CLINICAL VIGNETTE

Guillain-Barré Syndrome

Adam Solis-Cohen, MD1 and Farid Abdelmalak, MD1

1UCLA David Geffen School of Medicine, Department of Medicine

Case Report

A 43-year-old woman with no significant past medical history presented with acute onset of upper and lower extremity weakness for one week. She also noted numbness and sensory loss of bilateral upper and lower extremities. She was on vacation at Lake Havasu when her symptoms began. She initially noted numbness in her hands, which progressed up her arms, also including her mouth. She then reported lower extremity weakness with increasing difficulty walking. She presented to an ER in Arizona and was transferred to UCLA for further evaluation and treatment.

Admission Vital signs were within normal limits. Physical exam was significant for mild, bilateral lower extremity weakness and diffuse lower extremity sensory loss. Labs included unremarkable CBC, BMP, and coagulation panel. MRI brain and cervical spine were unremarkable for any pathology. Thoracic and lumbar spine MRI was positive for smooth and mild enhancement along the surface of the conus medullaris and the cauda equina nerve roots. These findings, although nonspecific, were consistent with Guillain-Barré syndrome. The patient underwent lumbar puncture and CSF was significant for albuminocytologic dissociation. EMG and Nerve conduction study showed polyradiculopathy with predominant axonal involvement, particularly in bilateral lower extremities. She was treated with 4 days of IVIG and noted improvement in motor function during treatment. She was transferred to acute rehabilitation for further care. She received another IVIG treatment at the rehab facility and continued to make gradual improvements in functioning with physical therapy.

Discussion

Guillain-Barré syndrome (GBS) is the most common and most severe acute paralytic neuropathy with incidence of about 100,000 people per year.1 The syndrome is also known as “acute inflammatory demyelinating polyneuropathy” and “acute motor axonal neuropathy” to describe different phenotypes.

Guillain-Barré syndrome is usually preceded by infection or other immune stimulation that induces an aberrant autoimmune response targeting peripheral nerves and their spinal roots.2 Molecular mimicry between microbial and nerve antigens is thought to be a driving force behind the development of GBS, as exemplified in the case of Campylobacter Jejuni infection. However, the interplay between microbial and host factors that dictates if and how the immune response is shifted towards unwanted autoreactivity is still not well understood.3 Moreover, genetic and environmental factors that affect individual susceptibility to develop the disease are unknown.4

Pathophysiology

Existing evidence supports a mechanism of inflammatory neuropathy due to cross reactivity between neural antigens and antibodies that is induced by specific infections.5 Glycans expressed on lipopolysaccharides of preceding infectious organisms are thought to be the common molecular mimic that triggers an immune response to these carbohydrate antigens located on microbial and axolemmal surface molecules.3 The mechanism is believed to be antibody mediated complement fixing damage to myelin sheath (schwann cell components) in the case of demyelinating polyneuropathy and to axons (axolemma) in the case of axonal polyneuropathy. This is supported by the discovery of specific antibody biomarkers to neuronal membrane gangliosides (GM1 and GD1a). In Miller Fisher syndrome, anti GQ1B was shown to be disproportionately enriched in motor nerves that innervate extraocular muscles.6 When compared to acute motor axonal neuropathy, the immunological cascade in acute inflammatory demyelinating polyneuropathy is less well understood due to a wider array of immune stimulants and to the lack of specific antibody biomarkers discovered.1

The clinical characteristics of GBS are a rapidly progressive bilateral weakness of the extremities, often with sensory and cranial nerve involvement 1-2 weeks after an infection (immune stimulation) with progression to peak clinical deficit in 2-4 weeks.1,7 Weakness is classically ascending, usually starting distally but can be proximal.1 Alternate presentations include Miller Fisher syndrome, which is characterized by the triad of ophthalmoplegia, ataxia, and areflexia. This is due to cranial nerve involvement resulting in facial, oculomotor, or bulbar weakness, which can extend to involve the limbs.1 Patients with GBS can also have sensory loss, ataxia, and features of autonomic dysfunction.1 Muscle pain or radicular pain is often present. Most patients have, or develop, reduced...
tendon reflexes in the affected limbs.² Twenty to thirty percent of patients develop respiratory failure requiring mechanical ventilation.¹ Progressive weakness usually continues for 4 weeks and can last up to 6 weeks. Severity and duration is highly variable and can range from mild weakness with spontaneous recovery to quadriplegia and ventilator dependent without signs of recovery for several months or longer. Although severe cases are uncommon, GBS is remarkably clinically diverse, and includes several distinctive variants and atypical cases.¹

The diagnosis of GBS is largely a clinical diagnosis but can be aided by cerebrospinal fluid (CSF) and electromyography (EMG). CSF studies show albuminocytologic dissociation—the combination of a normal cell count and increased protein level. Nerve conduction studies (NCS) abnormalities are most pronounced 2 weeks after the start of weakness.⁸ NCS allow clinicians to divide GBS into acute inflammatory demyelinating polyneuropathy, acute motor axonal neuropathy, or acute motor and sensory axonal neuropathy.⁹

NCS in patients with acute inflammatory demyelinating polyneuropathy show features of demyelination, including prolonged distal motor latency and reduced nerve conduction velocity.⁸ Features of axonal Guillain-Barré syndrome (acute motor axonal neuropathy or acute motor and sensory axonal neuropathy) are decreased motor, sensory amplitudes, or both.¹⁰

Treatment

Treatment of GBS requires supportive care, IVIG or plasma-pheresis. Combination of plasma exchange followed by IVIG is not significantly better than plasma exchange or IVIG alone.¹¹ Supportive care includes monitoring of respiratory function with maximal inspiratory pressures and vital capacity to predict diaphragmatic strength and timely transfer to ICU if needed.¹ Negative inspiratory force (NIF) is a bedside test to measure respiratory muscle function. Normal is usually greater than 60cm water. If the NIF is dropping or nears 20cm water, respiratory support needs to be available. The Erasmus GBS classification of Guillain-Barré syndrome: pathogenesis, diagnosis, treatment and prognosis. J Peripher Nerv Syst. 2014 Jan;19(2):88-103. doi: 10.1111/jnss.12074. PubMed PMID: 25039604.


