CHAPTER **22**

Guillain-Barré Syndrome: Literature Review and Case Presentation

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Guillain-Barré syndrome (GBS) was first described in 1859 by French Physician, Jean-Baptiste Octave Landry. Landry separated GBS from other forms of chronic neuropathies, unified the clinical features of the disease, and termed it the "Acute Ascending Paralysis." This clinical description accurately describes the progressive lower extremity ascending symptoms of pain and muscle weakness associated with GBS. In 1916, Gorges Guillian, Jean Alexandre Barré, and André Strohl published their observation of two soldiers who presented with paralysis and areflexia with albuminocytological dissociation causing increased spinal fluid protein concentration (1-3).

The onset of GBS can occur at any age with an annual incidence of 0.4 to 1.7 cases per 100,000, and is slightly more prone to occur in males than females. GBS is a disorder in which the body's immune system attacks the peripheral nervous system. This onset is associated with an infection (commonly gastrointestinal or respiratory) in 65% of patients. The initial symptoms of GBS are tingling, numbness, and pain in the lower extremities, alone or in combination. Subsequently, weakness of the lower extremities can occur bilaterally and can worsen over time as the symptoms ascend up the legs (2). In many instances, the symmetrical weakness and abnormal sensations can spread to the upper body and extremities. After the first clinical manifestations of the disease, the symptoms can progress over the course of hours, days, or weeks. In most cases, by the third week of the illness, 90% of all patients are at their weakest. As the neuropathy ascends, it can interfere with a patient's normal breathing, blood pressure, and/or heart rate. In rare cases, the patient's symptoms can increase in intensity to where the disorder can become life threatening.

In 8% of cases, the weakness only affects the lower extremities, which can cause paraparesis or even parapalegia. Approximately 1 of 3 people with GBS are able to continuously ambulate. Once the progression of the weakness has ceased, it will persist in a "plateau phase" before improvement occurs (4). The plateau phase can persist for a time period of 2 days to 6 months. Pain-related symptoms may affect more the 50% of people with GBS (4).

The pathophysiology of GBs is based upon an autoimmune disorder that attacks the myelin sheath covering nerve fibers. Anti-ganglioside antibody tests are often performed, but are not specific in their confirmation of a diagnosis of GBS (5). Generally, plasma tests are performed to exclude the possibility of another cause for weakness, such as a low sodium and potassium serum levels (6). Magnetic resonance imaging (MRI) of the spinal cord is performed to discern between GBS and other conditions causing lower extremity weakness, such as compression of the spinal cord. If MRI shows increased signal intensity of nerve roots, GBS may be suspected (3). Lumbar cerebrospinal fluid (CSF) analysis and nerve conduction studies are supportive diagnostic tests commonly performed in the diagnosis of GBS (5). Normal CSF results do not exclude GBS. CSF is unremarkable in 50% of people with GBS within the first few days of the onset of symptoms and 80% after the first week of onset of symptoms (3).

GBS is first managed using immunotherapy. Plasmapheresis and intravenous immunoglobulins (IVIg) are often used in combination. Both immunotherapy treatments are equally efficacious, irrespective of combining both treatments or if administered on their own (7).

Although pain is prevalent in patients with GBS, there is currently no standard recommended type of analgesic for treatment (8). Following the acute symptomatic phase, approximately 40% of patients with GBS require intensive rehabilitation via a multidisciplinary team (9). The primary focus is on improving activities of daily living (9).

CASE PRESENTATION

The patient is a 47-year-old female with a history of non-insulin dependent diabetes mellitus who presented in July 1999 with a chief complaint of sudden and progressive numbness and tingling to the left foot and heel pain. She had a past medical history of GBS diagnosed 5 years previous. She stated her blood glucose was well controlled with metformin. She stated her symptoms started 3 months prior to her first visit to our clinic. She denied any history of head trauma, stroke, alcohol or drug abuse. On the first physical examination, she demonstrated sufficient protective threshold after evaluation with a Semmes-Weinstein 5.07g monofilament. All of her peripheral pulses were palpable to

bilateral lower extremities. She was diagnosed with painful peripheral neuropathy and plantar fasciitis. Her initial treatment plan was Kenalog-10 corticosteroid (injected) for bilateral heel pain and topical capsaicin for neuropathic pain management.

The patient returned 2 months later for follow-up, and stated her neuropathic pain had improved 25% from her last visit using the capsaicin. In addition, she stated her over-the-counter orthotics improved her heel pain. Physical examination of her neuropathy demonstrated that she had now lost protective and epicritic sensation to the first and second left toe. At the time, the plan was to control her blood glucose tightly in concert with her primary care physician and to prescribe extra-depth, extra-wide diabetic shoes to prevent complications from her neuropathy. The hope was tighter glucose control would improve her neuropathic pain to the left foot.

At the next visit, one month later, the patient reported neuropathic pain and tingling to the right fifth toe. Her heel pain had resolved by this point, and her chief complaints were limited to the now bilateral neuropathic pain. The patient developed callosities secondary to pressure she could no longer sense to the right foot. Conservative management with callus debridement and custom inserts for her shoes was provided.

One month later, she returned to the clinic, now with pain described as burning in the right foot, and spreading from the toes, to now the plantar aspect of the right foot over the heel. A triple-phase bone scan was ordered and demonstrated an acute inflammatory process to the right heel. MRI studies revealed a possible stress fracture to the right calcaneus with high signal intensity within the heel. Hemoglobin A1C was ordered and demonstrated tight blood glucose control. A rocker-bottom type boot was prescribed to off-load the heel, and the patient was instructed to return for follow-up.

The patient returned in 4 weeks with no worsening or improvement in her right heel pain. At this time, the patient has numbness in both feet, and a possible stress fracture to the right calcaneus. A rheumatoid panel, sickle cell panel, and uric acid were ordered, and none were contributory to elucidating the etiology for the patient's spreading neuropathic pain. Her hemoglobin A1C was 8.2 mg/dl. Therefore, she was diagnosed with diabetic peripheral neuropathy and arthropathy. Accommodations using plastazote were made to the patient's rocker-bottom boot and provided 2 months of heel pain relief.

Once her heel pain resolved 8 months in total after the original diagnosis of right heel stress fracture, she initiated physical therapy. Her waxing and waning neuropathic pain became concerning for possible diabetic neuropathic bone changes consistent with Charcot neuroarthropathy. Over the next 2 years, conservative management with sharp callus debridement, custom-molded diabetic shoes with custom orthotics, and regular visits to her podiatrist modified her pain to reach a steady-state that she could live with. She was able to continue with her regular activities of daily living. There were no progressive radiographic changes to suggest any surgical intervention.

After that improvement period in her status of 2 years, she came to our clinic with left ankle and calf burning pain. The patient stated she experienced a transient ischemic attack but it resolved without any obvious physical limitations other than neuropathy, which was present before that incident. An arterial and venous ultrasound study was performed and showed no vascular pathology to contribute to her presentation. Observation, 81 mg aspirin therapy and anti-inflammatories were sufficient to resolve her ankle and calf pain.

The progression of her neuropathy made our office concerned for a proximal etiology for her neuropathic pain at the level of the spinal cord. Lumbar MRI was ordered and revealed stenosis of the L4-L5 neural foramen and compromise of the central canal. She was referred to her primary care physician, who diagnosed the patient with GBS. Our office referred the patient to physical therapy specialists to prevent progression of the patient's disease.

Our office would see this patient on a regular basis, and conservative management of her neuropathy and neuropathic complications prevented quick progression of her symptoms. She returned to our clinic every 3 months for 15 years and to date she has not required surgical intervention. A combination of physical therapy, custom-molded shoes, and oral anti-inflammatories has afforded this a patient a reasonable quality of life given her diagnosis of GBS.

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