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Mini Case Study

Unusual Presentation of Guillain-Barré Syndrome as Miller Fisher Subtype

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Abstract

We report an unusual presentation of Guillain-Barré syndrome as Miller Fisher subtype, as indicated by ocular weakness, exhibited through ptosis, double vision and decreased peripheral vision due to limited extraocular muscle movement. The patient was an 80-year-old Caucasian male, a smoker with a significant medical history of coronary artery disease, hypertension, diabetes, hyperlipidemia and atherosclerosis of the aorta. Miller Fisher syndrome presented shortly after symptoms of low-grade fever and persistent mucoid cough for 5 days, suggestive of an upper respiratory infection

Keywords: *Guillain-Barré Syndrome (GBS); Miller Fisher Subtype; Human Immunodeficiency Virus (HIV)*

Introduction

The acute immune-mediated polyneuropathies are classified under the eponym Guillain-Barré Syndrome (GBS); once considered a single disorder, it is now known to be a heterogeneous syndrome with several variant forms. Frequently, GBS reveals in the form of the acute monophasic paralyzing disease caused by a previous infection. The axonal forms of GBS are also well recognized in addition to the common demyelinating form. In our case, the first presentation was ocular, which is very uncommon.

The mechanism for GBS necessitates an antecedent infection evoking an immune response. The sharing of cross-reactive epitopes causes activation of peripheral nerve components, resulting in acute polyneuropathy. The immune response is directed toward the myelin or the axon of the peripheral nerve. Antecedent infections are believed to alter the immune response that leads to acute polyneuropathy. Approximately two-thirds of patients report an antecedent respiratory tract or gastrointestinal infection. Campylobacter infection is the most commonly identified precipitant of Guillain-Barré and can be detected in as many as 30 percent of cases. Other precipitants include cytomegalovirus, Epstein-Barr virus, Mycoplasma pneumoniae, and influenza-like illnesses. GBS can also occur in association with human immunodeficiency virus (HIV) infection, predominantly in those who are not profoundly immunocompromised. Following another triggering event like surgery, trauma, immunization, or bone-marrow transplantation; a small percentage of patients may develop GBS.

Hospitalization

The patient was initially in rehabilitation following a hip surgery that was complicated by a stress-induced gastric ulcer. Upon admission to rehab, all vital data and blood indices were within normal limits aside from a mild iron deficiency, reflected by hemoglobin of 9.

The mild iron deficiency was likely a sequela of recent blood loss due to the gastric ulcer for which he had esophagogastroduodenoscopy and cauterization, followed by two units of packed red blood cells while in the hospital. During the course of rehabilitation, the patient's performance status markedly improved. The patient had expected findings of mild upper respiratory infection upon physical exam but insisted on discharge. A few days later, the patient's symptoms worsened, and he was readmitted to the hospital with significant paralysis. The patient underwent seven sessions of plasmapheresis along with intravenous immunoglobulin (IVIg) and steroid therapy. Following treatment, the patient underwent physical and occupational therapy in the hospital for medical optimization. The course of the disease has been regressive, with ocular symptoms the last to resolve. Diagnostic tests included lumbar puncture, CT of the brain, MRI, electrophoresis and a nerve conduction test.

Diagnosis

Traditionally, the main diagnostic tool for GBS is a clinical diagnosis with appropriate history and physical exam. Diagnosis focuses on symmetric or modestly asymmetric distal muscle weakness and absent or depressed deep tendon reflexes. Weakness is variable, from mild difficulty with walking to nearly complete paralysis of all muscles of the extremities, progressing to facial, respiratory and bulbar muscles. However, some GBS variants present with local or regional involvement of particular muscle groups or nerves, and several have prominent cranial nerve involvement; the variable initial presentations can hinder early diagnosis.

Supportive features include the following:

- An elevated CSF protein (> 45 mg/dL) with a normal CSF white blood cell count.
- In the demyelinating forms of GBS, electrodiagnostic studies demonstrating abnormalities including motor conduction block, slowing of motor and sensory nerve conduction, temporal dispersion, and prolonged distal latencies. In the axonal forms of GBS, nerve conduction studies showing the decreased amplitude of motor responses, with normal conduction velocities.
- Contrast enhancement of the spinal nerve roots, cauda equina, or cranial nerve roots on MRI.
- Detection of serum IgG antibodies to GQ1b, supporting the diagnosis of the GBS variants Miller Fisher syndrome, Bickerstaff encephalitis, and pharyngeal-cervical brachial weakness.

Cerebrospinal fluid

In patients with Guillain-Barré Syndrome, lumbar puncture shows elevated CSF protein with a normal CSF white blood cell count. This finding is recognized as albuminocytologic dissociation, and it is seen in 50 to 65 percent of patients with GBS in the first seven days after the onset of symptoms, and more than 75 percent of patients in the third week.

Electrodiagnostic studies

Electrodiagnostic studies are the most specific and sensitive tests for the diagnosis of GBS; they establish the underlying pathophysiology as either demyelinating or axonal. The performance of a detailed neurophysiologic study enables diagnosis of pediatric GBS in as many as 90 percent of cases during the first week of symptoms. Changes are virtually universal by the second week of illness, by which time a definitive diagnosis can almost always be made. Electrodiagnostic studies are uncomfortable and can be technically challenging in small children; they should therefore be undertaken only by individuals with appropriate pediatric expertise.

Peripheral nerve demyelination

Peripheral nerve demyelination, which can be accompanied by conduction block, often primarily affects proximal nerve roots and the terminal segments of motor nerves. Nerves terminating intramuscularly predominantly result in unrecordable motor responses.

Magnetic resonance imaging

Spinal MRI with the administration of gadolinium frequently shows enhancement of the spinal nerve roots and cauda equina during the first weeks after the onset of GBS in children. The enhancement may be diffuse or predominantly involve the ventral (anterior) nerve roots, and less often the dorsal (posterior) roots. In certain cases, nerve root enhancement might be deferred and can only be observed on a repeat MRI. Enhancement of cranial nerve roots may also be seen in some cases, reflecting more diffuse nerve involvement.

Antibodies

Immune reactions directed against epitopes in Schwann cell surface membrane or myelin can cause the acute demyelinating form of GBS, while immune reactions against epitopes contained in the axonal membrane cause the acute axonal forms of GBS. Antibodies against GQ1b, a ganglioside component of nerves, are present in the vast majority of patients with Miller Fisher syndrome.

Treatment

Compared with that of other forms of Guillain-Barré Syndrome, the course of illness for patients with Miller Fisher syndrome is generally milder, with most patients experiencing complete recovery inside of 6 months, even with no treatment. Regardless, close monitoring should still occur in these patients, as a subset of MFS patients has been known to develop respiratory failure, weakness of the limbs, and facial or bulbar muscle paralysis [1]. In the event that a patient with MFS should have a more severe disease course with respiratory involvement, signs that may suggest impending respiratory failure include the patient being unable to count to 20 out loud on a single breath and marked neck muscle weakness. Patients with a negative inspiratory force of less than 60 cm H_2O or forced vital capacity under 15 cc/kg of ideal body weight should be intubated and mechanically ventilated. Feeding or nasogastric tubes should be considered for patients with severe dysphagia to prevent aspiration [2].

Effective treatment of GBS involves administration of either IVIg or plasmapheresis. Steroids (prednisolone, methylprednisolone, etc.) have not been shown to improve outcomes. While IVIg and plasmapheresis have demonstrated similar efficacy, IVIg is preferred in certain situations. In patients with Miller Fisher Syndrome, IVIg has not been shown to improve outcomes, with patients recovering fully from ataxia and ophthalmoplegia within 1 year without treatment [3].

Conclusion

Due to the commonly encountered cases of non-specific weakness and fatigue. clinicians shall place Guillain barre syndrome in the differential diagnoses. It is a commonly underdiagnosed problem. Good history taking and examination is so far the best modality of early suspicion. More research input must be geared towards various associated etiologies and presentation as well as treatment.

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