Trailer

Last year, my grandmother had a group of friend's visiting. When they left, she took a nap and woke up with no muscle function in her whole right side and was unable to open her eyes. Respiratory distress came later on within the hour. The sudden onset of Guillain-Barré Syndrome which didn't allow any of her nerves to send the signals to her brain to move. She went through intense therapies, medications, and rehab to gain the strength back. I work in the Neuro Intensive Care Unit at UVA and am/was very interested in her condition as it deteriorated her so rapidly with no onset warnings. I work in the Neuro ICU at UVA Medical Center and see other patient's come in and out of our unit with Guillain-Barré and all of their results vary so drastically. I would love to look into this topic more.

Guillain–Barré Syndrome

Personal Story:

My grandmother has never been one to sit down and enjoy a peaceful afternoon. An active life has always been a set standard for her through years of working in a factory, active work within the community conducting vegan food preparation classes, planning trips for seniors, as well as maintaining a home and garden. One afternoon, she invited friend's over for dinner. Post them leaving, she took an afternoon nap, which was completely out of her character. Upon waking up, she was unable to move her entire right side and had limited mobility with both legs. Her eyes were beginning to swell shut and all speech was beginning to slur. She managed to call my mother and immediately take her to the local doctor's office as they may have thought it was an onset of stroke or allergic reaction. The doctor immediately sent her to the Emergency room where upon arriving within 30 minutes had lost complete right side function and was unable to walk or see. The medical center admitted her directly into the ICU for respiratory distress and intubated her within minutes of her arrival. Upon a battery of tests conducted throughout the night, the doctors were still completely positive of a direct diagnosis for her as the onset was so sudden. At the end a week diagnostic's concluded that she had a rare type of Guillain-Barré syndrome (GBS).

Anatomical & Physiological Influences:

Typically, patients diagnosed with GBS will present with acute inflammatory demyelinating polyradiculoneuropathy (AIDP) within 2-4 weeks after having a respiratory or gastrointestinal illness with complaints of finger dysesthesias and proximal muscle weakness of the lower extremities.The weakness may progress over hours to days to involve the arms, truncal muscles, cranial nerves, and muscles of

respiration. Variants of GBS may present as pure motor dysfunction or acute dysautonomia. (Hughes RA)

Lower extremity weakness usually begins first and ascends symmetrically and progressively over the first several days. Upper extremity, trunk, facial, and oropharyngeal weakness is observed to a variable extent. Frequent complaints of paresthesias, objective sensory changes are minimal. Reflexes are absent or reduced early in the disease course. Hyporeflexia or areflexia of involved areas represents a major clinical finding on examination of the patient with GBS. Pathologic reflexes, such as the Babinski sign, are absent. Hypotonia can be observed with significant weakness. Facial weakness (cranial nerve VII) is observed most frequently, followed by symptoms associated with cranial nerves VI, III, XII, V, IX, and X. Involvement of facial, oropharyngeal, and ocular muscles results in facial droop, dysphagia, dysarthria, and findings associated with disorders of the eye (Andary). Ophthalmoparesis may be observed in up to 25% of patients with GBS. Limitation of eye movement most commonly results from a symmetrical palsy associated with cranial nerve VI. Ptosis from cranial nerve III (oculomotor) palsy also is often associated with limited eye movements (Andary).

Causes:

The exact cause of Guillain-Barre syndrome isn't known. The disorder usually appears days or weeks after a respiratory or digestive tract infection. Rarely, recent surgery or immunization can trigger Guillain-Barre syndrome. In Guillain-Barre syndrome, your immune system — which usually attacks only invading organisms — begins attacking the nerves (Mayo Clinic). In the Peripheral Nervous System, neuroglia called Schwann Cells. These cells surround large axons of peripheral neurons in lipid-rich sheaths. They are tight coverings at have layers of cell membranes and wrap around the axon like a bandage, called Myelin (Shier). In AIDP, the most common form of Guillain-Barre syndrome, the myelin sheath is damaged. The damage prevents nerves from transmitting signals to your brain, causing weakness, numbness or paralysis. Some other causes that may GBS may be triggered by include Influenza virus, Epstein-Barr virus, HIV, Mycoplasma pneumonia, surgery, Hodgkin’s lymphoma, and most commonly campylobacter(undercooked poultry)(Mayo Clinic).

Symptoms and Signs:

Symptoms of GBS can get worse quickly. It may take only a few hours for the most severe symptoms to appear. Muscle weakness or loss of muscle function (paralysis) affects both sides of the body and usually starts in the legs and spread to the arms. If the inflammation affects the nerves of the diaphragm and chest and there is weakness in those muscles, the person may need breathing assistance.

Other signs and symptoms may include muscle tenderness or pain that could feel like a cramp, tingling or numbness where you may have a loss of sensation, loss of tendon reflexes in the arms and legs, uncoordinated movement, abnormal heart rate, and/or low blood pressure or poor blood pressure control. One needs to be aware also if there's onset of blurred vision, double vision, clumsiness and/or falling, difficulty moving facial muscles, muscle contractions, or if they can feel palpitations of heart beat sensations. More drastic emergency symptoms would include not being able to take a deep breath, if their breathing temporarily stops, having difficulty breathing or swallowing, drooling, fainting, or feeling light headed when they stand (Jasmin).

Diagnosis/Differential Diagnosis Options:

As previously mentioned, the medical center took roughly a week to correctly diagnose my grandmother's condition that was potentially fatal. There are many specific types of GBS, however, some of the main include **Acute inflammatory demyelinating polyradiculoneuropathy (AIDP),** the most common form in the U.S. The most common sign of AIDP is muscle weakness that starts in the lower part of your body and spreads upward. Another includes, **Miller Fisher syndrome (MFS),** in which paralysis starts in the eyes. MFS is also associated with unsteady gait. MFS occurs in about 5 percent of people with Guillain-Barre syndrome in the U.S. but is more common in Asia. This is what she was ultimately diagnosed with. There's also **Acute motor axonal neuropathy (AMAN)and acute motor-sensory axonal neuropathy (AMSAN)**, which are less common in the U.S. but more frequent in China, Japan and Mexico.(Mayo Clinic)

Regarding a differential diagnosis, some patients with atypical presentation, incomplete weakness, and some inconsistencies in their physical examination are frequently diagnosed as having a psychological reaction or hysteria, and the diagnosis is very difficult. They are frequently sent home from the emergency department and then return with persistent or progressive symptoms hours to days later (Hughes RA).

- Guillain-Barré syndrome (GBS) is generally diagnosed on clinical grounds. Basic laboratory studies, such as complete blood counts (CBCs) and metabolic panels, are normal and of limited value in the workup.
- Sometimes, Electromyography (EMG) and nerve conduction studies (NCS) can be very helpful in the diagnosis. Abnormalities in NCS that are consistent with demyelination are sensitive and represent specific findings for classic GBS. Delayed distal latencies, slowed nerve conduction velocities, temporal dispersion of waveforms, conduction block, prolonged or absent F waves, and prolonged or absent H-reflexes are all findings that support demyelination. Needle EMG may be normal in acute nerve lesions, and it may take 3-4 weeks for fibrillation to develop. In the acute phase, the only needle EMG abnormality may be abnormal motor recruitment, with decreased recruitment and rapid firing motor units in weak muscles. Unfortunately, electrodiagnostic studies can be completely normal in acute GBS and a normal study does not

rule GBS. Frequent evaluations of pulmonary function parameters should be performed at bedside to monitor respiratory status and the need for ventilatory assistance.

- Lumbar puncture for cerebrospinal fluid (CSF) studies is recommended. During the acute phase of GBS, characteristic findings on CSF analysis include albuminocytologic dissociation, which is an elevation in CSF protein (>0.55 g/L) without an elevation in white blood cells. The increase in CSF protein is thought to reflect the widespread inflammation of the nerve roots.
- Imaging studies, such as Magnetic Resonance Imaging (MRI) and Computed Tomography (CT) scanning of the spine, may be more helpful in excluding other diagnoses, such as mechanical causes of myelopathy, than in assisting in the diagnosis of GBS (Albers).

Treatment:

Patients who are diagnosed with GBS should be admitted to a hospital for close monitoring until it has been determined that the course of the disease has reached a plateau or undergone reversal. Although the weakness may initially be mild and nondisabling, symptons can rapidly change over just a few days. Approximately one third of patients require admission to an ICU, primarily because of respiratory failure. After medical stabilization, patients can be treated on a general medical/neurologic floor, but continued vigilance remains important in preventing respiratory, cardiovascular, and other medical complications. Continued care also is needed to minimize problems related to immobility, neurogenic bowel and bladder, and pain. Early recognition and treatment of GBS also may be important in the long-term prognosis, especially in the patient with poor clinical prognostic signs, such as older age, a rapidly progressing course, and antecedent diarrhea.

Immunomodulatory treatment has been used to hasten recovery. Intravenous immunoglobulin (IVIG) and plasma exchange have proved equally effective. Corticosteroids (oral and intravenous) have not been found to have a clinical benefit in GBS.Consequently, this class of drugs is not currently employed in treatment of the syndrome. A few studies have investigated other medications to treat GBS; however, the trials have been small and the evidence weak, highlighting the need for further investigation of potential treatment options (Visser). The decision to use immunomodulatory therapy is based on the disease's severity and rate of progression, as well as on the length of time between the condition's first symptom and its presentation. Risks, such as thrombotic events associated with IVIG, should be taken into consideration. Patients with severe, rapidly progressive disease are most likely to benefit from treatment, with faster functional recovery. (Jaysena RA)

Personal Impact:

With my grandmother's case, she never experienced a sense of fatique and was rare to take afternoon naps. There's an issue of persistent fatigue after recovery from GBS. Studies have suggested that a large percentage of patients continue to have fatigue-related problems, subsequently limiting their function at home and at work, as well as during leisure activities. Treatment suggestions range from gentle exercise to improvement in sleep patterns to relief of pain or depression, if present (Garssen). I know that generally speaking, I am not in the statistical range to experience GBS, however it doesn't take one moment for me to think about the sudden onset with no potential warning signs that she endured to play a substantial role in my own life with the current amount of sleep deprivation of having one child and being pregnant with another. GBS can strike anyone, regardless of age or sex. GBS is not contagious, but it may follow a bacterial or viral infection, such as campylobacterial infection (caused by a bacteria found in undercooked food, especially poultry) or Epstein-Barr virus (EBV). Although GBS is not inherited, there may be a genetic predisposition to the disease (The Foundation For Peripheral Neuropathy). I currently work in the Neuro Intensive Care Unit at UVA Medical Center and see cases such as my grandmother's so few and far between that it strikes my interest to know additional causes, as there are none currently known.

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