

Pulse-steroid therapy in a 37-year-old man with acute motor and sensory axonal neuropathy: A case report

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Key Clinical Message

We report a case of 37-year-old man admitted with acute motor and sensory axonal neuropathy (AMSAN) which was treated with pulse-steroid therapy after the plasmapheresis and intravascular immunoglobulin. The improvement of the symptoms of AMSAN after pulse-steroid therapy may represent a therapeutic alternative for this variant of Guillain-Barré syndrome.

KEYWORDS

acute motor and sensory axonal neuropathy, Guillain-Barré syndrome, peripheral neuropathy, pulse-steroid therapy

1 | INTRODUCTION

A 37-year-old Saudi Arabian man was admitted to our hospital with acute weakness and numbness of the face and bilateral lower limbs associated with back pain that had persisted for the preceding 3 days. Based on electrophysiologic data and cerebrospinal fluid findings, he was diagnosed with a variant of Guillain-Barré syndrome (GBS) termed acute motor and sensory axonal neuropathy (AMSAN). Plasmapheresis and intravascular immunoglobulin were administered and achieved some symptomatic improvements, but his symptoms later progressed. Subsequently, he was given pulse-steroid therapy, which dramatically improved his symptoms. GBS is an acute autoimmune polyradiculoneuropathy for which plasmapheresis and intravascular immunoglobulin administration are considered the standard treatment. Its variant, AMSAN, is characterized by acute-onset distal weakness, diminished or absent deep tendon reflexes, and sensory symptoms. In this case report, we describe the dramatic improvement of the symptoms of AMSAN after pulse-steroid therapy, which may represent a therapeutic alternative for this variant of GBS.

Guillain-Barré syndrome (GBS) is a relatively uncommon immune-mediated neurologic disorder characterized by demyelination of the spinal nerve roots and peripheral nerves.

GBS manifests as a rapidly progressive ascending symmetrical limb weakness with or without sensory loss associated with diminished or absent tendon reflexes.¹ Acute motor and sensory axonal neuropathy (AMSAN) is a rare variant of GBS characterized by acute-onset distal weakness, diminished or absent deep tendon reflexes, and sensory symptoms. The prevalence of AMSAN is relatively small: the variant accounts for <10% of all cases of GBS.² *Campylobacter jejuni*, cytomegalovirus, and Epstein-Barr virus infection are the most common triggers of GBS, and in particular AMSAN.³

Guillain-Barré syndrome is diagnosed on the basis of a clinical evaluation, cerebrospinal fluid (CSF) findings, and the results of nerve conduction studies. In association with supportive care, immunotherapy via intravenous immunoglobulin (IVIG) administration or plasmapheresis is the standard treatment for GBS.⁴ Pulse-steroid therapy is not a standard treatment for GBS; however, a minority of patients exhibit a significant symptomatic improvement after steroid administration. In this report, we describe the case of a 37-year-old man with AMSAN who experienced a dramatic improvement of his symptoms after pulse-steroid therapy. The report highlights the potential utility of pulse-steroid therapy in patients unresponsive to the standard management approach.

2 | CASE REPORT

2.1 | Presentation

A 37-year-old Saudi Arabian man with no prior medical conditions was admitted to our hospital with acute numbness and weakness of the face and bilateral lower limbs associated with back pain that had persisted for the preceding 3 days. He had a history of tonsillitis and gastroenteritis, which had resolved 2 weeks before the onset of numbness and weakness. He had suffered a choking episode and was experiencing shortness of breath on exertion. His lower-limb muscle weakness had started distally and ascended over a period of 4 days. Subsequently, he started to have difficulty walking and using the stairs but exhibited no loss of balance or axial weakness. There was no evidence of diplopia or autonomic dysfunction.

2.2 | Signs and laboratory findings

The patient was conscious, alert, and oriented. A systemic examination yielded no remarkable findings, and the patient was afebrile. His higher mental function was intact. The patient had bilateral facial weakness, hypoesthesia, bilateral lower limbs power 2 out of 5, upper limb power 3 out of 5, peripheral diminished sensation all over, diminished reflexes in both the upper and lower limbs. There were no extraocular movement abnormalities or signs of autonomic dysfunction. The results of baseline laboratory investigations were unremarkable. Based on electrophysiologic data and CSF findings, the patient was diagnosed with AMSAN, a variant of GBS.

2.3 | Management

Plasma exchange was initiated; however, the patient's condition worsened, and he started to develop respiratory failure, necessitating intubation and mechanical ventilation in the intensive care unit (ICU). Despite the completion of two cycles of plasma exchange (10 sessions), his symptoms continued to progress. Therefore, the patient underwent IVIG at a dose of 20 mg/kg/d for 5 days. The patient showed only mild symptomatic improvements, and tracheostomy was required. Because the patient's symptoms did not improve as expected, he was given intravenous pulse-steroid therapy at a dose of 1 g/d for 4 days. Subsequently, after the second dose, the patient started to demonstrate significant improvement over all. He started to have significant improvement of lung function thus was extubated, started to set at the edge of the bed, power in the lower limbs improved to be 4+, and he started to walk with assistant. Then, the patient discharged to the ward after 45 days of ICU admission and then discharged home after 37 days in the ward. Total days of hospitalization were 84 days.

3 | DISCUSSION

Guillain-Barré syndrome is an acute-onset peripheral polyneuropathy that usually starts in the legs and progresses proximally. Weakness and sensory loss are the usual presentations of GBS. This weakness often ascends to the respiratory muscles, leading to respiratory failure: up to 30% of patients with GBS require mechanical ventilation.⁵ GBS has four clinical subtypes: acute inflammatory demyelinating polyneuropathy (AIDP); acute motor axonal neuropathy (AMAN); AMSAN; and Miller Fisher syndrome (MFS). AIDP is the most common subtype, affecting 85%-90% of patients with GBS. AMAN is commonly found in China and Japan, and has been shown to have a strong association with *C jejuni* infection. AMSAN is very similar to AMAN; however, it is characterized by sensory fiber involvement. The last variant, MFS, manifests as a triad of ataxia, areflexia, and ophthalmoplegia.

Diagnosis of GBS is based on the findings of a clinical evaluation, CSF testing, and nerve conduction studies. In addition, computed tomography scanning of the brain is essential to rule out raised intracranial pressure. Treatment with IVIG or plasma exchange in combination with supportive care is considered the standard management for GBS.⁶ Many studies have demonstrated no apparent benefit of corticosteroid therapy in the treatment of GBS. A systematic review of six randomized controlled trials involving 195 corticosteroid-treated patients and 187 controls confirmed that there were no significant differences in outcomes between the two groups.⁷ In 2016, a systematic review of studies of moderate-quality evidence conducted by Hughes et al⁸ concluded that corticosteroids alone did not significantly accelerate the recovery or affect the long-term outcomes of patients with GBS. Indeed, according to studies of low-quality evidence, oral steroids delayed recovery. However, in some published case reports, steroid therapy was very effective. In one report, a 27-year-old man with GBS exhibited dramatic symptomatic improvement after the administration of pulse-steroid therapy.⁹ Despite this, the mortality rate of GBS remains high at around 2%-12%: 20% of patients with the condition remain permanently disabled, and 40% require admission for intensive rehabilitation.¹⁰

4 | CONCLUSION

This case highlights the potential utility of pulse-steroid therapy for the treatment of AMSAN, especially in patients unresponsive to IVIG or plasmapheresis. Although pulse-steroid therapy is not a standard treatment for GBS, future research is warranted to determine whether pulse-steroid therapy can be added to the standard treatment of some variants of GBS.

CONFLICT OF INTEREST

We have no conflict of interest to declare.

AUTHOR CONTRIBUTION

RA: involved in writing the case report and submitting the case for publication. SA: involved in writing the case report. MB: involved in editing and supervision.

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