

Amsan Variant of Guillain-Barrésyndrome: A Case Report

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Abstract: *Guillain-Barré syndrome (GBS) is a rare and potentially life-threatening medical condition resulting from an individual's immune system's attack on the peripheral nerves. It is the result of an auto-immune response leading to axonal degeneration or demyelination. Acute motor and sensory axonal neuropathy (AMSAN) is a variant of GBS with the poorest prognosis. It is characterized by rapid-onset motor weakness, loss of deep tendon reflexes and sensory changes. Immunotherapy is considered a standard treatment for GBS. Here, the reported case is of AMSAN variant of GBS in a 42-year-old patient, who developed bilateral facial nerve palsy and treated with intravenous immunoglobulin (IVIg).*

Keywords: immune system, deep-tendon reflexes, lower and upper limbs, muscle movement

1. Introduction

Guillain-Barré syndrome (GBS) is a rare and potentially life-threatening medical condition resulting from an individual's immune system's attack on the peripheral nerves. It is an acute peripheral neuropathy that results in tingling sensation and muscle weakness, particularly in the limbs, of abrupt onset. GBS is often preceded by viral or bacterial infection. The symptoms gradually spread to the upper body. The symptoms can quickly progress over a few hours or can take a few weeks to develop. Sometimes, the disease is also associated with changes in sensory functions resulting from alterations in autonomic nervous system responses. Acute motor and sensory axonal neuropathy (AMSAN) is a variant of axonal GBS characterized by acute onset of motor weakness, reduction of deep tendon reflexes and sensory abnormalities. Annually, GBS affects between 1 out of 91,000 and 1 out of 55,000 people. AMSAN and acute motor axonal neuropathy (AMAN) variants of GBS account for only 3-5% of cases of GBS in the West but are more frequent (30%-50% of GBS cases) in Asian and Latin American countries.¹ The case is of AMSAN variant of GBS with bilateral facial nerve palsy in a 42-year-old patient.

2. Case Report

A 42-year-old male patient was presented to casualty with complaint of difficulty in swallowing solid foods. He was then transferred to the intensive care unit where he reported weakness in both lower limbs for 3 days. He was unable to stand from squatting position and frequently fell while walking. The patient had mild difficulty in gripping slippers while walking. He also reported tingling numbness in toes and fingers. Vitals examination revealed that the patient was hypertensive (160/90 mmHg). Romberg's sign was positive and the strength in both lower limbs was less (4/5). The patient was diagnosed with AMSAN variant of Guillain-Barré syndrome. He was switched to liquid foods and administered intravenous immunoglobulin (IVIg) at 0.4 mg/kg/day for 5 days. His condition improved over the next two days; however, the patient complained of rashes 2 days following first IVIg transfusion. Moreover, the patient acutely developed facial paralysis on both sides with slurred

speech. Cerebrospinal fluid (CSF) analysis revealed high protein in CSF (896.6 mg/dl), suggesting nerve inflammation. Glucose in urine and CSF was also higher than the normal ranges. His blood pressure improved after amlodipine (5 mg) and telmisartan (40 mg) therapy was initiated. Patient's random blood glucose (RBS) level was high (314 mg/dl). A rapid-acting insulin was administered to control blood glucose level. His RBS level improved during the course of his hospital stay. There was no respiratory involvement. During hospital stay, the patient revealed that he had been drinking alcohol for 10 years and quit drinking 15 days prior to hospitalization. The attending physician thus suggested ultrasonography to assess his liver condition. USG revealed diffuse fatty liver with mild hepatomegaly. However, there was no symptomatic manifestation of alcoholic liver disease. The patient's muscle power improved after 12 days, with recovery of normal strength in both lower limbs (5/5) and his speech and facial movements also improved. He was able to walk without difficulty during tandem gait test: suggesting no more sensory involvement and thus, recovery. His blood pressure (126/80 mmHg) and blood glucose (141 mg/dl) level were also reported to be normal prior to discharge. The patient was prescribed amlodipine, pantoprazole, metformin hydrochloride and multivitamin supplements for supportive care, upon discharge. He was asked to report after 15 days.

3. Discussion

Traditionally, GBS is described as a post-infectious, immune-mediated process following *Campylobacter jejuni* infection.² However, it is rarely associated with other causative factors. In our patient, no previous incidence of *C. jejuni* infection was reported. Early symptoms of GBS include pain, tingling sensation and weakness in the limbs. Bilateral facial paresis is common in GBS. Due to the rapid development and progression of symptoms, recovery is slower than the initial damage caused. There is no sure-fire treatment for GBS; however, plasma exchange (PE) and intravenous immunoglobulin (IVIg) have shown benefits in large randomized trials.³ There is an ongoing debate among practitioners regarding the preferred choice of therapy. Various trials comparing IVIg and PE have yielded

conflicting results. The American Academy of Neurology (AAN) guideline on GBS concludes that IVIg is easier to administer and has a safer side-effect profile than PE despite being equally effective.⁴ Hence, many practitioners prefer IVIg as the first choice of treatment. AMSAN is considered to be a rare form of GBS, and it usually has a more serious clinical course and slower recovery than the other variants of GBS. In AMSAN, there is an additional sensory fiber involvement.⁵ In this patient, AMSAN was confirmed and AMAN was ruled out after thorough examination of the clinical features. However, the pathogenesis of AMSAN is not clear in this case. After a thorough examination of the patient's medical history, it was concluded that he did not have any recent infection that could lead to axonal degeneration or demyelination. Further studies for a better pathogenic understanding of AMSAN is suggested. The patient had uncontrolled diabetes mellitus (DM) at the time of admission. It has been demonstrated that DM is likely to influence the clinical and electrophysiological patterns of GBS either by contributing to the pathophysiology of neuropathy or by causing subclinical diabetic neuropathy.⁶Hyperglycemia was managed in the patient effectively and he also responded fairly to IVIg therapy. This emphasize on the need to approach DM in patients with GBS appropriately to hasten the recovery process and prevent further complications. It is also advisable to conduct follow-up on patient's self-care activities and monitor compliance to the prescribed regimen, to minimize the risk of hospitalization and prevent readmissions. The symptoms of DM-related neuropathy and GBS often overlap. The former was ruled out in the patient since the symptoms were acute and progressed rapidly. There is a strong association between hypertension and GBS. A previous study suggests activation of sympathetic nervous system at a peripheral level in certain GBS subjects, leading to sustained hypertension.⁷ In GBS, hypertension has also appeared to be related to increased renin-angiotensin activity.⁸ Management of blood pressure in such patients is thus critical to ensure prevention of hypertension-related neuropathies and organ damage. Our patient presented with elevated blood pressure and it was promptly managed. Treatment with IVIg or PE has not reduced the mortality in this disease. GBS-related complications and prolonged length of stay result in mortality.⁹ Most GBS-related deaths are due to respiratory failure, cardiovascular disturbance, and thromboembolism.¹⁰ No respiratory changes were reported in the patient; nonetheless, his blood pressure was elevated. After treatment was initiated his vitals were checked regularly. Signs of respiratory muscle weakness could worsen the condition. Providing appropriate supportive care is vital to ensure better outcome. Elevated CSF protein levels in GBS is thought to result from the inflammation of nerve roots that leave the spinal cord and pass through CSF. When the nerve roots are inflamed in this disease, proteins leak in to CSF.¹¹ Nerve biopsies are not performed routinely in GBS patients; but they are cardinal in distinguishing the different variants of the disease.¹² Since in this patient the etiopathogenesis of the disease was not clear, a histopathological study could be helpful in determining how the disease manifested. AMSAN has a worse prognosis than other GBS variants. Despite the acute onset and progression of symptoms, it has a long-term impact on the patient's life. Constant follow-up is essential to warrant a good quality of life and prevent readmissions.

4. Conclusion

GBS is a rare disease with a long-term impact on patients' lives. While its diagnosis is usually based on clinical evidence and CSF protein analysis, a detailed examination across other aspects is necessary to gather more evidence and fine-tune the therapeutic approach for better outcomes. Supportive care and counselling are equally crucial to keep in the check the complications. The patients and their carers should be comprehensively briefed about the treatment and warning signs. The primary aim is to minimize the long-term impact of the disease.

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