AUCTORES
Globalize your Research

Journal of Surgical Case Reports and Images

Kiran Dudhat *

Open Access

Case Report

A Rare Case Report Presentation on Guillain-Barré Syndrome in Young Male

Pinal Viradiya, Tisha Lathia, Kushal Parekh, Kiran Dudhat*

School of Pharmacy, RK University, Kasturbadham, Rajkot, Gujarat- 360020, India.

***Corresponding Author:** Kiran Dudhat, School of Pharmacy, RK University, Kasturbadham, Rajkot, Gujarat- 360020, India.

Received Date: September 17, 2024; **Accepted Date:** September 30, 2024; **Published Date:** October 09, 2024

Citation: Pinal Viradiya, Tisha Lathia, Kushal Parekh, Kiran Dudhat, (2024), A Rare Case Report Presentation on Guillain-Barré Syndrome in Young Male, *J, Surgical Case Reports and Images*, 7(9); **DOI:10.31579/2690-1897/218**

Copyright: © 2024, Kiran Dudhat. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Abstract

Guillain-Barré Syndrome (GBS) is an immune-mediated illness, can result in motor disability and even death. Muscle weakness is one of its symptoms, which calls for an immediate diagnosis and course of treatment. Treatment options include plasma exchange and IV immunoglobulin therapy, both of which work best when started early. Increasing knowledge about GBS is essential for better patient outcomes and early diagnosis. To ensure that those who are impacted receive the best care possible, further research is required to improve therapeutic approaches and deepen understanding. In this case report, we have discussed the case of a 16-year-old male patient who had Guillain-Barré Syndrome and was given IV immunoglobulin therapy.

Keywords: gbs; immunoglobulin therapy; emg/ncs; inflammatory demyelinating polyneuropathy

Introduction:

GBS is thought to be an immune-mediated illness that targets the peripheral nerve system, which are the nerves that are not part of the brain or spinal cord. It can take days or weeks for symptoms to appear. Varied geographic regions experience the condition in varied proportions for its demyelinating and axonal manifestations, and there are easily distinguishable clinical variations such as Miller Fisher syndrome. GBS can result in severe muscular weakness, and 5% of patients pass away. Acute motor axonal neuropathy (AMAN) and acute inflammatory demyelinating polyradiculoneuropathy (AIDP) are the two most prevalent subtypes. The first presenting symptom in the majority of individuals with typical Guillain-Barré syndrome is rapidly progressing bilateral weakness. weakness is traditionally defined upward, typically beginning in the distal lower extremities, however it may begin closer to the body, in the legs or arms. The later pattern may provide a misleading clinical a pyramidal lesion's impression, or at the level of the spinal cord or higher), however this is readily accounted for by focused conduction block at the cervical and lumbar levels nerve roots as opposed to the entire length of the nerve fibre. [1]. GBS is a global disease that affects men more often than women, with a median yearly incidence of 1.3 cases per 100,000 people. It is believed that GBS is an autoimmune illness brought on by a previous viral or bacterial infection. Commonly recognized antecedent pathogens include Campylobacter jejuni, cytomegalovirus, Epstein-Barr virus, and Mycoplasma pneumoniae. It is obvious that a fundamental factor in the development of the disorder—at least in the case of Campylobacter jejuni infection—is the molecular mimicry between microbial and neuronal antigens. It is still unclear, though, how and when the immune response is changed toward undesirable autoreactivity due to the interaction of microbial and host components. Moreover, it is unknown whether hereditary and environmental variables influence a person's propensity to get the illness. Most people (>99%) who are exposed to an immunological stimulation as a result of infections linked to Guillain-Barré syndrome, including C jejuni, do not develop unwanted autoimmunity. [1,2]. Different cultures have developed GBS at different genetic loci, especially those pertaining to immune response control. Research on GBS has revealed aberrant expression of cytokine-coding genes, and investigations utilizing an animal model of autoimmune neuritis have confirmed that Th1/Th2/Th17/Treg cytokines are regulated. Although HLA alleles have been linked to GBS risk, opinions among populations about their significance are divided. Certain GBS subtypes have been linked to SNPs in cytokine-coding genes, namely IL-17 and TNF-α, suggesting that these subtypes may have distinct processes. A substantial percentage of the gene abnormalities in GBS cases could be explained by epigenetic processes. Studies conducted in vitro have revealed that GBS patients' peripheral blood cells and immune cells respond differently. [11]. Patients with GBS have complement, immunoglobulin, and macrophage infiltration peripheral nerve deposits.

GBS patients' blood also contains activated T cells, inflammatory cytokines, and anti-ganglioside antibodies. Neuronal membranes contain large amounts of gangliosides, sialylated glycosphingolipids that may be involved in signal recognition mechanisms. Numerous autoimmune disorders have been linked to the idea of molecular mimicry, with the function of molecular mimicry being established at several stages. Two key conditions must be met for a cross-reactive immune response to be triggered against autologous antigens: the antigens must be sufficiently dissimilar from the microbial to cause the immune system to break tolerance to self, and the antigens must be similar for immune responses to cross-react. In certain cases, lipopolysaccharide (LPS), which is present in the cell walls of C jejuni strains, has been shown to trigger anti-ganglioside antibodies. Through complement-dependent activity, anti-ganglioside antibodies mediate the illness course and contribute to the pathogenesis of GBS. IgG and complement deposits are observed on the axolemma at the nodes of Ranvier motor fibers in autopsy studies of AMAN, accompanied with little demyelination and lymphatic infiltration. In AMAN and Miller Fischer syndrome, C jejuni attached to motor nerve terminals destroy neuromuscular junctions in a complement-dependent manner, resulting in damage to neuronal and perisynaptic Schwann cells. Therapeutic applications of complement inhibitors are being investigated. [3,12]. Early diagnosis of Generalized Brain Syndrome (GBS) is important since it helps identify subgroups, shortens the disease's length and severity, and decreases the number of patients who need mechanical breathing. Different neurophysiological diagnostic criteria have been presented, and the diagnosis is made using Nerve Conduction Studies (NCS) and electromyography (EMG). By detecting anomalies in motor nerves to describe demyelination and categorizing axonal GBS into AMAN and AMSAN, NCS aids in differential diagnosis. Studying at least three sensory and four motor nerves, F-waves, and H reflexes improves NCS's diagnostic capacity. [3]. More than two-thirds of patients had increased concentrations of CSF protein, which is non-specific. Since axonal GBS is pathophysiologically characterized by axonal degeneration and reversible conduction failure at the axolemma of the node of Ranvier, Uncini et al. identify a trap when diagnosing GBS with regard to electrodiagnosis. Reversible conduction failure patterns should be taken into account as reliable electrodiagnostic standards change. [13]. As supplemental testing for GBS, CSF and serum analysis are also performed. CSF protein rises in nearly three-fourths of patients, with mononuclear cell count staying normal in the initial days of illness setting. [14]. PE and IVIg are examples of traditional therapy for GBS patients. IVIg, obtained from healthy donor blood, neutralizes pathogenic antibodies and inhibits complement system activation. It is recommended to take it within two weeks of becoming weak, at a dose of 0.4 g kg−1 day−1 for five days. IVIg or PE alone is more effective than their combination, according to a significant clinical investigation. Due to its effectiveness, safety, and accessibility, IVIg is used extensively across the world; but, in the future, its dosage and cost may need to be adjusted. [4,15]. There are ten main kinds of inhibitory glycoproteins known as interferons (IFNs). IFN-β, which is present in humans in natural forms as IFN-β, IFN-β-1b, and IFN-β-1a, is thought to be involved in GBS because of its possible advantages as a cellular immunomodulator. It could lower the incidence of GBS by blocking antigen presentation, T cell proliferation, adhesions, producing more anti-inflammatory cytokines, and altering macrophage characteristics. [16]. Patients with severe autoimmune illnesses can benefit from high-dose cyclophosphamide since it has a strong immunosuppressive effect and can improve their quality of life. Prophylactic CY treatment, however, can improve the clinical and histological aspects of GBS, as demonstrated by three case reports of patients with simultaneous GBS and systemic lupus erythematosus. However, extended treatment is linked to infections and neoplastic disorders.[17].

Case Report

A 16-year-old man from Rajkot, Gujarat presented with bilateral lower limbs weakness for 4 days and upper limbs weakness, diarrhoea and vomiting for 2 days. He was on a mixed diet and had no significant family or personal history. He has no social history. On examination, he was found to be conscious and well-oriented. Initial vitals, Biochemical parameters and Abdominal USG Report was normal. C-Reactive Protein (CRP) was found to be normal. CSF Sugar was found to be 94 mg/dl which was higher (40-80 mg/dl), while Protein-CSF was 29 mg/dl which was normal (8-43 mg/dl). There was abnormal RBC level 5.59* 10^6/ cumm was noted. Power and tone of muscle of upper and lower limbs were normal. Level of MCV and MCH were lower than normal value, while RDW was higher than normal. On next day MRI brain and whole spine screening, EMG/NCS Report (upper and lower limbs) was recommended along with vitamin B12 serum. In MRI brain and Whole spine screening there was no evidence of acute infarct, haemorrhage or space occupying lesion, while sacralisation of L5 vertebra is noted and no significant disc herniation of nerve root compression is seen also visualised spine cord appears normal in signal intensity. vitamin B12 (Cyanocobalamin) serum level >2000 pg/ml which was higher than normal levels (197-771 pg/ml). EMG/NCS Report of upper and lower limbs showed, BIL median and ulnar nerve was normal DL with normal CMAPS and normal CV. BIL radial nerve shows normal DL with normal CMAPS, left peroneal nerve shows normal DL with normal CMAPS and normal CV while right peroneal nerve shows normal DL with normal CMAPS and decreased CV. One other hand left tibial nerve shows increased DL with normal CMAPS and decreased CV while right tibial nerve shows increased DL with normal CMAPS and normal CV. Sensory nerve conduction shows right median sensory nerve increased DL with normal snaps and decreased CV while left median and ulnar sensory nerve shows normal DL with normal snaps and normal CV. From this NCS findings of both upper limbs are within normal limits while for lower limbs are so distal, symmetrical, predominantly demyelinating variant of pure motor poly neuropathy was found. Treatment plan for curing the patient include; inj. IvIg (20mg) IV(OD), Inj.Pantoprazole (40mg) iv (12hrly), Inj.Ondansetron (8mg) iv (8hrly), tab. Calcium (500mg) (OD), Tab. Folic acid (5mg) (OD).

Discussion

Initially identified as an acute inflammatory demyelinating polyneuropathy, Guillain-Barré syndrome (GBS) is an acute self-limited polyneuropathy which was first described in 1916. It has developed into other subgroups, such as Miller Fisher syndrome and acute motor axonal neuropathy. Since the 1980s, acute-phase sera have included antibodies against glycolipids, indicating that GBS is an immune-mediated illness.[5] Globally, GBS is a common disease with a median incidence of 1.3 cases per 100,000 people (range: 0.4–4.0). The condition affects men more frequently than women, with peaks in young people and the elderly. In Western countries, there is no obvious seasonal correlation; this could be because the two common antecedent events—respiratory and enteric infections—have opposing seasonality. Campylobacter jejuni infection is the most commonly found cause of GBS, accounting for up to 41% of

cases and linked to more severe illness and extended impairment.[6] It is frequently linked to parturition, vaccination, and surgery. Patients progress quickly; 50% reach their clinical nadir in two weeks, and 90% do so in four. Between 80 and 90 percent of patients become immobile during the illness, and 50 percent of patients report significant pain. While sensory evaluation is normal in the early period, neurological exams frequently reveal proximal and distal weakness. The predominant condition is widespread areflexia or hyporeflexia, and cranial nerve weakening is frequent. Mechanical ventilation is necessary for approximately one-third of hospitalized GBS patients because of respiratory muscle or oropharyngeal weakness. Over 50% of individuals experience autonomic abnormalities, which can manifest as tachycardia or more severe dysfunction.[7] Supportive ancillary testing, such as CSF analysis and electrodiagnostic testing, is necessary for GBS. Although these tests are necessary due to their limits and the need for fast treatment, they may be normal in the early stages. Merely 50% of patients exhibit elevated CSF protein content, with over 90% exhibiting it at the clinical nadir. The absence of CSF pleocytosis in GBS raises concerns regarding lymphomatous, carcinomatous, or infectious polyradiculoneuropathy. The clinical impression that peripheral neuropathy is the cause of sudden motor paralysis is supported by electrodiagnostic testing. Technology is used in nerve conduction studies (NCS) to assist in differentiating between axonal and demyelinating forms of neuropathy. The acuity of a patient's symptoms may be determined with the use of needle electromyography. [8,9]. Two treatment modalities are available for Guillain-Barré syndrome (GBS): intravenous immunoglobulin (IVIG) and plasma exchange. Given over the course of five days, IVIG at a rate of two grams per kilogram is intended to eliminate pathogenic antibodies, humoral mediators, and complement proteins implicated in the pathophysiology of GBS. Both therapies work just as well, but their effects are amplified if they are given within two weeks. No evidence of benefit with corticosteroids has been shown, either alone or in conjunction with IVIG and plasma exchange. Treatment for GBS is typically seen to hasten recovery; compared to untreated patients, treated individuals achieve independent ambulation 32 days sooner. [10].

Conclusion

The case of the 16-year-old boy emphasizes the significance of early identification and treatment for acute inflammatory demyelinating polyneuropathies such as Guillain-Barré syndrome (GBS). Due to its varied clinical presentations and quick course, GBS requires urgent care in order to minimize complications and improve prognoses. The prognosis of patients can be improved by prompting the delivery of therapies like intravenous immunoglobulin, and healthcare providers need to be mindful of issues like infections and vaccines. To improve patient care and therapeutic approaches, more research is required. To minimize the effects of the syndrome and maximize recovery, medical experts and patient education must work together.

References

1. Willison HJ, Jacobs BC, van Doorn PA. Guillain-barre syndrome. The Lancet. 2016 Aug 13;388(10045):717-727.
2. Le Doare K, Heath PT. (2013). An overview of global GBS epidemiology. Vaccine.;31: D7-12.
3. Dash S, Pai AR, Kamath U, Rao P. (2015). Pathophysiology and diagnosis of Guillain–Barré syndrome–challenges and needs. International Journal of Neuroscience. 125(4):235-240.
4. Verboon C, Doets AY, Galassi G, Davidson A, Waheed W. et al. (2019). Current treatment practice of Guillain-Barré syndrome. *Neurology*.93(1): e59-e76.
5. Kusunoki S. (2016). History of Guillain–Barré syndrome. *Clinical and Experimental Neuroimmunology*. 7(4):305-311.
6. Hughes RA, Rees JH. (1997). Clinical and epidemiologic features of Guillain-Barré syndrome. *Journal of Infectious Diseases*. 176(Supplement_2):S92-S98.
7. Burns TM. (2008). Guillain-barré syndrome. InSeminars in neurology. (Vol. 28, No. 02, pp. 152-167). © Thieme Medical Publishers.
8. Ropper AH. (1992). The Guillain–Barré syndrome. New England journal of medicine. 1992 Apr 23;326(17):1130-6.
9. Albers JW, Kelly Jr JJ. (1989). Acquired inflammatory demyelinating polyneuropathies: clinical and electrodiagnostic features. Muscle & Nerve: Official *Journal of the American Association of Electrodiagnostic Medicine*.;12(6):435-51.
10. Nguyen TP, Taylor RS. Guillain barre syndrome.
11. Safa A, Azimi T, Sayad A, Taheri M, Ghafouri-Fard S. (2021). A review of the role of genetic factors in Guillain–Barré syndrome. Journal of Molecular Neuroscience.;71(5):902-920.
12. Shang P, Feng J, Wu W, Zhang HL. (2021). Intensive care and treatment of severe Guillain–Barré syndrome. Frontiers in pharmacology.; 12:608130.
13. Brown WF, Feasby TE. (1984). Conduction block and denervation in Guillain-Barré polyneuropathy. Brain.107(1):219-239.
14. Pithadia A, Kakadia N. (2010). Guillain-Barre syndrome (GBS). Phar- ´ macol Rep ;62:220–232.
15. Van Koningsveld R, Schmitz PI, Van der Meche FG, Visser LH, Meulstee J.et al. (2004). Effect of methylprednisolone when added to standard treatment with intravenous immunoglobulin for Guillain-Barré syndrome: randomised trial. The Lancet. 363(9404):192-196.
16. Créange A, Lerat H, Meyrignac C, Degos JD, Gherardi RK. et al. (1998). Treatment of Guillain-Barré syndrome with interferon-β. The Lancet. 1998 Aug 1;352(9125):368-369.
17. Mangano K, Dati G, Quattrocchi C, Proietti L, Mazzarino C. et al. (2008). Preventive and curative effects of cyclophosphamide in an animal model of Guillain Barre syndrome. *Journal of neuroimmunology*. 196(1-2):107-115.

This work is licensed under Creative Commons Attribution 4.0 License

To Submit Your Article Click Here: **Submit Manuscript**

DOI:10.31579/2690-1897/218

Ready to submit your research? Choose Auctores and benefit from:

- fast, convenient online submission
- rigorous peer review by experienced research in your field
- rapid publication on acceptance
- authors retain copyrights
- unique DOI for all articles
- immediate, unrestricted online access

At Auctores, research is always in progress.

Learn more https://auctoresonline.org/journals/journal-of-surgical-case-reports-and-images