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Case report

Acute Epstein-Barr virus infection presenting as Guillain-Barre syndrome

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ABSTRACT

An 18-year-old man presented with 5-days of a lower extremity rash, sore throat, rapidly progressive bilateral facial numbness and paresthesias in his distal extremities. His neurological examination acutely deteriorated to include moderate bilateral facial weakness in a lower motor neuron pattern, mild flaccid dysarthria, mild bilateral interossei weakness, and diffuse hyporeflexia. In addition to neurological examination, EMG results of acute demyelinating polyradiculoneuropathy were suggestive of Guillain-Barre Syndrome (GBS). Infectious laboratory testing demonstrated acute infection of Epstein-Barr Virus (EBV) with relatively low EBV DNA quantitative values. The patient subsequently developed fever and cervical lymphadenopathy during his hospital course.

Contrasting typical GBS, which presents weeks after an acute infection, the patient's presenting symptom of EBV infection was GBS. GBS as a presenting symptom of EBV has not previously been described. This case may represent a unique mechanism for the pathogenesis of GBS in acute infections as opposed to the traditional post-infectious antibody-mediated process.

Introduction

Guillain-Barre Syndrome (GBS) often presents as an acute, paralyzing illness triggered by an antecedent infection which provokes an immune response targeting the myelin or axon of a peripheral nerve which cross reacts with peripheral nerve components due to molecular mimicry [1]. Nearly a third of all GBS cases are preceded by a Human Herpes Virus, such as Epstein-Barr virus (EBV) which is known to cause infectious mononucleosis [2].

Case report

A healthy 18-year-old male with no significant medical history presented with a lower extremity rash, sore throat, rapidly progressive bilateral facial numbness and ascending paresthesias from distal extremities. At presentation, he had no discernable skin rash. He did not have fever, adenopathy, pharyngitis, or splenomegaly. Neurologic examination revealed moderate bilateral facial weakness in a lower motor neuron pattern, mild flaccid dysarthria, mild bilateral interossei weakness, and diffuse hyporeflexia. He had a leukocytosis of 12,600/μL, but other laboratory values were unremarkable including liver function enzymes. CT head revealed no intracranial pathology. Cerebrospinal fluid (CSF) revealed no pleocytosis, mildly elevated protein of 56, with negative culture. EMG was consistent with acute demyelinating polyradiculoneuropathy. His symptoms progressed over the first day including ptosis, ophthalmoparesis, bulbar weakness, quadriparesis, and areflexia, further confirming the diagnosis of Guillain-Barre syndrome (GBS). He ultimately required intubation on day 3 for airway protection.

A diagnosis of acute Epstein-Barr Virus (EBV) infection was made after serologic testing demonstrated a positive EBV IgM anti-VCA, and negative both IgG anti-VCA and anti-EBNA; EBV DNA quantification was positive (218 IU/mL). Over the next several days, he developed symptoms consistent with acute mononucleosis such as cervical adenopathy, fever to 38.8 °C, and a rash around his neck. He did not have pharyngitis or splenomegaly. At that time his EBV DNA increased to 505 IU/mL, before decreasing to <100 one week later. Additional infectious diseases tests were negative including Lyme disease, babesiosis, anaplasmosis, HIV, West Nile virus serology (including convalescent sera), CSF PCR for Varicella Zoster Virus, Cytomegalovirus, Enteroviruses, and West Nile Virus. The patient met the criteria for GBS based on his ascending paralysis and EMG that demonstrated low action potential amplitude of the ulnar muscle as well as blink reflexes.

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Quadriparesis and facial weakness improved two weeks after receiving intravenous immunoglobulin (IVIG). He initially required a tracheostomy and PEG tube for significant bulbar and diaphragm weakness. After inpatient rehabilitation he ambulated independently at discharge. Two months later he returned to school with near baseline function.

Discussion

This case is unique due to the timing of GBS symptoms prior to subsequent symptoms of mononucleosis in acute EBV infection. GBS typically occurs 10–14 days following a respiratory or gastrointestinal illness, and is commonly viewed as a post infectious antibody-mediated disorder caused by molecular mimicry between the lipooligosaccharides of infective organisms and the surface molecules on myelin or axons [1–3].

The EBV infection was deemed to be acute due to a combination of positive EBV IgM anti-VCA, and negative both IgG anti-VCA and anti-EBNA. Initial EBV DNA quantitative serology was 218 IU/mL (range: 100−5 million IU/mL). This suggests that the patient's immune system responded quickly to develop GBS before EBV infection manifested with symptoms. This immune response was quicker than typical GBS cases; thus, the immune system was potentially signaling a different mechanism than the traditional post-infectious antibody-mediated process. Aggressive infectious evaluation in GBS cases without a clearly identifiable trigger may yield a better understanding of different mechanisms driving this disorder.

To our knowledge this is the first reported case where GBS was the initial symptom of EBV. This case may represent a unique mechanism for the pathogenesis of GBS in acute infections.

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Ethical approval statement

Informed consent was obtained from the patient and his family prior to the publication of this case report.

Declaration of Competing Interest

All authors declare no conflicts of interest in the publication of this report.

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