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Rapidly Progressive Guillain-Barré Syndrome (AMSAN Subtype) With Remarkable Improvement on Plasmapheresis

Leyla Kedir Bereka[1], Tanymol Saju[2], Manish Kumar[3], Ammar Alomar[4*], Ghamdan Ali Alsadeh[5] and Aya Alsayyad[6]

[1,2]Department of medicine, Ibrahim Bin Hamad Obaidullah Hospital, Ras al Khaimah, United Arab Emirates

[3]Department of Anesthesia and Intensive care unit specialist, Ibrahim Bin Hamad Hospital, United Arab Emirates

[4]Department of neurology Consultant, Ibrahim Bin Hamad Obaidullah Hospital, Ras al Khaimah, United Arab

[5]Department of Anesthesia and critical care unit Consultant, Ibrahim Bin Hamad Obaidullah Hospital, Ras al Khaimah, United Arab Emirates

[6]Department of neurology specialist, Ibrahim Bin Hamad Obaidullah Hospital, Ras al Khaimah, United Arab

[*]Corresponding author:

Ammar Alomar,
Department of neurology Consultant, Ibrahim Bin Hamad Obaidullah Hospital, Ras al Khaimah, United Arab

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1. Abstract

Guillain-Barré syndrome (GBS) is a rare disorder that affects the peripheral nervous system. It typically presents with ascending limb weakness, areflexia, and, in severe cases, respiratory failure. Standard treatments for GBS include intravenous immunoglobulin (IVIG) and plasmapheresis, both of which have been shown to be equally effective in the literatures; however, responses can vary between patients. We report the case of a 42-year-old woman who experienced progressive limb weakness that led to respiratory failure, despite receiving IVIG therapy. Neurophysiological studies identified an acute motor sensory variant of GBS, known as acute motor sensory axonal neuropathy (AMSAN) which is characterised by more rapid progression with axonal degeneration and delayed recovery. Remarkably, the patient demonstrated significant improvement after undergoing plasmapheresis, highlighting the importance of personalised treatment for managing GBS.

2. Introduction

Guillain-Barré syndrome (GBS) is an acute immune-mediated neuropathy that leads to progressive muscle weakness and sensory disturbances. It is a rare disorder affecting approximately 1-2 individuals per 100,000 people annually, and often following a recent infection, particularly of the respiratory or gastrointestinal tract [1].

The clinical presentation usually features symmetrical limb weakness and areflexia; however, the syndrome can manifest in various ways, resulting in a wide range of initial symptoms. In severe cases, patients may experience respiratory failure. Approximately, one-third of the patients with GBS will require mechanical ventilation, and most GBS-related deaths occur due to respiratory failure [8,9]. GBS is classified into several subtypes, with Acute Inflammatory Demyelinating Polyneuropathy (AIDP) being the most common. This subtype is characterised by the demyelination of peripheral nerves and presents with ascending motor weakness and areflexia. Facial palsy is the msot common type of cranial nerve involvement (53%), followed by bulbar weakness, ophthalmoplegia, and tongue weakness. Other variants such as Acute Motor Axonal Neuropathy (AMAN) and Acute Motor-Sensory Axonal Neuropathy (AMSAN), are associated with axonal damage and can lead to more severe clinical outcomes. Miller Fisher syndrome, which is characterised by ophthalmoplegia and ataxia, is another less common variant [1,6]. This case report highlights the crucial role of plasmapheresis in managing severe GBS, par-

ticularly when IVIG alone is insufficient. By detailing the patient's clinical course and response to plasmapheresis, we seek to contribute to understanding the effective treatment strategies for severe GBS and highlighting the importance of timely and appropriate therapeutic interventions.

3. Case Presentation

A 42-year-old female with no significant medical history presented with acute bilateral weakness of the limbs associated with sensory disturbances that began two days later. The symptoms started in the lower limbs and progressed to involve both upper limbs. The patient was unable to walk on the morning prior to admission. She had a recent history of an upper respiratory tract infection (URTI) one week before the onset of symptoms. Upon admission, on physical examination, the patient was conscious and oriented, hemodynamically stable, with oxygen saturation maintained on room air; however, she had mild dyspnea. Neurological examination revealed proximal muscle weakness (3/5) and distal muscle weakness (4/5) in all limbs. The patient had hyporeflexia with no signs of bulbar involvement or cranial nerve deficits. Brain CT scan showed normal findings. Laboratory tests, including complete blood count, inflammatory markers, renal function tests, electrolytes, liver function tests, urine examination, and chest radiography were normal, ruling out common metabolic or infectious causes. Cerebrospinal fluid (CSF) analysis revelaed elevated protein levels (70.5 mg/dl), normal glucose levels (3.8 mmol/L), and normal cell count (3 mononuclear cells /mm3). These results clearly showed albuminocytological dissociation, indicating inflammatory polyneuropathy. The patient initially received IVIG, the standard first-line therapy for GBS, at a dosage of 0.4 g/kg per day over five days. However, on the third day of admission, she developed breathing difficulties due to progressive respiratory muscle weakness, hypoxaemia, and hypercapnia, necessitating intubation and mechanical ventilation. 0n the seventh day, nerve conduction studies were performed to confirm the diagnosis of GBS. The study assessed the median, ulnar, and peroneal nerves, revealing a diffuse axonal sensory-motor neuropathic process. There was severe impairment of the sensory fibres, which were completely lost, along with significant evidence of major axonal loss, as indicated by the marked reduction in compound muscle action potential amplitudes that exceeded moderate degree. This clinical presentation is consistent with the AMSAN phenotype associated with GBS, classified as moderately severe. The patient completed a course of IVIG; however, her condition did not improve. A trial to extubate her had failed. Given the inadequate response to IVIG, we performed five sessions of plasmapheresis starting on the 14th day after admission. Each session involved the exchange of 2–3 litres of plasma based on the patient's body weight (50 ml/kg). To optimise the plasmapheresis procedure, we utilised continuous flow rather than intermittent methods; using albumin as the exchange fluid. This approach proved effective and well- tolerated, significantly contributing to the patient's recovery. Plasmapheresis led to a significant improvement in the patient's condition, particularly in reducing respiratory muscle weakness and stabilising the respiratory status. Follow-up neurological examinations revealed substantial improvements, proximal muscle strength increased to 4/5, distal muscle strength remained at 5/5 in all limbs, and sensory disturbances and numbness were notably reduced. Additionally, her respiratory function markedly improved, allowing successful extubation within 3 weeks. Subsequently, the patient was transferred to the medical ward for ongoing management and rehabilitation of the motor deficits. A comprehensive discharge plan was established, which included scheduled follow-up appointments to monitor her recovery and rehabilitation progress. The patient visited the clinic one month after discharge, reporting that she returned to her normal daily activities. She complained of numbness only in the right foot, but no respiratory, speech or swallowing difficulties. Muscle strength was 5/5 in all limbs, except for mild weakness in the right foot with normal sensation.

Table 1: Results of CSF Analysis**.**

CSF Analysis	**Reference value**	Patient value
Pressure	10 - 25 cm	15cm
Appearance	Colourless	Colourless
Protein (mg/L)	150- 450	705.0
Glucose (mmol/L)	2.2- 3.9	3.8
CSF poly segmented Neutrophil	0	0
Glucose –CSF serum ration	115/3.8	40- 80
WBC	0-5	3
CSF RBC count	Nil	2000/uL
CSF culture and gram stain	No bacteria were observed	

Table 2: The table highlights the major differences between Intravenous immunoglobulin therapy and plasmapheresis.

Features	IVIG	Plasmapheresis
Molecular Target	Pathogenic autoantibodies and components of the immune system	Pathogenic autoantibodies, immune complexes, complement factors, cytokines, and other pro-inflammatory mediators
Mechanism(s) of Action	-Modulates pathogenic autoantibody production - Neutralizes autoantibodies - Inhibits complement activation - Modulates immune responses (e.g., cytokines and T-cell functions)	- Rapidly removes harmful substances from plasma, including pathogenic autoantibodies and inflammatory mediators -Provides immediate therapeutic effects
Relative Strengths	-Widespread availability - No specialized equipment required - Safe in pregnancy; side effects are uncommon	- Rapid therapeutic effect. - Effective in quickly reducing harmful circulating agents
Relative Weaknesses	- Variable efficacy (dependent on brand/batch) - Risk of thrombotic events (e.g., myocardial infarction, DVT), renal failure, and anaphylaxis in IgA deficiency - Meningismus may occur	- Requires specialized equipment; high costs - Risk of hemodynamic instability, dilutional coagulopathy, hypocalcemia, and thrombosis

DVT: Deep Venous Thrombosis, IVIG: intravenous immunoglobulin

4. Discussion

This case underscores the remarkable improvement observed in a 42-year-old woman with severe GBS following plasmapheresis. Initially, the patient received a full dose of IVIG without improvement; however her condition deteriorated due to respiratory muscle weakness. Plasmapheresis, which removes pathogenic antibodies from the blood, led to significant improvements in her condition. After five sessions, the patient showed substantial gains in muscle strength and reflexes, and was successfully weaned off mechanical ventilation. Nerve conduction studies showed features consistent AMSAN subtype, which is a severe form of GBS. In this form, the disease progresses more rapidly, with both sensory and motor fibres affected by marked axonal degeneration, frequently causing delayed and incomplete recovery. Although AMSAN is associated with antiganglioside antibodies, we we did not detect them in this case. The AMSAN subtype has also been associated with other infections, such as Haemophilus influenzae, Mycoplasma pneumoniae, and cytomegalovirus. Cytomegalovirus infections (CMV). Notably, CMV infections are associated with autoantibody production [3,4]. Plasmapheresis and IVIG are immunomodulatory therapies commonly used in the management of GBS. Both IVIG and plasmapheresis reduced the levels of circulating autoantibodies and immune mediators via different mechanisms and to varying degrees. Regarding IVIG, it contains purified human immunoglobulins, obtained from large pools of plasma donated by thousands of healthy donors. IVIG is thought to exert its therapeutic effects by inactivating immune mediators (e.g. immune complexes and antibodies targeting nervous system components) and modulating immune responses (e.g. impeded activation of the complement system, altered B- and T-cell activation, and decreased inflammatory cell adhesion and migration) [5]. In contrast to IVIG, plasmapheresis aims to eliminate immune mediators. Plasmapheresis is a highly effective rescue treatment for patients with acute exacerbation of neuroimmunological disease, targeting and removing circulating autoantibodies and inflammatory components that attack peripheral nerves from the bloodstream. During plasmapheresis, plasma is separated from the blood cells and discarded, effectively 'washing out' harmful components and thereby reducing their levels in the circulation. However, this 'washout effect' is typically transient. Patients may require multiple sessions to achieve sustained benefits, as the body continues to produce harmful antibodies and mediators [2,7]. The case highlights an important consideration in the treatment of GBS: the potential 'washout effect' of IVIG when followed by plasmapheresis. Plasmapheresis removes plasma containing residual IVIG antibodies, which may alleviate immune-mediated damage to the peripheral nerves. This 'washout effect' also enhances recovery by decreasing the concentration of pathogenic antibodies that interfere with nerve function. The positive response observed in this case highlights the importance of plasmapheresis as an adjunctive therapy, particularly when IVIG alone is inadequate [5]. The patient initially received IVIG, the standard first-line therapy for GBS, at a dosage of 0.4 g/kg per day

over five days. However, on the third day of admission, she developed breathing difficulties due to progressive respiratory muscle weakness, hypoxaemia, and hypercapnia, necessitating intubation and mechanical ventilation. on the seventh day, nerve conduction studies were performed to confirm the diagnosis of GBS. The study assessed the median, ulnar, and peroneal nerves, revealing a diffuse axonal sensory-motor neuropathic process. There was severe impairment of the sensory fibres, which were completely lost, along with significant evidence of major axonal loss, as indicated by the marked reduction in compound muscle action potential amplitudes that exceeded moderate degree. This clinical presentation is consistent with the AMSAN phenotype associated with GBS, classified as moderately severe. The patient completed a course of IVIG; however, her condition did not improve. A trial to extubate her had failed. Given the inadequate response to IVIG, we performed five sessions of plasmapheresis starting on the 14th day after admission. Each session involved the exchange of 2–3 litres of plasma based on the patient's body weight (50 ml/kg). To optimise the plasmapheresis procedure, we utilised continuous flow rather than intermittent methods; using albumin as the exchange fluid. This approach proved effective and well- tolerated, significantly contributing to the patient's recovery. Plasmapheresis led to a significant improvement in the patient's condition, particularly in reducing respiratory muscle weakness and stabilising the respiratory status. Follow-up neurological examinations revealed substantial improvements, proximal muscle strength increased to 4/5, distal muscle strength remained at 5/5 in all limbs, and sensory disturbances and numbness were notably reduced. Additionally, her respiratory function markedly improved, allowing successful extubation within 3 weeks. Subsequently, the patient was transferred to the medical ward for ongoing management and rehabilitation of the motor deficits. A comprehensive discharge plan was established, which included scheduled follow-up appointments to monitor her recovery and rehabilitation progress. The patient visited the clinic one month after discharge, reporting that she returned to her normal daily activities. She complained of numbness only in the right foot, but no respiratory, speech or swallowing difficulties. Muscle strength was 5/5 in all limbs, except for mild weakness in the right foot with normal sensation.

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7. Conclusion

This case report underscores the critical role of plasmapheresis in the management of severe GBS, particularly when IVIG treatment alone is inadequate. The successful application of plasmapheresis in the treatment plan of this patient highlights its significance as a key therapeutic option for patients experiencing severe symptoms. Although this report provides valuable insights into the efficacy of plasmapheresis, its findings are limited to a single patient. Further research is essential to optimize plasmapheresis protocols and evaluate long-term outcomes. Overall, this case emphasizes the necessity of timely and comprehensive treatment strategies for GBS, contributing to an evolving understanding of effective management approaches.

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