



Guillain-Barre Syndrome with Lymphocytic Pleocytosis of the CSF

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Abstract

Introduction: Guillain-Barre typically presents as a symmetric ascending weakness with areflexia. The cerebrospinal fluid typically shows albuminocytologic dissociation. In this case, we present an atypical presentation of severe Guillain-Barre with cerebrospinal fluid exhibiting lymphocytic pleocytosis.

Case report: 60-year-old man presented with progressive lower-extremity weakness that progressed to involve respiratory failure and areflexia over several weeks. Electromyography showed both demyelinating and axonal features. Lumbar puncture revealed a lymphocytic pleocytosis. Given the abnormalities on these tests with a clinical picture of Guillain-Barre the patient underwent extensive paraneoplastic testing and full neuro-axis imaging. Imaging revealed abnormal enhancement of ventral and dorsal nerve roots which is consistent with Guillain-Barre.

Conclusion: In conclusion, a diagnosis of Guillain-Barre syndrome may still be considered in a patient with clinical findings and EMG studies consistent with GBS but with a CSF profile that does not show the typical albuminocytologic dissociation. However, all other causes of CSF lymphocytic pleocytosis must be ruled out before coming to this conclusion.

Keywords

Guillain-Barre, Lymphocytic pleocytosis, Ascending weakness

Introduction

Guillain-Barre typically presents as a symmetric ascending weakness with areflexia that progress over a period of two to four weeks. Diagnosis is made by evaluating the clinical picture, as well as electromyography (EMG) and a lumbar puncture. EMG will show an acute polyneuropathy with demyelinating features. Typically, cerebrospinal fluid shows albuminocytologic dissociation, which is an elevated protein with normal white blood cell count. This case represents an atypical presentation of severe Guillain-Barre with cerebrospinal fluid exhibiting lymphocytic pleocytosis.

Case Report

A 60-year-old African American man developed insidious onset of lower-extremity weakness and ascending paresthesias. At baseline he was able to walk independently and perform all his activities of daily living. Imaging studies, including an MRI of the brain, did not show any acute abnormality. The patient was then discharged to a rehab facility where his respiratory status deteriorated. He was

intubated and transferred to a university hospital for evaluation for suspected Guillain-Barre syndrome.

On admission, he was awake and able to nod in response to simple questions. The patient had diffuse weakness and absent deep tendon reflexes in all extremities. A lumbar puncture was attempted on admission but was unsuccessful. He was subsequently started on a 5-day course of intravenous immunoglobulin. On day 6, EMG studies showed a motor neuropathy with both demyelinating and axonal features. Specifically, there were absent responses in the tibial nerves bilaterally, decreased CMAP amplitude the right peroneal and right median with prolonged duration. The right median also had slowed conduction velocity. Active denervation was seen on EMG in the left tibialis anterior, medial gastrocnemius, and right deltoid. The patient refused to continue with the sensory portion of testing. On day 8 of admission, lumbar puncture under fluoroscopy was performed and revealed a lymphocytic pleocytosis (WBC: 49, 97% lymphocytes, RBC: 43, protein: 266, CSF glucose: 86). MRI of the cervical and thoracic spine on day 9 of admission showed abnormal enhancement along bilateral ventral and dorsal nerve roots concerning for an inflammatory polyneuropathy, such as Guillain-Barre syndrome. Brain MRI on day 9 did not show any acute findings. A repeat lumbar puncture on day 14 of admission once again showed lymphocytic pleocytosis (WBC: 81, 97% lymphocytes, RBC: 0, protein: 109, glucose: 73). The CSF showed no phenotypically abnormal cell population. A sensory-motor neuropathy antibody panel was sent, which included anti-ganglioside antibody, and the entire panel was negative. All infectious, paraneoplastic, and autoimmune studies were negative, including an HIV test. The only exceptions were a positive HTLV antibody but western blot was negative and a positive West Nile IgG with a negative IgM antibody. Other serologies and antibodies tested that were negative included: VDRL, FTA, lyme, VZV, ACE, EBV, enterovirus, HSV, campylobacter, CMV, voltage gated calcium channel antibody, acetylcholine receptor antibody, C-ANCA, P-ANCA, X-ANCA, anti-sDNA, anti-SS A, anti-SS B, cryoglobulins, heavy metals, rheumatoid factor, and cytology. In addition, CT scans of the chest, abdomen, and pelvis did not show any malignancy.

Unfortunately, the patient did not show significant neurologic improvement during his hospital stay and continued to be ventilator dependent. He underwent a tracheostomy and was discharged to a long-term acute care facility for ongoing ventilator weaning.

Discussion

Guillain-Barre syndrome is an acute immune-mediated

polyneuropathy that presents as progressive symmetric muscle weakness with hyporeflexia or areflexia. Diagnosis is based on clinical features, CSF analysis and nerve conduction studies. The classic CSF finding is albuminocytologic dissociation, which is an elevated CSF protein with a normal white blood cell count. A study done at Massachusetts General Hospital of 110 patients with GBS showed that the cell count was less than 5 cells/mm³ in 87% of patients and greater than 30 cells/mm³ in 2% of patients [1]. Our patient met the clinical and EMG criteria for GBS as well as had an MRI suggestive of nerve root involvement. His CSF profile, however, was atypical for GBS. There are several explanations as to why the cerebrospinal fluid profile was seen.

CSF pleocytosis is commonly seen in patients with HIV [2] as well as HIV-associated GBS [3,4]. However, our patient tested negative for HIV. There are few reports of non-HIV infected patients with Guillain-Barre syndrome presenting with lymphocytic pleocytosis. A particular case of a child with recurrent GBS who presented with lymphocytic predominant CSF pleocytosis recovered rapidly following administration of IVIG [5].

Rauschka et al. [6] examined five cases in which there was a CSF pleocytosis or CSF-polymorphonuclear granulocytes [6]. These five cases were typical clinical cases of GBS but all patients died within 100 days of onset. The clinical course of each of these was more severe and CSF showed pleocytosis, but autopsy revealed there was not a significant difference in their pathology when compared with controls. Berciano et al. [7] describes another similar case in which the patient had a fulminant case of GBS and CSF pleocytosis was noted. Similar to these cases, our patient could have an atypical severe presentation of GBS [7].

Another consideration is an inflammatory process such as aseptic meningitis in the setting of IVIG. The rate of aseptic meningitis following IVIG infusion greatly varies. In a study of 384 patients with Kawasaki disease who received IVIG, 4 patients developed aseptic meningitis [8]. In another study of 54 patients receiving high-dose IVIG for various immune related neuromuscular disorders, 6 patients developed aseptic meningitis within 24 hours after completion of the infusion [9]. Of note, the patients who developed aseptic meningitis in these cases did develop clinical symptoms of meningitis while

our patient did not. We recognize that most patients with aseptic meningitis develop symptomatic headache but in this case the patient was intubated and had significant pain associated with his radiculitis that was being treated with pain medications that may have masked a headache.

In conclusion, a diagnosis of Guillain-Barre syndrome may still be considered in a patient with clinical findings and EMG studies consistent with GBS but with a CSF profile that does not show the typical albuminocytologic dissociation. However, all other causes of CSF lymphocytic pleocytosis must be ruled out before coming to this conclusion.

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