Myelitis: A Rare Presentation of Epstein Barr Virus

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Abstract

EBV associated nervous system complications includes encephalitis, meningitis, cerebellitis, polyradiculomyelitis, transverse myelitis, cranial and peripheral neuropathies, and psychiatric abnormalities are usually more commonly seen in immunocompromised patients and rarely in immunocompetent patients. Here we are reporting a 13 years old boy developed headache, malaise, sore throat and low back pain with radiation to both lower limbs. Next day he felt numbness below umbilicus followed by acute onset weakness in both lower limbs and urinary retention. Motor exam revealed proximal muscle power MRC grade 4/5 and distal power 1/5 in right lower limb and proximal power 4-/5 and distal power 0/5 in left lower limb with normal power in both upper limbs. Deep tendon reflexes were bilaterally normal except absent ankle reflexes. Both plantars were mute. All the modalities of sensation including pain, touch, temperature, joint position and vibration were impaired below umbilicus. Routine investigations were normal. The magnetic resonance imaging (MRI) of thoracic spine showed intramedullary lesion in conus, which was iso-hyperintense on T1-weighted and hyperintense on T2-weighted images extending from D12thoracic vertebral level to L1 with cord expansion (Figures 1, 2). The MRI features were suggestive of conus myelitis. Cerebrospinal fluid (CSF) analysis revealed increased protein, normal cells, glucose and Chloride. CSF Polymerase chain reaction (PCR) was positive for Epstein Barr virus. The clinical and imaging findings were consistent with the diagnosis of myelitis and responded well to steroid plus acyclovir treatment. The clinicians should be aware of such uncommon etiology of a common disease.

Introduction

The Epstein-Barr virus (EBV) is a member of the her-pesviridae family associated with central nervous system (CNS) diseases (encephalitis and meningitis), but rarely cause myelitis.1 The EBV associated CNS infections usually have devastating consequence in immunocompromised patients of nasopharyngeal carcinoma, burkit lymphoma, Hodgkin’s disease and lymphoproliferative disease. The incidence of EBV related neurological complications is underestimated. It varies between 1-18 percent of individual with infectious mononucleosis and includes encephalitis, meningitis, cerebellitis, polyradiculomyelitis, transverse myelitis, cranial and peripheral neuropathies, and psychiatric abnormalities.2 The pathogenesis of EBV associated CNS complications is unknown but may be due to direct invasion of neuron, infiltration of EBV infected lymphocyte and deposition of immune complexes in the endothelium causing injury to neuronal parenchyma. Other possibilities of EBV infected B cells are actively attacked by EBV specific cytotoxic T cells causing injury to neuronal tissue.3 Here we are reporting EBV associated myelitis in an immunocompetent adolescent boy and response to steroid plus acyclovir treatment.

Case vignette

A 13 years old boy developed headache, malaise and sore throat. Over next 2 weeks he developed low back pain with radiation to both lower limbs initially right followed by left. Next day he felt numbness below umbilicus in form of inability to feel clothes and touching with his own hands. Few hours later he developed acute onset weakness in both lower limbs in the form of inability to move his limbs in bed followed by difficulty in micturition leading to urinary retention. He required an indwelling catheter draining 800 ml urine on insertion. There was no h/o fever, loss of consciousness, visual blurring, seizure, upper limb weakness, trauma or tuberculosis.

General physical and other systems examination was unremarkable. On neurological examination higher mental function, speech and cranial nerves including fundi were normal. Motor exam revealed proximal muscle power MRC grade 4/5 and distal power 1/5 in right lower limb and proximal power 4-/5 and distal power 0/5 in left lower limb with normal power in both upper limbs. Biceps, triceps supinator and knee deep tendon reflexes were bilaterally normal while ankle reflexes were absent. Both plantars were mute. Other superficial reflexes were absent. All the modalities of sensation including pain, touch, temperature, joint position and vibration were impaired below umbilicus. There were no meningeal or cerebellar signs. Examination of spine revealed local tenderness without gibbus below L1 vertebral level and SLR test was positive bilaterally.

Hemogram, biochemistry including liver functions, renal functions, thyroid function tests and serum vitamin B12 level, HIV and VDRL were normal. Erythrocyte sedimentation rate (ESR) was normal (15 in first hour). The magnetic resonance imaging (MRI) of thoracic spine showed intramedullary lesion in conus, which was iso-hyperintense on T1-weighted and hyperintense on T2-weighted images extending from D12thoracic vertebral level to L1 with cord expansion (Figures 1, 2). The MRI features were suggestive of conus myelitis. Computed tomography (CT) of chest and abdomen was normal. The markers for autoimmune and connective tissue disorders (ANA, Anti-ds DNA, Anti-nucleosome, Anti-histones, Anti-Sm, Anti SS-A, Anti RO, Anti Scl-70, Anti Rib-PProtein, Anti-JO, Anti-SS-B) were negative. Lumbar puncture showed normal intracranial pressure (140 mm H2O). Cerebrospinal fluid (CSF) analysis revealed increased protein, normal cells, glucose and Chloride. CSF acid-fast bacillus (AFB) stain was
Fig. 1: The magnetic resonance imaging (MRI) of thoracic spine axial view showed normal intramedullary lesion in conus, which was hyperintense on T2-weighted images extending from D12 thoracic vertebral level to L1.

Culture for Mycobacterium tuberculosis and other bacteria were negative. Polymerase chain reaction (PCR) was positive for Epstein Barr virus and negative for other viruses including Herpes simplex virus, Enterovirus and Cytomegalovirus. CSF anti-AQP4 antibody (NMO antibodies) and oligoclonal bands were negative. The clinical and imaging findings were consistent with the diagnosis of myelitis. The patient was treated with high dose intravenous methylprednisolone (1000 mg per day for 5 days) and antiviral drugs acyclovir 30mg/kg /day for 2 weeks. At the time of discharge, he showed moderate improvement in muscle power MRC grade 4/5 in both lower limbs with minimal left side foot drop during walking. He was discharged on supportive treatment and physiotherapy was advised.

Discussion

EBV myelitis is rare in immunocompetent individual. The clinical feature of myelitis and MRI findings of increased signal in conus part of spinal cord were similar to those described by Merelli et al. EBV DNA Virus is positive in CSF mainly in immunocompromised patients with CNS disease but our patient was immunocompetent. This CSF abnormality indicated that Acute EBV myelitis was an inflammatory disease process due to white matter involvement. There was transverse expansion of the spinal cord due to EBV infection, manifesting as asymmetric motor and sensory symptoms in immunocompromised patient. Our patient also presented with asymmetric motor and sensory symptoms but he was not immunocompromised. Steroid, immunoglobulin and antiviral had limited role in presenting disease progression described earlier in the literature but our patient responded well to steroid plus antiviral therapy. However, more studies are needed for consistent favorable response.

Conclusion

EBV associated nervous system complications are usually more commonly seen in immunocompromised patients and rarely in immunocompetent patients. Here we are reporting a 13 years old boy who was diagnosed as EBV associated conus myelitis and responded well to steroid plus acyclovir treatment. The clinicians should be aware of such uncommon
etiology of a common disease and ask for CSF studies for various viruses to differentially diagnose for precise underlying pathogens.

References

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