CLINICAL VIGNETTE

Guillain-Barré

Angela Ruman, M.D.

Case Report

A 31-year-old Caucasian female presented to my office for Bell's palsy follow-up. She had recently been seen in the ER where the clinical diagnosis of right Bell's palsy was made and was started on oral steroids and valacyclovir.

The patient noted initial improvement following steroid treatment but reported worsening weakness of the right face associated with slight weakness in left face, bilateral lower extremity "pins and needles sensations" starting in the feet and moving up the legs to her hips, neck and back pain, and difficulty walking due to leg weakness.

The patient did not have any significant past medical or surgical history but reported an episode of food poisoning one week prior to her facial weakness. She denied any allergies to medications, and she was not taking any additional prescription or non-prescription medications. She was recently married and denied tobacco, alcohol or drug use and did not have any significant family medical history. Her exam was remarkable for blood pressure 120-140 systolic over 70-100 diastolic, HR 106 with normal temperature.

Her neuroexam revealed:
CN: Facial asymmetry with weakness of both the upper and lower halves of the right face. The uvula elevates midline. Head turn and shoulder shrug are 5/5 strength bilaterally and the tongue protrudes midline. Sensation was intact to light touch and pinprick in V1 V2 V3 distribution.
Motor Exam: Normal tone, normal bulk, strength is 5/5 proximally and distally in the upper extremities. In the lower extremities, the patient's strength is 4/5 proximally and distally. Sensory Exam: The patient is intact to light touch and proprioception throughout. Pinprick is diminished in a stocking pattern in the lower extremities bilaterally. Reflexes are 3+ bilaterally in biceps, brachioradialis and absent with patellar tendons and ankle jerks. Hoffman's is negative and the toes are down going.

Laboratory Data

The patient's laboratory data included a white count of 11.92 with normal Hgb, platelets, chemistries and routine urinalysis. She was admitted to the hospital given the progressive nature of her neurologic deficits.

Imaging

A contrast enhanced MRI of her C-spine was unremarkable. A contrast enhanced MRI of her brain showed bilateral leptomeningeal enhancement of upper cervical and facial nerves. A spinal tap was performed; revealing elevated CSF protein levels with normal WBC and RBC counts, suggestive of an inflammatory demyelinating polyneuropathy.

Hospital Course

She was started on a course of Intravenous Immune Globulin at 2.0 mg/kg divided over three days. An EMG study was consistent with demyelinating polyneuropathy. Her right-sided facial weakness and loss of sensation had minimal improvement during admission, but the weakness in her proximal leg muscles appeared to resolve, with decreased pain and imbalance during gait. She was discharged with instructions for a neurology follow-up as well as physical therapy.

Discussion

Guillain Barré Syndrome is heterogeneous grouping of immune-mediated processes generally characterized by motor, sensory, and autonomic dysfunction. In its classic form, GBS is an acute inflammatory demyelinating polyneuropathy characterized by progressive symmetric ascending muscle weakness, paralysis, and hyporeflexia with or without sensory or autonomic symptoms[2].

Guillain Barré Syndrome starts with a progressive phase, which lasts from a few days to four weeks. About 73 percent of patients reach a nadir of clinical function at one week and 98 percent at four weeks. The progressive phase is followed by a plateau phase of persistent, unchanging symptoms. Improvement begins within days of the plateau. The time to resolution of symptoms varies among patients[2].

Pain is another common feature of GBS, and it is seen in approximately 50% of patients and is sometimes described as severe. Pain is most severe in the shoulder girdle, back, and posterior thighs[2].
Two-thirds of patients who develop GBS have diarrhea or a respiratory illness several days or weeks before onset of symptoms. Campylobacter jejuni infection is one of the most common risk factors for GBS. GBS also develops after influenza or other viral infections such as cytomegalovirus and Epstein Barr virus. On very rare occasions, GBS may develop in the days or weeks after getting a vaccination[1, 2].

Some theories suggest an autoimmune mechanism, in which the patient's defense system of antibodies and white blood cells are triggered into damaging the nerve covering or insulation, leading to weakness and abnormal sensation.
An estimated 3,000 to 6,000 people develop GBS each year on average, and the incidence of GBS is reported to be 1.2–2.3 per 100,000 per year[1].

Plasma exchange therapy and intravenous immune globulin are effective for treatment of Guillain Barré syndrome. They may decrease autoantibody production and increase solubilization and removal of immune complexes. Both have been shown to shorten recovery time by as much as 50%. IVIG is easier to administer and has fewer complications than plasma exchange. The cost and efficacy of each are comparable. Combining PE and IVIG neither improved outcomes nor shortened illness duration[3,4].

Approximately 85 percent of patients with GBS achieve a full and functional recovery within six to 12 months[2]. The lesson learned from this case was to perform a full neurologic exam and pay careful attention to vital signs. Autonomic instability was present given the high blood pressure readings in addition to a more progressive neurologic condition affecting more than just cranial nerve VII.

REFERENCES

1. **Kuwabara S**. Guillain-Barré syndrome: epidemiology, pathophysiology and management. *Drugs*. 2004;64(6):597-610. Review. PubMed PMID: 15018590.
2. **Newswanger DL, Warren CR**. Guillain-Barré syndrome. *Am Fam Physician*. 2004 May 15;69(10):2405-10. Review. PubMed PMID: 15168961.
3. **Shahar E**. Current therapeutic options in severe Guillain-Barré syndrome. *Clin Neuropharmacol*. 2006 Jan-Feb;29(1):45-51. Review. PubMed PMID: 16518134.
4. **Hughes RA, Swan AV, Raphaël JC, Annane D, van Koningsveld R, van Doorn PA**. Immunotherapy for Guillain-Barré syndrome: a systematic review. *Brain*. 2007 Sep;130(Pt 9):2245-57. Epub 2007 Mar 2. Review. PubMed PMID: 17337484.

Submitted on October 28, 2011