediately started on dexamethasone (6 mg IV every 6 h), with significant resolution of symptoms the following day. Bradycardia resolved after the treatment. He finished 5 days of dexamethasone along with 10 days of ceftriaxone and acyclovir. The patient was discharged home, back to his baseline functional status with no neurological deficits. Unfortunately, he was lost to follow-up.

Discussion

Acute cerebellitis in adults is a rare syndrome. It is more common in children, and the exact prevalence of cerebellitis in adults is unclear [1,3]. The mechanism of the disease remains unclear. Samkar et al. reported that most patients had no identifiable causes, and viral infection contributed to 23% of the cases [1]. Other possibilities are attributed to medications such as isoniazid, especially in patients with renal failure [1].

Of all cases reported in the literature, there have been multiple presentations and outcomes. In a recent literature review, more than 80% of adults presented with nausea, vomiting, ataxia, and headache, and 29% of patients presented with altered mentation [1]. Our patient presented with vague symptoms of dizziness and fatigue, with questionable fever and the absence of other neurological symptoms. Therefore, there was initially no suspicion of acute cerebellitis.

For the majority of cases mentioned in the literature, MRI is considered the criterion standard to diagnose acute cerebellitis [1,2]. Most cases have abnormal signals in T1-weighted MRI images, with contrast in the cerebellar cortex or leptomeningeal enhancement [4,5]. Bilateral changes are more common,

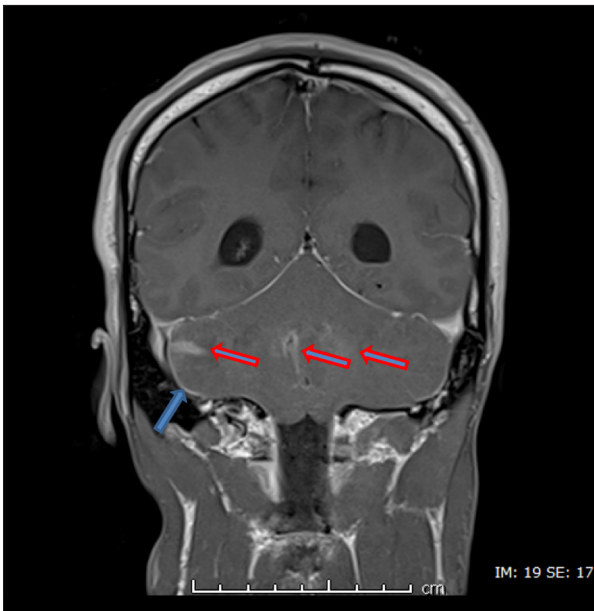


Figure 1. Coronal section MR T1-weighted sequence at the level of the occipital horn, with post-contrast enhancement and fat sat. Multiple subtle focal and patchy enhancement of the cerebellum were detected, mainly at its right side peripherally (indicated by red arrows), with subtle swelling and effacement of its folia. Meningeal enhancement is seen (blue arrow).

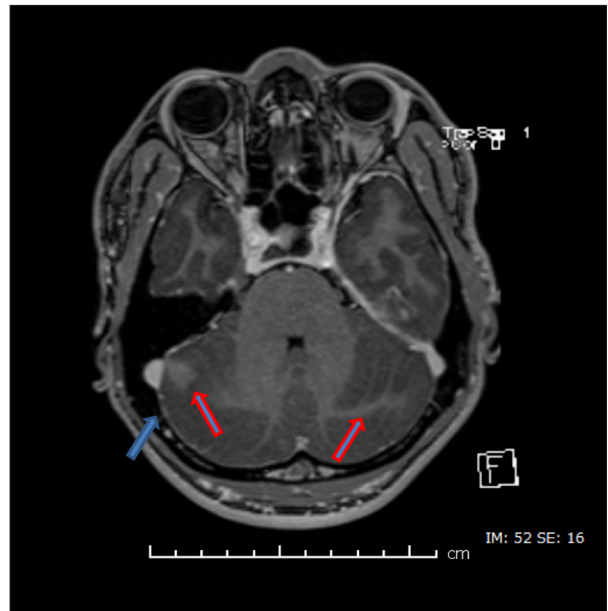


Figure 2. Axial cuts MR T1-weighted sequence at the level of the cerebellum, with post-contrast enhancement showing multiple foci and patchy enhancement of the cerebellum, mainly in the right side (red arrow). Enhancement of its meninges was also noted, indicated by a blue arrow, which are findings suggestive of non-specific cerebellitis.

but unilateral changes are reported in some cases [4]. However, there are rare cases in which MRI was unremarkable, and the diagnosis was established using other modalities, such as lumbar puncture [6].

CSF analysis is used to support the diagnosis and can help in identifying the etiology. Furthermore, serological markers for certain infections such as bacteria, fungi, and viruses can be helpful. In rare circumstances, stereotactic biopsy and histological finding can be used for a definite diagnosis [7].

There are various treatment options for acute cerebellitis. Antibiotics and antivirals should be initiated if there is high suspicion of direct invasion by certain organisms. However, the duration of treatment is not well defined. In addition, corticosteroids play an important role, especially in patients with diffuse brain edema. Surgical options are appropriate for severe complications such as hydrocephalus and herniation [1,4].

We report an unusual case of EBV-induced acute cerebellitis. This patient's initial presentation was atypical. He presented with vertigo, cerebellar signs, and bradycardia. He was diagnosed by MRI. CSF analysis showed lymphocytosis with negative EBV serology in CSF. However, EBV IgM and IgG were positive in serum. This favors the possibility that acute cerebellitis is due to a post-infectious phenomenon. However, serology is

not an optimal diagnostic method, and PCR is superior to serology. Serum PCR was not done in this case.

It was noted that bradycardia had resolved rapidly after initiation of treatment. Bradycardia is usually seen in brain pathology as part of the Cushing's reflex secondary to raised intracranial pressure. In addition, bradycardia is described if there is stretching of the brainstem centers, such as central or uncus herniation. In our case, the patient had bradycardia without intracranial tension, which denotes the possibility of the effect of the infection on the vasomotor regulatory center [8,9].

Serious cardiac arrhythmias, such as atrial fibrillation, have been reported with infectious mononucleosis. EB-associated pericarditis, pericardial effusion, or cardiac tamponade has been reported to cause atrial fibrillation [10,11]. Furthermore, complete heart block has been reported in Epstein-Barr myocarditis [12]. However, sinus bradycardia was benign and seemed self-limiting in this patient, and it resolved after starting the treatment.

Cerebellitis following EBV in adults is rare. The majority of cases that were reported in the medical literature included males under the age of 18 years [9,13]. Several cases have been reported in children following mononucleosis or after infection [14]. Unilateral cerebellitis secondary to EBV has been reported in immunocompromised adults with HIV [7].

There is no consensus on the best treatment of EBV-associated cerebellitis. Some studies do not recommend starting corticosteroid after EBV infection. However, it is recommended to initiate corticosteroid treatment in patients with encephalitis or acute demyelinating encephalomyelitis [4]. Furthermore, acyclovir should be considered in the setting of EBV CNS infection [14]. Corticosteroid and antiviral treatments are initiated based on clinical judgment.

Conclusions

The diagnosis of acute cerebellitis in adults following EBV infection can be difficult. Post EBV cerebellitis can happen in the

absence of preceding symptoms typical of infectious mononucleosis. A high index of suspicion is critical, especially in ataxic patients presenting with signs of cerebellar edema. Bradycardia, along with neurological findings, might be an initial presentation of acute cerebellitis, including the adult age group.

Department and Institution where work was done

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Conflict of interest

None.

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