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An Adult Case of Acute EBV Cerebellitis

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ABSTRACT

Acute neurological manifestations of infectious mononucleosis are uncommon and have been predominantly reported in the paediatric population. We report a case of acute Epstein-Barr virus cerebellitis in an adult in whom spontaneous resolution of symptoms and signs occurred after 2 weeks of supportive treatment. An infective cause for an acute cerebellar syndrome in an adult must always be considered in the differential diagnosis when appropriate.

LEARNING POINTS

- A high index of suspicion is needed in adults who present with a febrile syndrome and cerebellar signs.
- An acute cerebellar syndrome may be the only manifestation of Epstein-Barr virus infection.
- In certain cases, conservative management may be sufficient depending on the clinical severity.

KEYWORDS

Epstein-Barr virus, infectious mononucleosis, cerebellitis

CASE DESCRIPTION

A 42-year-old woman presented to the emergency department with a 1-week history of high-grade fever and lethargy. She denied any symptoms of sore throat, cough, myalgias or gastrointestinal disturbance. Her general practitioner had treated her with an empirical course of co-amoxiclav with no improvement in her symptoms. She denied any recent travel history.

She was admitted to hospital for further investigations. Two days after admission, she developed slurring of speech and unsteadiness which progressively worsened. She also complained of nausea and vomiting. Doxycycline was initiated on admission in view of an insect bite, but she was still spiking fever despite treatment. On examination, she was febrile and tachycardic with a pulse rate of 105 beats per minute. Her blood pressure was 105/80 mm Hg with no postural decline. Her heart rate was in dual rhythm and there were no murmurs. Her apex beat was not displaced. She had a respiratory rate of 16 breaths per minute. The breath sounds were symmetrically vesicular with no wheeze or crepitations.

Her abdomen was soft on superficial palpation. The liver and spleen were not palpable. She had no palpable lymph nodes in the cervical, axillary or inguinal regions. No rashes were noted.

A nervous system examination showed an alert and fully oriented middle-aged woman. Her speech was dysarthric but she had a normal cranial nerve examination with normal saccades. She had no neck stiffness and Kernig's and Brudzinski's signs were negative. Fundoscopy showed normal discs with bilateral venous pulsations. Upper and lower limb examination revealed normal tone and power with normoreflexia. Sensory examination was normal and plantars were down-going bilaterally.

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She had mild dystaxia in both upper limbs with no dysdiadochokinesia. The heel-shin test was impaired on both sides. Gait assessment showed a wide-based gait and the patient was unable to tandem gait.

METHODS

Initial laboratory investigations showed a normal white cell count with a mildly raised lymphocytic count of 3.73×10^9/l (normal range: 1.30–3.60×10^9/l). The liver and renal profiles were within normal limits. Inflammatory markers showed a mildly raised erythrocyte sedimentation ratio of 15 mm in the first hour (normal range 10–14 mm in the first hour). Magnetic resonance imaging (MRI) of the brain showed no abnormalities. A viral screen was requested including Epstein-Barr virus (EBV) IgM and IgG and cytomegalovirus IgM and IgG which were negative. A lumbar puncture was performed which showed a normal opening pressure, with a total number of cerebrospinal fluid (CSF) lymphocytes of 632×10^6/l (normal range: up to 5×10^6/l) with raised CSF protein of >1 g/l (normal range: less than 450 mg/l) with a CSF/serum glucose ratio of 0.5. Polymerase chain reaction (PCR) on CSF revealed an EBV titre of 7,843 copies/ml. The rest of the CSF viral screen was negative. On repeating the lumbar puncture 1 week later, the lymphocytic count had improved to $286\times10\times19^6$/l and the CSF protein level had decreased to 0.8 g/l. In view of the clinical picture and laboratory findings, a diagnosis of acute viral cerebellitis secondary to EBV was made.

The patient's symptoms improved gradually and she was discharged after a few days. She was reviewed a week later and had fully recovered with normalization in speech and absence of the previously elicited cerebellar signs. The patient refused another lumbar puncture which was aimed at showing complete resolution of the abnormal CSF.

DISCUSSION

In the adult population, the differential diagnosis of an acute cerebellar syndrome includes metabolic, vascular, neoplastic, inflammatory, autoimmune and infective causes, whether bacterial or viral[1, 2]. The latter include cytomegalovirus, coxsackievirus, poliovirus, rotavirus, echovirus, varicella zoster virus, herpes simplex virus and EBV. In this setting, acute cerebellitis may occur during the course of an infection, as a post-infectious manifestation or after immunization[3]. Patients usually present with a febrile illness, truncal ataxia, nystagmus, intention tremor, headache and altered mental state. Infectious mononucleosis commonly presents with an uncomplicated febrile illness, pharyngitis and local lymphadenopathy.

This case above focuses on a rare neurological presentation of infectious mononucleosis, both because it occurs in only 1% of cases and because these usually manifest in young patients in the post-infectious period. Diagnosis was made on the basis of the patient's symptoms, the presence of CSF lymphocytosis and by detection of virus by PCR in CSF. CT and MR imaging were normal and there was no clinical evidence of meningo-encephalitis. The patient was given supportive treatment and recovered spontaneously within 2 weeks. A repeat lumbar puncture at this point showed a reduction in EBV viral load in CSF.

Reported cases of EBV cerebellitis pertain mostly to the paediatric population and there are no established guidelines regarding treatment indications and regimens for EBV cerebellitis in adults. The use of steroids and acyclovir have been suggested in some case reports based on clinical trials, but their effect and benefit have yet to be ascertained[4, 5]. Severe neurological complications such as hydrocephalus, cerebellar oedema and brainstem herniation have been reported[6]. The use of intravenous immunoglobulins has been mentioned in post-infectious cases of EBV cerebellitis[7]. In our case, no specific treatment was administered and the patient improved spontaneously.

CONCLUSION

Since only a few cases of acute EBV cerebellitis in adults have been reported in the literature, this case aims at highlighting the need for a high index of suspicion for diagnosing this condition in patients who present with an acute cerebellar syndrome in the setting of a febrile illness. Further studies are necessary to establish treatment guidelines for such cases.

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