A Case Study Perspective: Navigating Guillain-Barre Syndrome

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Abstract

Background. Guillain-Barre syndrome (GBS) is one of the primary causes of acute paralysis, which can potentially distress any age group of the human population. It is a rare but serious post-infectious, immune-mediated neuropathy, resulting from the autoimmune destruction of nerves in the peripheral nervous system. Several infections have been linked with GBS, the most common being gastrointestinal and respiratory illnesses. Up to 70% of patients have reported an antecedent illness in the 1 to 6 weeks before the presentation of GBS. Despite the current understanding of the pathophysiology and available treatment options, the morbidity and mortality associated with GBS is quite high. Patient concerns and findings. Here, we present a case from

a tertiary care hospital, where a 16-year-old boy presented with presented with sudden onset bilateral lower limb weakness, immobility, dysphagia, fever, cough and bowel incontinence. Main diagnosis, interventions and outcomes. The CNS examination suggested diminished reflexes, tone, complete absence of power in all the limbs and facial nerve palsy. Pleural effusion and spinal cord edema were other investigative observations. These led to the diagnosis of Guillain-Barre syndrome, with lower respiratory tract infection. The patient was immediately transferred to the intensive care unit, wherein the necessary antimicrobials were initiated. This was followed by five cycles of plasmapheresis with a successful outcome. Conclusion. Although the patient response was satisfactory, vigorous monitoring for comorbidities and secondary infections should be necessitated.

Keywords: Neurodegenerative diseases; Guillain-Barre Syndrome; Autoimmune conditions; Respiratory infections; Plasmapheresis

Introduction

Guillain-Barré Syndrome (GBS), a rare autoimmune condition causes damage to the peripheral nervous system. The immune system accidentally targets the nerves, causing inflammation and damage, which is the hallmark of GBS [1]. The immune reaction to a preceding infection, most frequently by bacterial or viral agents - plays a role in the pathophysiology of GBS [2, 3]. Molecular mimicry, i.e. the similarity between infectious agent antigens and peripheral nerve proteins is assumed to be a major factor. A cross-reactive immune response that targets the infection as well as the nerves may result from these similarities. The myelin sheath, protecting the nerve fibres is the primary target of the immunological onslaught. A variety of neurological symptoms are brought on by this demyelination, which interferes with normal transmission of nerve impulses [4, 5]. Mild sensory or motor symptoms, such as tingling, weakness, or numbness in the extremities, are usually the initial signs of GBS. These symptoms worsen and may increase symmetrically as the condition advances [6]. In severe situations, muscle weakness can progress quickly and result in paralysis. Ascending paralysis, which frequently begins in the legs and spreads to the upper body and, in certain situations,

affects breathing muscles. GBS is a potentially fatal illness since respiratory collapse may require mechanical support [7, 8].

Clinical Findings and Diagnostic Assessment

A 16-year-old boy from a semi-urban area presented at our hospital with a sudden onset bilateral, lower limb weakness in the last two days which was gradually progressive. Initially, he was able to walk with support, but by end of the day, he could not even move the limbs. This was followed by sole movement of fingers associated with a difficulty in swallowing for two more days. Eventually, he could not even swallow saliva. His mother also reported a history of fever and cough in the last four days, with an increased stool frequency, marked by bowel incontinence.

At the time of arrival, the patient was conscious and oriented to time, place and person. His vital signs showed a normal temperature, a pulse rate of 70 bpm, blood pressure of 120/80 mm Hg, and oxygen saturation of 98% on room air. Upon systemic examination, the respiratory system exhibited bilateral air entry with clear noises, the gastrointestinal system demonstrated a soft and non-tender belly, and the existence of bilateral 7th cranial nerve palsy, suggestive of facial paralysis. The patient also presented with bilaterally absent plantar reflexes, total paralysis (0/5 power), and reduced tone in all four limbs. Ankle, brachioradialis, triceps, patellar, and biceps reflexes were among the superficial reflexes that were significantly reduced or absent. A bilateral pupil test revealed equal-sized pupils that responded slowly to light. A right-sided pleural effusion was suggested by a soft tissue opacity in the right lower lung zone and blunting of the right costophrenic angle shown on a chest X-ray. The results of the examination and diagnosis add to a thorough clinical profile of the patient.

Furthermore, aerobic culture from the tracheal aspirate identified Gram-negative bacilli, of which Acinetobacter baumannii and Klebsiella pneumoniae were isolated and showed heavy and limited growth, respectively. MRI of the cervical spine showed long-segment mild intra-medullary T2W and STIR hyperintensity, which indicated cord oedema. The hyperintensity extended from the cervico-medullary junction to the D1 vertebral level. Additionally, a long-segment mild hyperintensity was seen in the myelography. A right ventricular systolic pressure (RVSP) of 24 mm Hg and a left ventricular ejection fraction (LVEF) of 60% were recorded by the 2D ECHO. The electrocardiogram (ECG) revealed sinus tachycardia.

Anisopoikilocytes, tear drop cells, elliptical cells, neutrophilic leukocytosis, and normochromic normocytic anaemia were all seen in peripheral smears.

Leukocytosis, which is indicated by an elevated white blood cell (WBC) count, was mostly linked to an infectious cause. Thrombocytosis, i.e., a high platelet count, suggested a viral aetiology and an underlying autoimmune condition. Moreover, uraemia was detected, indicating acute renal impairment. Liver injury was indicated by elevated levels of transaminitis-related serum glutamic pyruvic transaminase (SGPT) and serum glutamic oxaloacetic transaminase (SGOT). Furthermore, red blood cells and pus cells in the urine (pyuria) were observed, indicating a urinary tract infection (UTI).

These diagnostic findings, laboratory parameters and examination, along with the clinician's judgement led to the diagnosis of Guillain-Barré Syndrome, along with lower respiratory tract infection (LRTI).

Therapeutic Interventions

During the five days of stay in the intensive care unit (ICU), the patient was treated with broad spectrum antibiotics and an antifungal - Inj. Colistin 3 mIU (TDS), Inj. Sultamicillin 1.5 g (BD) and Fluconazole 150 mg (OD) via Ryles tube. Five cycles of plasmapheresis were also performed. After being shifted to the general ward, the treatment plan consisted the following: Inj. Cefoperazone/Sulbactam 1.5 g (BD), Inj. Doxycycline 200 mg (BD) for 5/14 days, calcium plus vitamin D3 supplements, Inj. Pantoprazole 40 mg (BD), Inj. Enoxaparin 0.4 mg (OD), Acetylcysteine 600 mg (TDS), Liq. Paraffin 30 mL (BD), Neb. Duolin (TDS), Carboxymethylcellulose eye drops (TDS), Disodium hydrogen citrate syrup (TDS). Hence, the standard treatment protocol was followed for this patient.

After 14 days of treatment, the patient was stable and his symptoms had improved. Following this, the patient was discharged and was advised to follow-up with the physician after a week, to further discuss about IVIG treatment.

The table below, **Table 1** shows the treatment chart of the patient.

Table 1: Treatment regimen administered to the patient.

Drug	Dose	RA	Freq.	01	02	03	04	05	06	07	08	09	10	11	12	13	14
Cefoperazone/Sulbactam	1.5 g	IV	BD	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓
Doxycycline	200 mg	IV	Stat, BD	×	×	×	✓	✓	✓	✓	✓	×	×	×	×	×	×
Calcium + Vitamin D3	500 mg	RT	TDS	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓
Pantoprazole	40 mg	IV	BD	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓
Ondansteron	4 mg	IV	SOS	×	×	×	×	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓
NS/DNS/RL	500 mL	IV	Inf.	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓
Enoxaparin	0.4 mg	SC	OD	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓

Acetylcysteine	600 mg	RT	TDS	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓
Liq. Paraffin	30 mL	RT	BD	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓
Neb. Duolin	-	Nasal	TDS	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓
Carboxymethylcellulose	-	Eye drops	TDS	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓
Disodium hydrogen citrate syrup	2 tsf	RT	TDS	×	×	×	×	×	×	✓	✓	✓	✓	✓	✓	✓	✓

Discussion

A prompt differential diagnosis and treatment initiation are crucial to the management of GBS and related comorbidities. For the presented case, the team acted on the diagnosis and kept the patient under observation in the ICU. While being discharged, the patient's family was also referred to a neuro-physiotherapist to overcome possible long-term damage. Appropriate counselling to the patient was also provided about his medications and being alert to any deteriorating signs. A long-time lapse required for a definite diagnosis would add the potential risk of airway compression and distress, particularly in a such a patient presenting with pulmonary symptoms. The standard therapy for GBS usually comprises: (i) Prehospital and emergency department care, including intubation for hypoxia, rapidly declining respiratory function, poor or weak cough, suspected aspiration, (ii) Respiratory therapy, including tracheotomy for prolonged respiratory failure, especially if mechanical ventilation is required for more than 2 weeks, (iii) Prevention and treatment of infections which consists minimal sedation, frequent physiotherapy, antibiotic therapy, (iv) Immunomodulatory treatment, which is the mainstay intervention of this condition. It includes intravenous immunoglobulins (IVIG) and plasma exchange or plasmapheresis [9].

However, a serious drug-drug interaction had to be kept under surveillance, which was an increased bleeding risk associated with the simultaneous administration of Cefoperazone and Enoxaparin. A medication error in the treatment which had to be taken care of was the administration of a laxative (Liq. Paraffin) even though the patient presented with diarrhea and bowel incontinence. A few other points to be addressed were prescribing an oral alternative (such as rivaroxaban) to subcutaneous enoxaparin, monitoring for secondary infections, such as urinary tract infections and vigorous monitoring for liver function abnormalities, during admission and after discharge, due to continued enoxaparin prescription.

As it is typical to this syndrome, the patient presented with the classic signs and symptoms which further simplified the diagnostic decision-making. Moreover, almost 30% GBS patients develop respiratory failure and require ICU admission [10]. Mechanical ventilation is also necessitated in several cases and such poor conditions require sincere attention from the caregivers [8]. Ventilator-associated pneumonia and

nosocomial infections are another area of concern in an already debilitated patient and hence, early tracheostomy and proper prophylaxis with antibiotic and antifungal agents is highly recommended to avoid any complications [11]. According to a research study, systemic infections occurred in one-fifth of GBS patients admitted to the ICU. The risk was increased proportionally to the duration of ICU stay. Uncommon life-threatening conditions like pulmonary embolism and gastrointestinal bleeding were also common [12]. Giving due attention to the risk of deep vein thrombosis, our patient was started on anticoagulant treatment, which was also continue after discharge.

Conclusion

Though not very common, Guillain-Barré Syndrome (GBS) required urgent medical attention, in addition to the patient/family being aware of the seriousness of this condition. Early hospitalisation, a quick differential diagnosis, ICU admission, thorough antibiotic treatment and five-cycle of plasmapheresis in our patient helped him recover. A 14-day hospital stay also played a role in keeping the patient under core observation of the healthcare professionals and help reduce any likely future adverse event related to the condition. Early treatment is the key to managing GBS, especially for patients presenting with concomitant respiratory conditions.

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Ethical Clearance

Not Applicable

References

1. Dimachkie MM, Barohn RJ. Guillain-Barré syndrome and variants. Neurologic clinics. 2013;31(2):491-510.
2. Jasti AK, Selmi C, Sarmiento-Monroy JC, Vega DA, Anaya JM, Gershwin ME. Guillain-Barré syndrome: causes, immunopathogenic mechanisms and treatment. Expert review of clinical immunology. 2016;12(11):1175-89.
3. Finsterer J. Triggers of Guillain-Barré Syndrome: Campylobacter jejuni Predominates. International journal of molecular sciences. 2022;23(22).
4. Malek E, Salameh J. Guillain-Barre Syndrome. Seminars in neurology. 2019;39(5):589-95.
5. Rodríguez Y, Rojas M, Pacheco Y, Acosta-Ampudia Y, Ramírez-Santana C, Monsalve DM, et al. Guillain-Barré syndrome, transverse

myelitis and infectious diseases. Cellular & molecular immunology. 2018;15(6):547-62.

6. Uncini A, Yuki N. Sensory Guillain-Barré syndrome and related disorders: an attempt at systematization. Muscle & nerve. 2012;45(4):464-70.

7. Shahrizaila N, Lehmann HC, Kuwabara S. Guillain-Barré syndrome. Lancet (London, England). 2021;397(10280):1214-28.

8. Shang P, Zhu M, Baker M, Feng J, Zhou C, Zhang HL. Mechanical ventilation in Guillain-Barré syndrome. Expert review of clinical immunology. 2020;16(11):1053-64.

9. Chevret S, Hughes RA, Annane D. Plasma exchange for Guillain-Barré syndrome. The Cochrane database of systematic reviews. 2017;2(2):Cd001798.

10. Madden J, Spadaro A, Koyfman A, Long B. High risk and low prevalence diseases: Guillain-Barré syndrome. The American journal of emergency medicine. 2024;75:90-7.

11. Dharmayanti A, Astrawinata D. Ventilator-Associated Pneumonia (VAP) in a Patient with Guillain-Barre Syndrome. Acta medica Indonesiana. 2017;49(2):151-7.

12. Henderson RD, Lawn ND, Fletcher DD, McClelland RL, Wijdicks EFM. The morbidity of Guillain-Barré syndrome admitted to the intensive care unit. 2003;60(1):17-21.