Guillain- Barre Syndrome: A Rare Immune Mediated Polyneuropathy

Nama Ravi Sneha Keerthi*, Neelathahalli Kasturirangan Meera

Department of Pharmacy Practice, Visveswarapura Institute of Pharmaceutical Sciences, Bengaluru, Karnataka, INDIA.

ABSTRACT

Guillain-Barré Syndrome (GBS) is a rare autoimmune disorder associated with demyelinating polyneuropathy requiring early diagnosis and accurate treatment. From birth to 30 years, the annual incidence is fairly uniform at 1.3 to 1.9 per 100,000. Clinical hallmarks include symmetrical flaccid muscle paresis and areflexia in the presence of an increased cerebrospinal fluid protein and electrophysiologic studies demonstrating demyelination. A case of 29 year old female presenting to a tertiary care hospital with GB syndrome and hypothyroidism is described. The significance of an appropriate diagnosis by the physician and subsequent management by the clinical staff is reviewed.

Key words: Autoimmune, Areflexia, Demyelination, Demyelinating polyneuropathy, Flaccid muscle paresis, Hypothyroidism.

INTRODUCTION

Guillain-Barré Syndrome (GBS) is an acute inflammatory polyneuropathy with an autoimmune etiology.[1] It may manifest as limb pain, numbness, weakness, paresthesias, or cranial nerve dysfunction associated with abated or absent deep tendon reflexes. The incidence of GBS is 1.72 patients per 100,000 person-years, with 50% increase for every 10-year increase in age.[2] From birth to 30 years, the annual incidence is fairly uniform at 1.3 to 1.9 per 100,000.[3] Recent studies show that GBS can be classified into 4 main clinical and electrophysiological subtypes such as Acute Inflammatory Demyelinating Polyneuropathy (AIDP), Acute Motor Axonal Neuropathy (AMAN), Acute Sensory Axonal Neuropathy (AMSAN) and Miller Fisher Syndrome (MSF).[4] GBS can be caused by viral or bacterial infections, including influenza and *Campylobacter jejuni*. It has also been reported as an adverse event following immunization. The 1976 swine-type influenza-A/ New Jersy vaccine was associated with an excess risk of GBS of 1 per 100,000 vaccines.[5] When patients present with rapidly progressive paralysis, the diagnosis of Guillain-Barre syndrome needs to be made at the earliest and may require hospital admission for precise monitoring treatment and supportive care.[6] The diagnosis being challenging, a thorough medical assessment may be required to exclude "mimic disorders" like diphtheria. Nerve conduction studies, CSF analysis and electromyography are significant to establish diagnosis. The goal of treatment is to reduce the severity of the illness and to assist in the patient's recovery. High dose immunoglobulin therapy, physical therapy to increase muscle flexibility and strength and plasmapheresis are notable treatment for GBS.[7] The case of GB syndrome in a 29 year old female patient is discussed here.

CASE REPORT

A 29 year old female from South India presented with fever, pain and shaking of lower limbs (calf muscles), pain in upper limbs, difficulty in walking, eating and guiding food to the mouth since 3 days with loss of appetite and she had two episodes of vomiting, non-bile/ blood tinged, containing food particles. Increased pain on walking,

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Address for correspondence:
Sneha Keerthi,
Visveswarapura Institute of Pharmaceutical Sciences, 22nd main, 24th cross, BSK-2nd stage, Bengaluru- 560070, Karnataka, INDIA.
Phone no: +91 9739929736
E-mail: sneha.nr@gmail.com

www.ijopp.org

sudden in onset and gradually progressive was reported. Patient noticed shaking of lower limbs when she tried to stand up and walk and was unable to walk without support. No history of similar complaints or trauma was ever reported. She had a past history of hypothyroidism and was treated with T. Thyronorm for 3 months, 4 years ago. Physical examination revealed that she was moderately built, nourished, conscious, co-operative, well oriented to time, place and person. Further examination revealed reduced plantar reflexes, rhomberg's test positive, finger nose test impaired, intention tremors, hypotonia of all 4 limbs and neck rigidity. Deep tendon reflexes and sensory loss was variable in both upper and lower limbs. Abdominal examination, respiratory and cardiac function were found to be normal.

The laboratory findings showed normal blood picture with abnormal TSH, T_3 and T_4 values. CSF analysis showed the presence of occasional lymphocytes with good number of RBC's and an elevated protein level of 276 mg/dL (normal range <50 mg/dL). Electroneuromyography showed axon loss demyelinating neuropathy.

The course of treatment during hospital stay included IV ceftriaxone 2 g BID × 5 days converted to tab. cefixime + clavulanic acid 200 mg BID × 5 days, IV pantoprazole 40 mg OD, tab paracetamol 650 mg SOS, IV normal saline at 50 ml/hr, IV human normal immunoglobulin (1 vial of 100 ml=5 g), tab. thyronorm 50 mcg OD, injection mecolight 1 amp. In 100 ml NS OD × 8 days converted to syp. mecolight 4C 10 ml BID × 2 days. On day 3 CSF aspiration was done for microbiological, cytological and biochemical analysis. This was followed by administration of IV immunoglobulin of 2 vials each on 3rd and 4th day, 3 vials each on 5th and 6th day, 4 vials on 7th day of hospitalization with a test dose given before starting each vial at MICU.

On day 5 and 6, patient showed reaction to human immunoglobulin transfusion and it was de-challenged. The patient was administered with inj. Pheneramine maleate 1 amp stat and tab paracetamol 650 mg SOS and subsequently immunoglobulin transfusion was continued. Except for hypotonia of all left lower limbs, all other parameters showed clinical progress on treatment. Patient was discharged with advice to continue syp. mecolight 10 ml BID for a month and tab thyronorm 50 mcg OD.

DISCUSSION

GBS is an acute inflammatory demyelinating polyradiculoneuropathy affecting the peripheral nervous system and a trigger for the same is an acute infection. It can affect all age groups; incidence increases with age, and there is a minor peak among young adults.[8] Complications like ventilator failure and cardiovascular instability are often observed. This disorder has no cure, but several treatments can ease symptoms and abate the duration of the illness.[8] Use of steroid hormones have not shown to be beneficial in clinical trials. In the present case, a young adult female patient had history fever and two episodes of vomiting and her total count, differential count and ESR were found to be normal. However, electroneuromyography showed axon loss demyelinating neuropathy. The CSF analysis indicated the presence of lymphage with RBC's and an elevated protein value. These findings meet with the required diagnostic criteria to confirm GBS. Thyroid function tests revealed abnormal levels of TSH. However, no literature states co-relation between thyroid function and GBS. The patient was treated with IV immunoglobulins with a test dose prior to every dose of administration. However, the patient developed fever and rashes which were treated after temporary de-challenge of immunoglobulin. By the end of hospital stay, the patient showed an overall improvement in the movement of all 4 limbs, finger nose test, eating and guiding food to the mouth and walking. Areflexia, a major complaint observed on the day of admission was also resolved. At discharge she was prescribed with a vitamin syrup and thyroid supplementation. The recovery period in general may take few weeks or as long as few years. After few years of the initial attack, about 3% may suffer a relapse of muscle weakness and tingling sensations. Individuals with GBS also face emotionally painful periods along with physical difficulties. Sudden paralysis and dependence on others for daily activities makes it difficult for them to accept, hence may need psychological counseling to help them adapt. Scientists are working on finding new treatments. GBS after a viral or bacterial infection suggests that certain characteristics of some viruses and bacteria may activate the immune system inappropriately.[7]

CONCLUSION

GBS is a rare and serious disorder hence it is very important to identify and refer the affected patients to undergo necessary investigation and receive appropriate therapy at the earliest.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

ABBREVIATIONS USED

GBS: Gullain- Barre Syndrome, **AIDP:** Acute inflammatory demyelinating polyneuropathy, **AMAN:** acute motor axonal neuropathy, **AMSAN:** Acute sensory axonal neuropathy, **MSF:** Miller Fischer syndrome, **CSF:** Cerebrospinal fluid, **TSH:** Thyroid stimulating hormone, **ESR:** Erythrocyte sedimentation rate.

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