A Case of Atypical Presentation of Guillain-Barré Syndrome Concomitant with Severe B12 Deficiency and Spinal Stenosis

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Abstract

Guillain-Barré Syndrome (GBS) is an acute immune-mediated inflammatory polyneuropathy, often provoked by a preceding infection.

We present a case of atypical GBS with concomitant B12 deficiency and severe spinal canal stenosis with impingement and clumping of the cauda equina in a 70-year-old male, with a past medical history significant for hypertension, asthma, and atrial flutter who presented with lower extremity and right-hand weakness. After two weeks of an extensive workup, discussions with countless physicians, and a planned transfer to another hospital, a repeated lumbar puncture (LP) gave the diagnosis of GBS, and plasmapheresis treatment was initiated.

A missed diagnosis of GBS delays care and increases mortality and morbidity. This unique case reinforces the importance of considering concomitant diagnoses alongside a main diagnosis to prevent financial, health, and psychological impacts on patients. Although the initial workup for GBS was negative, it is known that the acuity of symptoms influences the findings on LP. The patient was found to have severe lumbar stenosis and severely low vitamin B12 levels, with the notable exception of any anemia; however, his symptoms could not have been fully explained by those findings, so initiating treatment with scheduled laminectomy was premature considering the lack of clinical foundation. Furthermore, this case enforces the importance of knowing the limitations of diagnostic tests, as many highly specific tests for conditions may be negative if performed too early following symptom onset.

This case highlights the challenges involved in diagnosing GBS with concomitant conditions, such as B12 deficiency and severe spinal canal stenosis. Nonetheless, it is imperative to investigate beyond the initial diagnosis to ensure accurate diagnosis and appropriate treatment management.

Introduction

Guillain-Barré syndrome (GBS) is an immune-mediated acute inflammatory polyneuropathy with an incidence in Europe and North America reported to be 0.8 - 1.9 per 100,000 person-years. It is classically associated with molecular mimicry due to *Campylobacter Jejuni*, but has also been associated with various other infectious materials. The pathophysiology is theorized to encompass antibodies and lymphocytes, where the antibodies target axons and/or myelin, and the lymphocytes work to damage the axons that are targeted by the lymphocytes. GBS can be subdivided into acute motor and sensory axonal neuropathy (AMSAN), acute motor axonal neuropathy (AMAN), and acute inflammatory demyelinating polyneuropathy (AIDP), but the symptoms between each are similar.[1] The most common symptoms of GBS are paraesthesias, flaccid weakness, difficulty with facial movements and swallowing, trouble with bowel and bladder control, and severe pain.[2]

Disease Topic

Although every medical student is required to know the classic presentation of GBS, many presentations of GBS are non-classical.

Physicians teaching students must realize that, although important to know for board exams, knowing only the classical presentation can lead to pitfalls in diagnosis, and wasted money and resources to the patient and the rest of the healthcare system.

All physicians must keep a broad differential to ensure that the obscure presentations of various conditions are not overlooked, so diagnostic errors do not occur.

Many diseases may present non-classically, and the ability to recognize inconsistencies in the non-classical presentations ensures that diagnostic accuracy remains at an appropriate level.

Patient Presentation

A 70 year old man with past medical history significant for hypertension, hyperlipidemia, generalized anxiety disorder, atrial flutter s/p ablation, BPH presented with the chief complaint of left lower extremity right upper extremity weakness that started 1 day prior. HE was unable to rise from the toilet due to these symptoms, and it also woke him from his sleep. He denied any falls or trauma and states he has chronic pain at baseline and is able to perform all ADLs independently.

In the emergency department he was hypertensive at 141//90, tachycardic with a pulse of 105, afebrile, respiratory rate of 18, and spo_2 of 99%. Pertinent physical exam findings included LLE motor strength of 4/5 and left foot drop. A CTA head and neck, CT head, MRI cervical, thoracic, lumbar spine were ordered. Significant findings included MRI results showing severe spinal canal stenosis with impingement and clumping of the cauda equina at L2 and L3 with evidence of multilevel degenerative disc disease. Neurosurgery was consulted and a L2-L5 decompressive lumbar laminectomy with in situ fusion was scheduled for that week.

As he remained in the hospital, his weakness was progressing distal to proximal and he had severe pain rated 14/10 that was not relieved by pain medications. Neurology was consulted and evaluated the patient exploring the differential diagnosis of GBS or b12 deficiency. New physical exam findings at this time included decreased grip strength, notable right shoulder pain, and decreased lower extremity motor strength and foot drop bilaterally. Lumbar puncture results were not consistent with the potential diagnosis of guillain barre syndrome, and b12 was found to be critically low at 67 and b12 injections were initiated for 3 days. Due to the these findings potentially being the cause of his symptoms, the laminectomy was not recommended and subsequently canceled and PT/OT was initiated.

Despite the injections, his B12 continued to drop (67→1400→600). Due to the continued rapid drop, IM B12 was continued for 7 more days. Upon further workup he was found to have intrinsic factor antibodies, however it could be a false positive as he received IM B12 the day before and it still did not explain the clinical picture. Myopathy was ruled out by negative anti-jo1, anti-ro1, anti-la, ANA, ESR. CPK was found to be elevated. At this time the patient's symptoms were improving and a plan to discharge to inpatient rehab was being discussed amongst the care team.

Within days the patient was experiencing excruciating pain, unrelieved by pain medication and the continuation of narcotics due to the risk of addiction was discussed. Rheumatology and Hematology were consulted to assist in making the diagnosis and more extensive labs were drawn. EMG was performed and showed classic length dependent axonal demyelinating neuropathy. Due to there being no clear diagnosis despite continuous and thorough evaluation and testing, escalation of care and transfer to a different hospital was planned.

Prior to transfer, repeat lumbar puncture was done 10 days after the initial one and showed cytoalbuminologic dissociation consistent with GBS. The patient then received 5 days of plasmapheresis and was admitted to inpatient rehabilitation following the completion of treatment.

Results

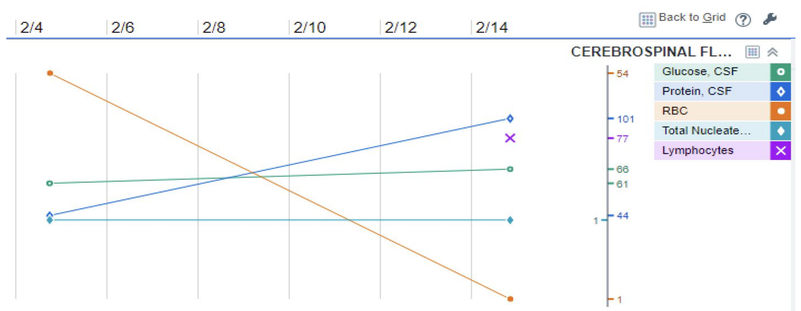

Figure 1. Cerebrospinal Fluid Analysis on 2/4 vs. 2/14

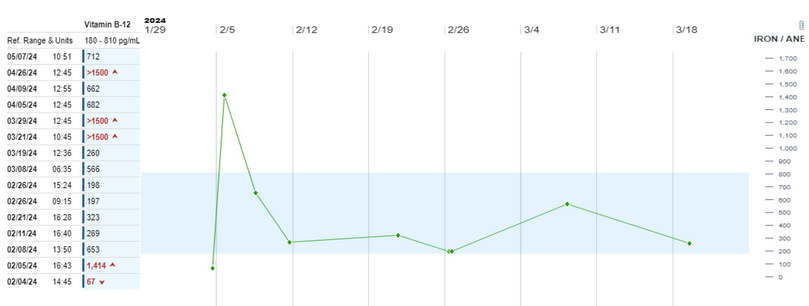

Figure 2. Vitamin B12 Levels

Discussion

A missed diagnosis of GBS delays care and may increase mortality and morbidity.[3] Several factors, such as neuropathic pain, asymmetric pattern of weakness, intact reflexes, the involvement of cranial nerves, and the lack of a neurological evaluation, have been noted to delay diagnosis and treatment.[4] Therefore, early neurological evaluation and treatment is imperative in mitigating the potential consequences associated with GBS.

Nevertheless, the timing of symptoms may influence the findings on LP and, thus, pose challenges to obtaining an accurate diagnosis. Albuminocytological dissociation is characteristic of GBS; however, it is only present in ~64% of patients with GBS.[5] Additionally, only 50% of patients have increased CSF protein levels within the first three days following the onset of symptoms, compared to 80% after the first week.[5] Therefore, while LP findings may help support the diagnosis of GBS, they should not be solely relied on for the diagnosis.

Moreover, patients may also present with concomitant syndromes, as seen by this patient with pernicious anemia and severe spinal canal stenosis. These conditions have the potential to mask the symptoms of GBS and further delay diagnosis. Thus, it is critical to investigate beyond the initial diagnosis to ensure accurate diagnosis and treatment management.

Ultimately, these circumstances can adversely affect patients by prolonging their hospital stays, increasing testing, and causing distress due to the lack of a diagnosis. A person is a unit of the body, mind, and spirit; therefore, healthcare professionals must consider all aspects of the human being when providing quality care. Hence, this unique case reinforces the importance of considering concomitant diagnoses alongside a primary diagnosis to prevent financial, health, and psychological impacts on patients.

References

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