

Guillain Barre

Short Case:

Patient is a 55 y/o F with PMH of HCV and depression who presents to the ED from outside ED for evaluation of lower extremity weakness with concern for Guillain Barre syndrome (GBS). Patient states that she has had 1 week of increasing numbness and tingling in the lower extremities and over the past 2-3 days is associated with progressive weakness in her legs as well. She denies fever, recent illness, n/v/d/c, chest pain, or difficulty breathing. On exam she has 3+ strength in the b/l LEs, absent patellar reflexes, and decreased sensation to light touch. She is unable to ambulate due to weakness. Lumbar puncture performed at OSH is unremarkable.

After discussion with my attending, I posed the question, “isn’t GBS a purely motor deficit? How can it be GBS if she has sensory deficits as well”. And like so many before her, her response was “maybe you should read a little about it.” And that’s what I did.

Classic Guillain Barre Syndrome:

GBS is one of the most frequent causes of acute flaccid paralysis and is a serious neurologic emergency. The GBS that we are all taught about in med school is the type characterized by progressive, usually symmetric, ascending weakness, mostly preceded by an illness or infection. Weakness is usually accompanied by absent or decreased reflexes and paralysis can eventually progress to include any muscle group. Paresthesias frequently accompany these symptoms but sensory abnormalities are generally mild and not profound. Disease usually progresses over a course of 2 weeks, and by 4 weeks since onset, symptoms usually reach a peak. Patients can also have autonomic dysfunction with resultant tachy/bradycardia, hyper/hypotension, arrhythmias, and urinary retention. It also is interesting to note that fever should be absent at the onset of symptoms.

Lumbar puncture should be performed in patients with suspected GBS, and the classic finding is elevated protein with normal white blood cell count (albuminocytologic dissociation). This can be seen in the majority of patients 1 week after symptom onset. Other laboratory tests can be obtained, but are not necessary for the diagnosis.

GBS variants:

The Miller Fisher syndrome (MFS) usually presents with ophthalmoplegia, ataxia, and areflexia. Unlike what we were taught in medical school, only about one quarter of these cases will develop extremity weakness. The main distinguishing characteristic of this variant is its oculomotor involvement. Additionally, another variant called Bickerstaff encephalitis is similar to MFS in that there is ophthalmoplegia and ataxia, but different in that there is encephalopathy and hyper-reflexia. You can test for these variants by testing serum IgG antibodies to GQ1b.

Acute motor axonal neuropathy (AMAN) and acute motor and sensory axonal neuropathy (AMSAN) are similar to the classic variant but are almost always preceded by C. jejuni infection. In AMAN, sensory nerves are not affected and reflexes tend to be

preserved. AMSAN is the more severe variant in which motor and sensory fibers are affected with axonal degeneration that leads to delayed and incomplete recovery

Finally, the pharyngeal-cervical-brachial variant is characterized by weakness of the oropharyngeal, neck, and shoulder muscles, usually with associated difficulty swallowing. There is also usually accompanying facial weakness. The main distinguishing factor of this variant is that there is no lower extremity weakness or hyporeflexia.

Treatment:

While patients remain in the ED, the mainstay of treatment is supportive care. Many of these patients can rapidly progress to respiratory failure, it is important that they are watched closely and supported appropriately. **Do not hesitate to intubate if needed.** Vital signs should also be monitored closely, because as discussed earlier, autonomic dysfunction is not uncommon.

Plasmapheresis or IVIG administration are also mainstays of treatment. Plasmapheresis works by removing circulating antibodies and complement, and studies have shown that the optimal number of exchanges is 4 times over 8-10 days. Plasmapheresis also has better efficacy if started within 7 days of symptom onset. It has also been shown that IVIG is just as effective as plasmapheresis in treatment and there is no benefit to combined therapy with both of them. IVIG is usually administered at 0.4g/kg/d. Choice of treatment usually comes down to availability, but IVIG is much easier to administer. Additionally, it was previously thought that steroids played a role in treatment of GBS, but this is no longer the case and they are not recommended.

References:

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2. Vriesendorp, FJ. *Treatment and Prognosis of Guillain Barre Syndrome in Adults*. In: UpToDate, TePas E (Ed), UpToDate, Waltham, MA. Available at: https://www.uptodate-com.foyer.swmed.edu/contents/treatment-and-prognosis-of-guillain-barre-syndrome-in-adults?source=search_result&search=guillain+barre&selectedTitle=3~150 Accessed May 30, 2015
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