Peripheral Neuropathy

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KEYWORDS

• Peripheral neuropathy • Guillain-Barré syndrome
• Charcot-Marie-Tooth disease • CIDP • POEMS

Patients who have peripheral neuropathy present in a variety of scenarios to the office, to the emergency department, and a times in the setting of other major medical problems. The majority of patients can be diagnosed, classified, and managed based on history and physical examination.

PATIENT ONE: ACUTE PERIPHERAL NEUROPATHY

Question: How to Approach a Patient who Presents with Symptoms of Acute or Rapidly Developing Peripheral Neuropathy?

Case presentation 1

A 31-year-old farmer presents with 48 hours of progressive difficulty described as initial trouble with getting up out of a chair and going up and down stairs with subsequent trouble walking, decline in balance, and heaviness in the arms. The patient noted several days of a febrile illness with abdominal cramps, loose stools, and malaise before the onset of these symptoms. Past medical and family histories are unremarkable.

Examination shows 4/5 strength involving arms and legs with slightly greater weakness proximally than distally. There is reduced position and vibration sense at the toes and ankles. Muscle stretch reflexes are absent throughout. Cranial nerve function is normal and forced vital capacity (FVC) is 5 L.

Discussion

This patient presented with an acute progressive quadriparesis heralded by a gastrointestinal illness. The examination suggests that the weakness likely was at the level of the motor unit (anterior horn cell, roots, peripheral nerve, neuromuscular junction, or muscle). The presence of somewhat more weakness proximally than distally raised the question of a myopathy but the finding of sensory deficit in the feet pointed toward a localization, including sensory and motor function. Thus the likely localization was the sensory motor peripheral nerve and or the roots. As all neurologists have seen, there are patients who present with an acute myelopathy in whom the examination looks more peripheral than central. Patients who have spinal cord infarction or transverse myelitis and even some who have compressive lesions may present with an

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acute progressive quadiparesis, reduced stretch reflexes, reduced muscle tone, and variable form of sensory deficit, and it may be challenging to know with certainty that the problem is peripheral rather than a myelopathy. As a general rule, even if patients are experiencing the phenomenon of acute spinal shock with reduced tone and hypoactive muscle stretch reflexes, the majority of such patients display an extensor toe sign in the early phase of their illness. Also, as a general rule, if clinicians are not completely certain that a problem is peripheral, then it behooves them to obtain immediate MRI of the spine.

The presence of proximal weakness more than distal or, in some patients, arm weakness worse than leg can make the diagnosis of peripheral neuropathy less obvious. Many clinicians consider patients who have distal more than proximal and leg more than arm weakness as reflecting the distribution of peripheral neuropathy, but a significant proportion of patients who have acute peripheral neuropathy may present otherwise.

The differential diagnosis for the patient (discussed in Case Presentation 1) is headlined by acute inflammatory demyelintating polyradiculoneuropathy (acute inflammatory neuropathy or Guillain-Barré syndrome [GBS]). The antecedent illness, time course for presentation, and preponderance of motor involvement all are in favor of GBS.

As such, the patient should be admitted to an ICU setting in which he can have meticulous nursing care and vigilant monitoring for respiratory, bulbar, and autonomic dysfunction. For the patient discussed in Case Presentation 1, I would push for an immediate MRI of the entire spine with gadolinium. Routine laboratory tests should be sent off and supplemented with studies (discussed later) for conditions that may mimic GBS depending on the level of certainty of the diagnosis. Tests that can provide evidence to support the GBS diagnosis include a spinal fluid evaluation. I would obtain a cerebrospinal fluid (CSF) examination after looking at the MRI study. Also, blood for GM1 ganglioside antibodies can be useful especially in patients who have a gastrointestinal prodromal illness when looking for *Campylobacter jejuni*–related GBS. Often the establishment of Campylobacter-related illness helps with the long-term prognosis and realistic expectations for the time course of recovery.

What is Known About Guillain-Barré Syndrome and How Should Patient Evaluation and Treatment be Handled?

GBS is considered the prototype ''postinfectious'' neurologic disorder. The majority of patients describe an antecedent febrile illness, followed in days or weeks by the development of ascending paralysis, which seems to be on the basis of an acute inflammatory peripheral neuropathy. Although most skilled neurologists recognize GBS when they see it, the disorder is heterogeneous and diverse in its antecedent events, clinical presentations, and natural course. Even the name is diverse, with GBS, Landry-Guillain-Barré-Strohl syndrome, acute inflammatory demyelinating polyradiculoneuropathy, and acute inflammatory neuropathy all synonymous.

History of Guillain-Barré syndrome

Octave Landry[1] is credited with the earliest description of what has come to be recognized as GBS. In 1859 Landry reported a condition, which he called *acute ascending paralysis*, describing a patient who had a febrile illness in the springtime, which was followed by the development of sensory symptoms within 2 months. One month later, the patient developed subacute progressive inability to walk due to leg weakness, respiratory failure, and death. Landry pointed out that bowel and bladder function were spared, sensory involvement was mild, and muscles respond to faradic electrical