CASE REPORT

Epstein-Barr virus-associated cerebellar ataxia

Khalid Ali,¹ Charlotte Lawthom²¹Department of Neurology, Morriston Hospital, Swansea, UK²Department of Neurology, University Hospital of Wales, Cardiff, UK

Correspondence to

Dr Khalid Ali,
khalid_w2003@yahoo.co.uk

SUMMARY

Cerebellar ataxia is a common neurological presentation. It can be acute, subacute or chronic. Neurological complications of Epstein-Barr virus (EBV) are well-recognised with a variety of presentations. Acute cerebellar ataxia is a rare, but an established complication. It has been described as the sole manifestation of EBV infection without the systemic features of infectious mononucleosis. The pathophysiology is not clear. The course of the illness may last for a few months with a benign outcome, though serious complications can happen. We present a case of a 38-year-old man who presented with an acute cerebellar ataxia owing to EBV infection, along with a review of the literature.

BACKGROUND

A male patient presented with an acute cerebellar ataxia, the initial thinking was that of a paraneoplastic disorder, which led to extensive investigations which put the patient and his family under tremendous pressure emotionally.

We later arrived to the diagnosis of EBV-associated cerebellar ataxia. This is a benign condition with good recovery. We suggest the clinicians who receive such cases to consider monospot test with constant observation.

CASE PRESENTATION

A 38-year-old man presented to our department with a 2-week history of gait disturbance. He had initially developed mild headache and felt lethargic. Within 2 days, this condition was replaced by unsteadiness and clumsiness. He worsened over 2 weeks and needed to use a walking frame to mobilise. He had a history of psoriasis. He did not take any medications, but used ciclosporin previously. He has no family history of significance, in particular, no history of ataxia. He consumed little amount of alcohol and he did not smoke.

On examination, he was afebrile with normal general examination. He had slow saccadic eye movements both in the horizontal and vertical meridian with no nystagmus. He was mildly dysarthric. He had no pyramidal or extrapyramidal signs. Sensory examination and in particular his proprioception were normal. He had mild finger nose and heel shin ataxia. He struggled to stand and managed to walk few steps only with a wide-based gait using a frame and needing supervision. Romberg test was negative.

INVESTIGATIONS

Full blood count, urea and electrolytes, liver and thyroid function tests were normal. C reactive

protein was 1 mg/l. No abnormality was detected on MRI of the brain with contrast. Cerebrospinal fluid analysis (CSF) showed a normal opening pressure, CSF glucose of 3.5 mmol/l (plasma glucose 5.1 mmol/l) with CSF protein of 1.07 g/l. There were 18 lymphocytes in the CSF. CSF cytology showed no malignant cells. Oligoclonal bands were negative in CSF. The CSF was negative for HSV1 and HSV2 and enterovirus PCR. Antineuronal antibodies against Yo, Hu and Ri antigens were negative and he had a negative anti-GAD antibodies. CT scan of his chest and abdomen and a FPG-PET CT scan showed mild to moderately enlarged lymph nodes and mild splenomegaly. Lymph node histology showed strong positivity for EBV, which was suggestive of acute EBV lymphadenitis. This was also supported serologically with a positive serum monospot test with raised EBV capsid antigen IgM in his serum. A bone marrow biopsy showed no evidence of lymphoma. His HIV test was negative.

TREATMENT

The patient was initially treated with 14 days of acyclovir. He was later given 5 days course of intravenous immunoglobulins.

OUTCOME AND FOLLOW-UP

His gait gradually improved over few weeks time. He was free of symptoms at 4 months follow-up outpatient appointment. He is doing well at 12 months follow-up.

DISCUSSION

We report a case of acute cerebellar ataxia in a young healthy man. The differential diagnosis includes vascular, inflammatory, autoimmune, toxic, neoplastic, paraneoplastic, infective and genetic causes. Supported by the initial investigations, paraneoplastic cerebellar degeneration was highly suspected. The CT and the PET CT scan of the whole body confirmed lymphadenopathy and splenomegaly. Lymph node biopsy failed to reveal neither primary nor secondary neoplasm. Instead, it showed strong evidence of acute EBV infection. This was later confirmed serologically.

Infectious mononucleosis is a common benign viral infection. It has a large spectrum of systemic complications. Neurological complications of EBV infection are rare, but recognised with an incidence of 0.37–7.3%. Meningitis, encephalitis, mononeuritis, Guillain-Barre syndrome, ophthalmoplegia, optic neuritis and acute demyelinating encephalomyelitis (ADEM) have been described. Acute cerebellar ataxia is a rare neurological complication of

To cite: Ali K, Lawthom C. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-009171

EBV infection;^{1–3} it was first described by Landes and colleagues in 1941.⁴

The condition has a male predominance and affects young adults.^{1–13} Kuwahara *et al*¹² reported a 61-year-old woman with this condition. The pathophysiology is not fully understood whether it is due to direct invasion of the cerebellum by the EBV or delayed postinfectious autoimmune mechanism. Several case reports detected antibodies against EBV in the CSF suggesting a direct viral infection of the cerebellum.^{1–3} The condition can present in the context of infectious mononucleosis with fever, pharyngitis, lymphadenopathy and atypical lymphocytosis, but acute cerebellar ataxia can be the only feature of the infection without the other systemic manifestations.^{1–3}

The condition has been described as benign with a good recovery within 3–13 weeks.^{3–5–8} Cerebellar swelling leading to obstructive hydrocephalus has been described. MRI scan and particularly DWI sequence (diffusion-weighted imaging) can be helpful to predict early cerebellar cytotoxic oedema and swelling.^{11–13} Treatment with corticosteroids has been used with doubtful benefit, the evidence behind which is very limited.^{11–13}

Learning points

- ▶ Neurological complications of Epstein-Barr virus (EBV) are rare, but recognised.
- ▶ EBV-associated cerebellar ataxia should be considered in the differential diagnosis of acute cerebellar ataxia. Monospot test is easy and not an expensive test to perform. This will save the patients', their relatives' and the doctors' time, money and anxiety.
- ▶ EBV-associated cerebellar ataxia has a benign course.
- ▶ There is no satisfactory evidence to support any treatment, but steroids have been tried in severe cases with some success.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Lascelles RG, Longson M, Johnson PJ, *et al*. Infectious mononucleosis presenting as acute cerebellar syndrome. *Lancet* 1973;2:707–9.
- 2 Fujimoto H, Asaoka K, Imaizumi T, *et al*. Epstein-Barr virus infections of the central nervous system. *Intern Med* 2003;42:33–40.
- 3 Erzurum S, Kalavsky SM, Watanakunakorn C. Acute cerebellar ataxia and hearing loss as initial symptoms of infectious mononucleosis. *Arch Neurol* 1983;40:760–2.
- 4 Landes R, Reich JP, Perlow S. Central nervous system manifestations of infectious mononucleosis. *JAMA* 1941;116:2482–4.
- 5 Wadhwa NK, Ghose RR. Acute cerebellar ataxia and infectious mononucleosis. *Postgrad Med J* 1983;59:457–8.
- 6 McCarthy CL, McColgan P, Martin P. Acute cerebellar ataxia due to Epstein-Barr virus. *Pract Neurol* 2012;12:238–40.
- 7 Kramer DS, Smitnik LM, John K, *et al*. Acute cerebellar syndrome in infectious mononucleosis: documentation of two cases with Epstein-Barr virus infection. *J Natl Med Assoc* 1985;77:305, 307–8.
- 8 Klockgether T, Döller G, Wüllner U, *et al*. Cerebellar encephalitis in adults. *J Neurol* 1993;240:17–20.
- 9 De Fraiture DM, Sie TH, Boezeman EH, *et al*. Cerebellitis as an uncommon complication of infectious mononucleosis. *Neth J Med* 1997;51:79–82.
- 10 Yabuki S, Kazahaya Y, Ikeda K. Acute cerebellar ataxia associated with infectious mononucleosis—a case report and review of the literature. *No To Shinkei* 1983;35:337–42.
- 11 Van Lierde A, Righini A, Tremolati E. Acute cerebellitis with tonsillar herniation and hydrocephalus in Epstein-Barr virus infection. *Eur J Pediatr* 2004;163:689–91.
- 12 Kuwahara S, Kawada M, Uga S, *et al*. A case of cerebellar meningo-encephalitis caused by Epstein-Barr virus(EBV): usefulness of Gd-enhanced MRI for detection of the lesions. *No To Shinkei* 2000;52:37–42.
- 13 Göhlich-Ratmann G, Wallot M, Baethmann M, *et al*. Acute cerebellitis with near-fatal cerebellar swelling and benign outcome under conservative treatment with high dose steroids. *Eur J Paediatr Neurol* 1998;2:157–62.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow