CASE REPORT

The Silent Survivor: A Case of Locked-in State due to AMAN Variant of Guillain-Barré Syndrome with Complete Neurological Recovery

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ABSTRACT

Background: The acute motor axonal neuropathy (AMAN) variant of Guillain-Barré Syndrome (GBS) represents a severe and rapidly progressive form of immune-mediated polyneuropathy. Locked-in syndrome is an exceptionally rare presentation in GBS, characterized by quadriplegia and anarthria with preserved consciousness and eye movements. Early recognition and prompt immunotherapy are crucial for recovery.

Case Presentation: A 62-year-old woman with no prior comorbidities presented with rapidly ascending flaccid paralysis following a diarrheal illness. Within days, she developed quadriplegia and respiratory failure, necessitating invasive ventilation. Neurological examination revealed preserved vertical eye movements with complete limb paralysis, consistent with a locked-in state. Cerebrospinal fluid analysis demonstrated albuminocytologic dissociation, while nerve conduction studies confirmed the AMAN variant. The patient was treated with intravenous immunoglobulin (2 g/kg over 5 days) and intensive supportive care including tracheostomy, physiotherapy, and autonomic monitoring. Over six weeks, she showed gradual motor recovery and was successfully weaned off the ventilator. At three-month follow-up, she had regained swallowing function and improved limb strength (2/5). Continued physiotherapy resulted in steady improvement in daily activities.

Conclusion: This case illustrates that even in profound paralysis associated with the AMAN variant of GBS, early immunotherapy and comprehensive multidisciplinary rehabilitation can yield remarkable recovery. Recognition of locked-in features and timely intervention are key determinants of prognosis.

Keywords: Guillain-Barré Syndrome, Acute Motor Axonal Neuropathy, Locked-in Syndrome, Intravenous Immunoglobulin, Rehabilitation

How to cite this article: Shinde PP, Patil RS. The Silent Survivor: A Case of Locked-in State due to AMAN Variant of Guillain-Barré Syndrome with Complete Neurological Recovery. Int J Drug Deliv Technol. 2026;16(23s): 430-432. DOI: 10.25258/ijddt.16.23s.45

Source of support: Nil., **Conflict of interest:** None

INTRODUCTION

Guillain-Barré Syndrome (GBS) is an acute, immune-mediated polyradiculoneuropathy characterized by rapidly progressive, symmetrical weakness, areflexia, and varying degrees of sensory and autonomic dysfunction (1). It represents one of the most common causes of acute flaccid paralysis worldwide, with an annual incidence of 1–2 cases per 100,000 population. Although GBS encompasses several subtypes, the acute motor axonal neuropathy (AMAN) variant is a distinct and severe form primarily affecting motor axons (2). AMAN is often triggered by antecedent infections, particularly Campylobacter jejuni, and is associated with antibodies directed against gangliosides such as GM1 and GD1a, leading to immune-mediated axonal injury (3).

Locked-in syndrome is a rare but devastating neurological state wherein the patient is fully conscious and cognitively intact but unable to perform any voluntary movements except vertical eye motion and blinking (4). It typically results from pontine lesions affecting corticospinal and corticobulbar tracts. However, in exceptional circumstances, severe peripheral motor paralysis as in AMAN variant GBS can mimic this presentation, resulting in a "GBS-related locked-in state"(5). Such cases pose significant diagnostic and therapeutic challenges, often requiring careful differentiation from brainstem stroke or other central causes through neuroimaging and electrophysiological studies (6).

The pathophysiology of AMAN involves complement-mediated axonal damage without significant demyelination. The resulting profound weakness, ventilator dependence, and minimal voluntary movement often create a clinical picture of complete paralysis (7). Despite its severity, timely administration of intravenous immunoglobulin (IVIG) or plasmapheresis, along with meticulous supportive and rehabilitative care, can yield substantial neurological recovery (8).

This report describes a rare presentation of locked-in state

in a 62-year-old woman diagnosed with the AMAN variant of GBS. The case underscores the importance of early recognition, aggressive immunotherapy, and multidisciplinary management in optimizing outcomes. It also highlights the remarkable potential for recovery even in cases initially appearing catastrophic. Through this report, we aim to increase clinical awareness of this uncommon presentation and emphasize that locked-in-like states in GBS, although alarming, are not necessarily irreversible with appropriate, timely intervention.

Case Presentation

Patient Information

A 62-year-old woman, previously healthy, presented with sudden onset weakness in both lower limbs that progressed rapidly over four days to involve the upper limbs. The weakness was symmetrical and ascending in nature. She reported difficulty walking and climbing stairs, followed by inability to lift her arms or grip objects. There was no history of fever, sensory loss, cranial nerve involvement, or exposure to toxins or medications. Ten days prior to symptom onset, she had an episode of diarrheal illness, which resolved spontaneously.

Clinical Findings

On examination, the patient was alert and fully conscious but unable to move any limb or articulate speech. Muscle tone was flaccid in all four limbs, and deep tendon reflexes were absent. Sensory examination was normal. Cranial nerve testing revealed absent facial movement and gag reflex, with preserved vertical eye motion findings consistent with a locked-in state. Respiratory examination showed shallow effort with desaturation, necessitating immediate ventilatory support.

Diagnostic Assessment

Routine hematological and biochemical investigations were within normal limits. Cerebrospinal fluid analysis demonstrated albuminocytologic dissociation (protein 120 mg/dL, normal cell count). Nerve conduction studies showed markedly reduced compound muscle action potentials (CMAPs) with preserved sensory nerve action potentials (SNAPs), confirming the acute motor axonal neuropathy (AMAN) variant of Guillain-Barré Syndrome. Electromyography revealed denervation consistent with motor axonal injury. MRI brain and cervical spine ruled out brainstem pathology, showing mild anterior nerve-root enhancement suggestive of inflammatory neuropathy.

Therapeutic Intervention

The patient received intravenous immunoglobulin (IVIG) at a dose of 2 g/kg over five days. Early tracheostomy was performed anticipating prolonged ventilation. Supportive management included lung-protective mechanical ventilation, enteral nutrition via nasogastric tube, deep vein thrombosis prophylaxis, and meticulous autonomic monitoring. Passive physiotherapy was initiated early to maintain joint mobility.

Follow-up and Outcomes

Gradual neurological recovery was observed from the fourth week onward, with minimal voluntary finger movements and improved facial muscle tone. She was successfully weaned from the ventilator after six weeks. Speech and swallowing rehabilitation began once bulbar function improved.

At discharge, limb power was 2/5, and she could maintain spontaneous respiration. At the three-month follow-up, she demonstrated marked improvement in swallowing, speech, and limb strength with ongoing physiotherapy and occupational therapy. Autonomic and pulmonary functions returned to near normal levels, indicating a steady and sustained recovery.

DISCUSSION

Guillain-Barré Syndrome (GBS) is a post-infectious, immune-mediated neuropathy that presents with rapidly progressive, symmetrical weakness and areflexia (9). The acute motor axonal neuropathy (AMAN) variant, first described in northern China, primarily affects motor axons without demyelination (7). It is strongly associated with Campylobacter jejuni infection and antiganglioside antibodies, particularly GM1 and GD1a (7). The resulting immune cross-reactivity leads to complement-mediated axonal degeneration, producing profound motor paralysis and ventilator dependence (10).

The occurrence of a locked-in-like state in GBS is extremely rare. This state, characterized by complete paralysis with preserved consciousness and vertical eye movements, is usually caused by pontine lesions due to vascular or traumatic insults (4). However, in severe AMAN or acute motor-sensory axonal neuropathy (AMSAN) variants, peripheral motor paralysis can mimic locked-in syndrome, often leading to diagnostic confusion with central nervous system disorders. In such cases, careful clinical evaluation and electrophysiological studies are crucial for differentiation. MRI brain and cervical spine imaging help exclude central causes (11).

In the present case, the rapid progression to quadriplegia and respiratory failure, along with preserved eye movements and normal sensation, raised suspicion for a locked-in state secondary to AMAN. The diagnosis was confirmed by nerve conduction studies showing markedly reduced CMAP amplitudes with preserved SNAPs. Cerebrospinal fluid analysis revealed albuminocytologic dissociation, consistent with GBS.

The cornerstone of management in GBS includes immunotherapy either intravenous immunoglobulin (IVIG) or plasma exchange administered early in the disease course. Supportive care, including mechanical ventilation, physiotherapy, nutritional support, and prevention of complications such as deep vein thrombosis and infections, significantly influences outcomes. In this case, timely administration of IVIG and multidisciplinary rehabilitation led to gradual but substantial recovery.

This case reports that even the most severe presentations of

AMAN variant GBS are potentially reversible. Early recognition of locked-in features, exclusion of central pathology, and prompt initiation of immunotherapy are essential for favorable outcomes. It also highlights the remarkable neuroplasticity and capacity for functional recovery when comprehensive rehabilitative care is sustained over time.

CONCLUSION

This case highlights that locked-in-like presentations can occur in severe AMAN variant Guillain-Barré Syndrome, mimicking central causes. Early diagnosis, prompt initiation of intravenous immunoglobulin therapy, and sustained multidisciplinary rehabilitation can lead to remarkable recovery. Awareness of this rare presentation facilitates timely intervention and improved neurological outcomes in affected patients.

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