Severe Progressive Back Pain Causing the Misdiagnoses of Guillain-Barré Syndrome (Twice)

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Recommended Citation


ISSN: 2769-2779

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Abstract

Introduction:

Guillain-Barré syndrome (GBS) is a rare autoimmune demyelinating disease. Symptoms vary widely and commonly include ascending bilateral weakness, pain and hyporeflexia. Approximately 30% will develop respiratory failure contributing to the high morbidity and mortality. Advancements in diagnostic and treatment have greatly decreased mortality to now less than 10%. However, GBS is still often misdiagnosed, thus delaying care.

Case Description:

A 42-year-old female presented with 1 week of back pain that now is affecting her hands and feet. Her neurological exam was within normal limits. Imaging included brain CT and MRI, chest and abdomen CT, were all unremarkable along with labs. She was discharged home with pain medication for unspecified back pain. The next day, the patient presented back to the hospital with progressive weakness and pain now occurring throughout her body. After additional workup was negative, the physicians were now considering drug addiction as a diagnosis. She was discharged home and told to follow up with her primary physician, who urgently referred her to a neurologist. The following morning, the patient saw a neurologist who recommended a lumbar puncture and nerve conduction study but were not performed due to insurance complications. She was now unable to walk and told to immediately return back to the hospital. Upon arrival, her neurological exam deteriorated further, and she was in respiratory distress. A lumbar puncture showed albuminocytologic dissociation. A nerve conduction study showed multifocal demyelinating peripheral neuropathy confirming GBS. Intravenous immunoglobulin was initiated but despite several doses, her symptoms continued to worsen. She was intubated and transferred to a tertiary hospital for plasmapheresis. On Day 14 she had a tracheostomy and a percutaneous endoscopic gastrostomy tube placed and was discharged to a Long-Term Acute Care Hospital on Day 25.

Discussion:

GBS is often misdiagnosed especially when patients present with atypical symptoms, like in this case, back pain. After the more common etiologies for back pain are ruled out, and with a high index of suspicion, initiating early neurology consultation and early therapy will help slow progression. Severe progressive pain, including back pain, is a common presenting symptom and is caused from the demyelination of peripheral nerves. Multiple medical textbooks and resources emphasize the more typical symptoms such as progressive ascending neuropathy, yet few highlight the pain severity. Current studies are underway to help address this matter including increasing physicians’ awareness and comfort with diagnosing and treating GBS.

Keywords
Guillain Barre, autoimmune, neuropathic pain, ascending neuropathy

Conflict of Interest Statement
No financial conflicts of interest to report.

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This case report is available in Advances in Clinical Medical Research and Healthcare Delivery: https://scholar.rochesterregional.org/advances/vol2/iss1/7
Introduction

Guillain-Barré syndrome (GBS) is a rare autoimmune demyelinating disease, occurring at approximately 1.72 patients per 100,000 person/year.\(^1\) GBS is often preceded by an infection, surgery, trauma, or vaccination.\(^2\) However, the absence of any previously defined etiology does not necessarily exclude GBS.\(^3\) The most common symptoms include ascending symmetric bilateral limb weakness and hyporeflexia or areflexia.\(^1\) Less common symptoms include severe pain, fever, dyspnea, and paralysis.\(^3\)

Up to 30% will develop respiratory failure contributing to the high morbidity and mortality.\(^4,5\) The recent advancements in diagnostics and treatments have greatly decreased mortality to now less than 10%.\(^6\) A common cause of increased mortality is the failure to appropriately diagnose GBS and initiate early therapy. Those who are diagnosed late will likely have a longer hospitalization course and a slower recovery period.

Having an atypical presentation such as non-specific back pain, normal deep tendon reflexes, late onset weakness, or cranial nerve abnormalities that mimic stroke, are factors that delay the diagnosis of GBS.\(^5,7,8\) It can take up to 5 visits to the Emergency Department (ED), with 2 visits being the average, before GBS is even considered.\(^1\) Other etiologies that can manifest similarly to GBS include peripheral vascular disease, heavy metal or drug intoxication, spinal cord ischemia, transverse myelitis, stroke, polymyositis, or severe metabolic derangements.\(^1\)

In 2015, Dubey et al. showed in their study that among the 69 patients that were eventually diagnosed with GBS, the diagnosis was considered in only 34 (49.3%) of the patients on the initial ED visit.\(^1\) Reasons for this result included patients presenting with atypical symptoms and delayed consultation with a neurologist. In the same study, those who were assessed early by a neurologist during the initial hospitalization, were 71.2% more likely to be discharged home rather than to a long term acute care hospital or inpatient rehab.\(^1\) Additionally, residual symptoms such as neuropathy or weakness were more likely to be present on discharge for those with delayed treatment.\(^1\) In this case study, we describe a patient with GBS that was diagnosed after her third hospital visit, which contributed to her complicated clinical course and her prolonged recovery.

Case Presentation

ED Visit 1: A 42-year-old female, with essential hypertension, presented to a community hospital with a one week history of severe back pain that radiated to her hands and feet. She denied any recent illnesses, injuries, medication changes or vaccinations. On examination, her blood pressure was elevated at 160/107 mmHg, her other vital signs were unremarkable. Her cranial nerves, motor function, and sensation were intact with normal deep tendon reflexes. Imaging included brain computed tomography (CT) and magnetic resonance imaging (MRI) to exclude
stroke, along with chest and abdominal CT to exclude aortic artery pathology, were all negative along with her labs. GBS was on the differential diagnosis but was placed much lower as she did not have the typical manifestations. Without any meningeal signs, the staff presumed a lumbar puncture would be low yield. She was discharged home with opiate pain medication for unspecified back pain.

**ED Visit 2:** The next day, the patient presented back to the hospital with progressive pain and paresthesia throughout her arms and legs along with increasing weakness. Aside from mild ataxia on tandem heel walk, her neurologic exam was, again, unremarkable. An MRI of her cervical, thoracic and lumbar spine was performed to evaluate her vertebrae and spinal cord as well as a repeat CT brain, which were all negative. The physicians were now considering drug addiction as a possible diagnosis because opiate pain medication was the only treatment that relieved her discomfort. She was discharged home and instructed to follow up with her primary physician. She saw her primary physician who then referred her urgently to a neurologist.

**ED Visit 3:** The following morning, the patient saw a neurologist at their private office, who suspected GBS and recommended a lumbar puncture and nerve conduction study. Unfortunately, it would have to be postponed until approved by insurance. At this point, she was unable to walk and in a wheelchair. The neurologist advised her to return back to the hospital immediately. On arrival, she noticed that her neurologic exam had deteriorated further with areflexia on her lower extremities. She also was in respiratory distress and subsequently admitted to the Intensive Care Unit. A lumbar puncture was performed showing mild albuminocytologic dissociation: 179 mg/dL (normal range 15-45). A nerve conduction study followed that showed multifocal demyelinating peripheral neuropathy consistent with GBS.

Intravenous immunoglobulin (IVIG) was initiated but despite receiving multiple doses, her symptoms continued to worsen. She was intubated and transferred to a tertiary hospital for plasmapheresis. She had a prolonged hospitalization with a tracheostomy and a percutaneous endoscopic gastrostomy tube placement and was ultimately discharged to a long term acute care hospital on Day 25.

**Discussion**

An important aspect of this clinical case was the patient’s initial chief complaint of severe progressive pain that started in her back. This atypical presentation of GBS misled the initial physicians. However, progressive severe pain, including back pain, is a common presenting symptom that is seen in more than 50% of cases and is caused from the autoimmune demyelination of the peripheral nerves.\(^6\),\(^8\) Furthermore, one third of cases will have pain preceding limb weakness and neuropathy.\(^7\) The reason why this is problematic is because many medical textbooks, journals, and medical resources place the most emphasis on the more
typical symptoms such as progressive symmetric bilateral neuropathy, areflexia, cranial nerve abnormalities, and respiratory dysfunction. Yet, there is hardly any emphasis on the pain severity with GBS, which delays the diagnosis during its early phases.7

Another key factor that delayed this patient’s diagnosis was that the community hospital did not have inpatient neurology services readily accessible. A neurologist may have provided additional insight into the atypical symptoms of GBS in this case and recommend further testing (i.e. lumbar puncture) and starting treatment early. The challenging part for physicians is deciding which symptoms warrant neurology consult and further workup, especially during the initial stages. Current studies are underway to increase physicians’ awareness and comfort with diagnosing and treating GBS.1

The neurologist that the patient saw had advised the hospital to perform the lumbar puncture and the nerve conduction study, which helped diagnose and confirm GBS. It is likely her symptoms had been ongoing for longer than a week prior to testing, which increases the sensitivity of these tests.8 However, if the tests were performed within a week of the patient’s symptoms or performed in cases of mild GBS disease, the results might not have been diagnostic. Repeating diagnostic studies at a later time (approximately 2-3 weeks later) would be helpful to confirm the diagnosis.8 Nonetheless, GBS is a clinical diagnosis and treatment should be started promptly with IVIG or plasmapheresis, especially when symptoms rapidly progress.8 Treatment should not be withheld while waiting for confirmation testing.

In conclusion, this case report highlights the challenges physicians have in the diagnosis of GBS in patients presenting with atypical symptoms such as back pain. Although medical literature does not emphasize pain being associated with GBS as much as other common symptoms, pain is still frequently seen. GBS is rare and if there is a high index of suspicion, neurology should be consulted promptly to determine if further testing, including lumbar puncture, is warranted after the more common etiologies are excluded.

Conflict of Interest Statement
No financial conflicts of interest to report.
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