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. 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. Vital signs 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 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. 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. There is an ongoing debate among practitioners regarding the preferred choice of therapy. Various trials comparing IVIg and PE have yielded
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.

References


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