Case Series

Varied Presentation of Guillain-barre Syndrome : Case Series and Review of Literature

Sumanyu Saxena[1], Prakhar Kumar[2]

Abstract

Background : We encountered a few cases with varied clinical presentation of Guillain-Barre Syndrome (GBS) in a Tertiary Care Centre and they were analysed to correlate certain clinical features with outcome at discharge.

Materials and Methods : Out of all the GBS patients admitted in the medical wards, 5 patients with varied presentation were analysed. Detailed history and physical examination was taken and necessary lab investigations were done including CSF study and Nerve conduction study.

Results : All the patients included in the study, irrespective of the clinical presentation and varied symptoms were diagnosed early with prompt initiation of treatment, were discharged with no or minimal residual symptoms.

Conclusion : There is a high percentage of Motor axonal variant of GBS in this study. There is predominance of Male patients and that too in a young adult age group.

Key words : Guillain-Barre Syndrome, Albumino-Cytologic Dissociation, Polyradiculopathy.

Guillain-Barré Syndrome (GBS) is an eponym, which contains multiple acute immune mediated poly neuropathies, although rare but serious post-infectious neuropathies. Resulting from autoimmune nerve destruction in the peripheral nervous system, it causes symptoms such as tingling, numbness, weakness and can even progress to quadriparesis[1]. GBS is one of the causes of acute, acquired flaccid, neuromuscular paralysis. Considered to be immune-mediated neuropathies, GBS and its variants are theorized to be post-infectious. Molecular mimicry is believed to play a key role in pathogenesis, as evidenced from animal models, where epitopes on peripheral nerve are targeted by and cross react with an immune response to an antecedent infection or other event[2]. For example, the lipo-oligosaccharide present in the outer membrane of Campylobacter jejuni is similar to gangliosides, which are components of peripheral nerves. Therefore, an immune response to C jejuni gastrointestinal infection can lead to a cross-reaction on host nerves[3]. GBS has been linked to numerous infections, most common being gastrointestinal or pulmonary illnesses. C jejuni can generate antibodies to specific gangliosides, including GM1, GD1a, GalNac-GD1a, and GD1b, which are strongly associated with AMAN and AMSAN. A large proportion of patients, up to 70% report an antecedent illness in the 1-6 weeks prior to presentation of GBS[4].

Editor's Comment :

- Although GBS and its variants may present with numerous presenting complaints, a strong suspicion based on detailed history and examination along with support from Electrodiagnostic studies and Lab evaluation, the precise diagnosis can be made out in time before it becomes life-threatening and hinders day to day life.

Other common infections like Influenza A & B, Cytomegalovirus, HIV, COVID-19 and Zikavirus have also been associated with GBS. Patients with the common Acute Inflammatory Demyelinating Polyneuropathy (AIDP) form have prominent demyelination on electro diagnostic studies and lymphocytic infiltration on sural nerve biopsies, while those with other forms such as Acute Motor Axonal Neuropathy (AMAN) form have prominent axonal loss without lymphocytic infiltration or complement activation and few degenerating nerve fibres. Demyelination is thought to start at the level of the nerve roots where the blood-nerve barrier is deficient. The breakdown of the blood-nerve barrier at the dural attachment allows transudation of plasma proteins into the cerebrospinal fluid. Demyelination blocks electrical saltatory conduction along the nerve. This causes conduction slowing and leads to muscle weakness. Distribution of GBS is worldwide with

Department of Medicine, Jawaharlal Nehru Medical College, AMU, Aligarh, Uttar Pradesh 202002
[1]MBBS, Junior Resident and Corresponding Author
[2]MBBS, MD, Assistant Professor
Received on : 26/10/2022
Accepted on : 09/07/2023

How to cite this article : Varied Presentation of Guillain-barre Syndrome : Case Series and Review of Literature. Saxena S, Kumar P. *J Indian Med Assoc* 2025; **123(8):** 57-62.

overall incidence ranging from 1 to 2 cases per 1,00,000 per year, incidence being slightly higher in males than in females[5].

Case Series

Case 1 :

A 24-year-old male presented with complaints of 1 episode of fever (unrecorded, low grade not a/w chills or rigor) around 10 days back, then he developed insidious onset weakness in Right upper limb. Two days later he developed weakness in Left upper limb and then 4 days later he developed weakness in bilateral lower limbs. This weakness was progressive in nature such that initially he could perform all the activities but within 2 days of onset of weakness he was even unable to hold food items in both his hand, comb his hair or wear his clothes. He was also unable to walk without support or get up from squatting position. Along with the weakness patient also developed pinching type of pain in his upper back area which was non-radiating and not associated with any hand or shoulder movement. There was no history of any sensory deficit, bladder or bowel incontinence. There was no history of any trauma or antecedent diarrhoea.

Examinations — On examination, patient was conscious and all higher functions and cranial nerves were intact. Single Breath Count (SBC) test was 16. On inspection, spine was normal and no deformity was apparent. On motor system examination, bulk, tone and nutrition was normal in both upper limbs. Power was 3/5 in both UL at shoulder, elbow and wrist in all range of motion. Hand grip was significantly reduced. In both lower limbs also, bulk, tone and nutrition was normal whereas power was 4/5 in both LL at hip, knee and ankle in all range of motion. All the deep tendon reflexes (biceps, triceps, supinator, knee and ankle) were mute bilaterally and plantar reflex was bilaterally flexor. Sensory examination including cerebellar signs was unremarkable.

Patient was investigated thoroughly including routine investigations which were normal. Radiographs of D/L and L/S spine were normal. Serum vitamin B12 levels were normal. CSF examination showed a total cell count of 02 cells and a protein level of 50mg/dl. Patient underwent Nerve Conduction Study(NCS) of all 4 limbs was done which showed CMAP amplitude was depressed in B/L median, ulnar, peroneal and tibial nerves, prolonged distal motor latency. F-wave and H-reflex was absent in all tested nerves. These findings were consistent with Acute Motor Axonal Neuropathy (AMAN) and patient was started on Intravenous Immunoglobulin (IVIG) at 2gm/Kg over 5 days. Patient showed improvement in his weakness by 4th day of treatment and backache was also improved. On 8th day of admission power in all 4 limbs had improved and he could walk without support and was able to perform personal activities. On a 2 week follow up patient was stable and had no residual weakness.

Case 2 :

A 14-year-old Male presented with the complaints of fever (unrecorded, low grade not associated with chills or rigor) around 4 days back, following which he developed insidious onset weakness in bilateral lower limbs resulting in difficulty in walking. One day later he also developed complaint of tingling sensation in both his upper limbs however there was no complaint of loss of power or restriction of any activities. He also complaint of pain abdomen and unable to micturate.There was no history of bowel incontinence or antecedent diarrhoea. There was no history of any backache or trauma.

Examinations — On examination, patient was conscious and oriented to time, place and person. All higher functions and cranial nerves were intact. Single breath count test was 22. On motor system examination, bulk, tone and nutrition was normal in both upper limbs. Power was 5/5 in both UL at shoulder, elbow and wrist in all range of motion. Hand grip was normal. In both lower limbs also, bulk, tone and nutrition was normal whereas power was 3/5 in both LL at hip, knee and ankle in all range of motion. All the deep tendon reflexes in all 4 limbs (biceps, triceps, supinator, knee and ankle) were absent. Plantar reflex was absent. Sensory examination including cerebellar signs was unremarkable.

Patient's investigations revealed normal serum B12 level, normal MRI Brain with spine screening. NCS revealed CMAP amplitude was depressed in B/L median, ulnar, peroneal and tibial nerves and prolonged distal motor latency. F-wave was inconsistent in all the tested nerves. These findings suggested a diffuse motor axonal neuropathy. CSF analysis showed total cells 10 and albumin level of 168mg/dl. Patient was started on IVIG at 2gm/Kg over 5 days. Patient showed improvement in his weakness by 6th day of treatment such that Patient could walk without support. On a 2 week follow-up patient had no residual weakness.

Case 3 :

A 33-year-old Male presented with the complaints of acute onset weakness of bilateral lower limbs for 2 days which was progressive in nature such that within a period of 2 days he was unable to walk without support and couldn't rise up from squatting position. Patient also complaint of weakness in both upper limbs for 2 days such that he could not perform his personal activities or raise his arms above his head. Patient also developed complaint of slurring of speech for 12 hours however there was no history of drooling of saliva. Patient had a history of painful lesions on his trunk 1 week back which was consistent with Herpes Zoster. There was no history of bowel or bladder involvement or any antecedent diarrhoea.

Examinations — On examination, patient was conscious and oriented to time, place and person. All higher functions were intact. There was loss of naso-labial fold prominence on the left side and slurring of speech, suggesting Facial nerve involvement. However taste sensation was intact. Single breath count test was 17. On motor system examination, bulk, tone and nutrition was normal in both upper limbs. Power was 4/5 in both UL at shoulder, elbow and wrist in all range of motion. Hand grip was normal. In both lower limbs also, bulk, tone and nutrition was normal whereas power was 4/5 in both LL at hip, knee and ankle in all range of motion. All the deep tendon reflexes in all 4 limbs (biceps, triceps, supinator, knee and ankle) were absent. Plantar reflex was flexor. Sensory examination including cerebellar signs was unremarkable.

Patient's investigations revealed normal routine investigations. B12 levels were normal. CSF analysis showed Total cells 25 with N20L80 differential and CSF albumin level was 800mg/dl. NCS of all 4 limbs showed Axonal changes in upper limb nerves involving motor and sensory fibres with prolonged distal motor latency and either absent or impersistent F-wave and H-reflex, suggesting radiculopathy. Patient was diagnosed as a case of Post Herpes Zoster Acute Inflammatory Demyelinating Polyradiculopathy with Facial nerve involvement and was started on IVIG at 2gm/Kg over 5 days. Patients weakness and speech improved after 7 days and was discharged on 9th day witch no residual paralysis or difficulty in speech.

Case 4 :

A 36-year-old Male presented with complaints of bilateral lower limb weakness for the last 5 days which was progressive in nature such that he could not walk without support within a span of 5 days. There was also difficulty in passing urine and stools for 2 days. Patient had a history of fever with chills 10 days back and was tested positive for NS1 Antigen and was conservatively managed.

Examinations — On examination, patient was conscious and oriented to time, place and person. All higher functions and cranial nerves were intact. On motor system examination, bulk, tone and nutrition was normal in both upper limbs. Power was 4/5 in both UL at shoulder and 3/5 in elbow and wrist in all range of motion. Hand grip was normal. In both the lower limbs, bulk and nutrition was normal whereas tone was decreased. Power was 2/5 in both LL at hip, knee and ankle in all range of motion. All the deep tendon reflexes in all 4 limbs (biceps, triceps, supinator, knee and ankle) were absent. Plantar reflex was flexor. Sensory examination including cerebellar signs was unremarkable.

Patient's investigations revealed normal routine investigations. MRI brain was normal. CSF analysis revealed total cell count 02 cells and protein 100mg/dl. Nerve conduction studies of bilateral ulnar, median and tibial nerve showed prolonged distal motor latency. CMAP amplitude and F-wave absent in b/l peroneal nerve and depressed in bilateral median, ulnar and tibial nerve. H-reflex bilaterly absent. Patient was diagnosed as a case of Post Dengue Fever Guillain-Barre Syndrome. Patient was started on IVIG at 2gm/Kg over 5 days. Two days after admission, patient developed weakness in bilateral upper limbs and difficulty in breathing and was placed onto Mechanical Ventilation. As the weakness persisted even after complete course of IVIG, patient was started on alternate day Plasmapheresis. Meanwhile tracheostomy was done. After 2 sessions patient showed improvement in weakness and patient was weaned off from ventilator after 4th session of plasmapheresis (15th day). Patients power improved to 4/5 in all limbs was discharged 5 days later.

Case 5 :

A 25-year-old Male presented with the complaints of acute onset weakness in all 4 limbs. Weakness was initially noticed by him in the R lower limb, which progressed to L upper limb, then the L lower limb and then to the R upper limb all within 9 days. Patient was unable to walk even without support or do any of his personal activities. There was no history of any

antecedent fever, diarrhoea or trauma of any kind. There was no history of any back pain. There was no history of bowel or bladder incontinence.

Examinations — On examination, patient was conscious and oriented to time, place and person. All higher functions and cranial nerves were intact. Single breath count test was 20. On motor system examination, bulk, tone and nutrition was normal in both upper limbs. Power was 2/5 in both UL at shoulder, elbow and wrist in all range of motion. Hand grip was weak. In both lower limbs also, bulk, tone and nutrition was normal whereas power was 2/5 in both LL at hip, knee and ankle in all range of motion. All the deep tendon reflexes in all 4 limbs (biceps, triceps, supinator, knee and ankle) were absent. Plantar reflex was flexor. Sensory examination including cerebellar signs was unremarkable.

Patient's routine investigations were normal. Serum B12 levels were normal. CSF analysis was done which showed total cell count of 5 and a protein level of 29mg/dl. NCS of all 4 limbs was done which showed prolonged distal motor latency of bilateral median, ulnar and tibial nerve. CMAP amplitude and F-wave absent in b/l peroneal nerve and depressed in bilateral median, ulnar and tibial nerve in both motor and sensory fibres. Patient was diagnosed as a case of AMSAN variant of GBS. Patient was started on IVIG at 2gm/Kg over 5 days. Patient showed improvement in his weakness by 4th day of treatment such that Patient could walk without support and was discharged on 8th day of admission with no complaints.

Age/ Sex	Antecedent Infection	Type of Weakness	CSF (Albumin/ Cells)	NCV Findings	Treatment Given	Outcome
24/M	N/A	Descending	40/02 (WNL)	AMAN	Inj IVIG	Cured
14/M	N/A	Ascending + Urinary Retention	168/08 (Albumino-cytologic dissociation)			
33/M	Herpes Zoster	Ascending + Facial Nerve Paralysis	800/15 (Albumino-cytologic dissociation)	AMAN	Inj IVIG	Cured
36/M	Dengue Fever	Ascending +B/B Involvement+ Respiratory Paralysis	100/02 (Albumino-cytologic dissociation)	AMAN	Inj IVIG+ Plasmapheresis +Mechanical Ventilation	Cured
25/M	N/A	Ascending +Sensory Involvement	29/05 (WNL)	AMSAN	Inj IVIG	Cured

Typical features of GBS include progressive and symmetric muscle weakness and absent /depressed deep tendon reflexes, may also have sensory symptoms and dysautonomia. Present within a few days-week after onset of symptoms, progressing over a period of two weeks. By 4 weeks, more than 90% of patients have reached the nadir of the disease. Weakness may vary from mild difficulty in walking to near complete paralysis of all limb, facial, respiratory, and bulbar muscles, depending on disease severity and clinical subtype. Classically, there is flaccid proximal and distal arm and leg weakness. Weakness is usually symmetric, starting in legs, but begins in arms or facial muscles in 10% of patients. Most patients progress to weakness in both arms and legs by the nadir. Facial nerve palsies occur in more than 50% with AIDP and oropharyngeal weakness eventually occurs in 50%. Occulomotor weakness occurs in about 15% of patients. Decreased or absent deep tendon reflexes in the arms or legs are found in approximately 90% of patients at presentation[4]. Most patients will develop hyporeflexia as symptoms progress to the nadir. Autonomic dysfunction may also develop in some patients with AIDP and may be prominent features in some variant forms of GBS. Paresthesias in the hands and feet are reported by more than 80% of patients, but sensory abnormalities on examination are frequently mild. Pain due to nerve root inflammation, typically located in the back and extremities, can also be a presenting feature and is reported during the acute phase by two-thirds of patients with all forms of GBS[6]. The prevalence of autonomic dysfunction ranges from 38 to 70% of patients with GBS[7], including Ileus, Hypertension, hypotension, Fever, tachycardia or bradycardia or Urinary retention. Patients with dysautonomia tended to have more frequent cardiogenic complications, hyponatremia and a higher burden of disability. Unusual features of GBS include papilledema with severely elevated Cerebro-spinal Fluid (CSF) protein, facial myokymia, hearing loss, meningeal signs, vocal cord paralysis, and mental status changes[8].

Common variant forms include : Acute Motor Axonal Neuropathy (AMAN), Acute Motor And Sensory Axonal Neuropathy (AMSAN).

Other rarer forms are Miller Fisher syndrome (MFS) & Bickerstaff Brainstem Encephalitis (BBE), Acute pan dysautonomia, Pure sensory GBS, Facial diplegia and distal limb paresthesia and Acute bulbar palsy.

Diagnosis of GBS in suspicious cases is based on

clinical features that are consistent with the syndrome and supported by diagnostic testing such as Electrodiagnostic Studies (EDx) and Cerebrospinal Fluid (CSF) analysis. Diagnostic criteria for GBS, originally proposed for research in 1978 by the National Institute of Neurological Disorders and Stroke (NINDS) [140], are widely used in clinical practice. CSF protein elevations may vary from 45 to 200 mg/dL for most patients, but protein elevations as high as 1000 mg/dL (10 g/L) have also been described. In one-third to upto one-half of patients, if CSF analysis is done earlier than one week from symptom onset, a normal CSF protein can be seen and therefore does not exclude the diagnosis of GBS. CSF cell count in GBS is usually normal (ie, <5 cells/mm^3). The albuminocytologic dissociation varies by time since symptom onset. In the first week of illness, it may be present in 50-66% of patients and in the third week in ≥75% of patients[9]. Electrodiagnostic studies commonly suggest Prolonged or absent F waves and absent H reflexes as the earliest findings, conduction blocks and Increased distal latencies with temporal dispersion of motor responses, Significant slowing or absent response on nerve conduction velocities not seen until the third or fourth week. Needle EMG of weak muscles show reduced recruitment or denervation[10].

Diagnostic imaging is typically reserved for patients with atypical symptoms to exclude alternative causes.

All patients with GBS should be monitored for deterioration with supportive care to address symptoms or their progression. Progressive neurologic weakness may help identify patients at risk for respiratory failure. More frequent neurologic evaluations (eg, every four to eight hours) are performed for those at high risk for deterioration and for those who may be rapidly worsening. For DVT prophylaxis, we use low molecular weight heparin and intermittent pneumatic compression or compressive stockings until patients are able to walk independently, unless a contraindication exists. Pain occurs in approximately two-thirds of patients with acute GBS and those in the recovery phase. Pain in GBS may be both somatic, related to inflammation of nerves, and neuropathic, secondary to axonal degeneration and can be managed by gabapentin, NSAIDs, opiods or morphine. Disease-modifying treatment in GBS include immunotherapy with either Plasma Exchange (PLEX) or Intravenous Immune Globulin (IVIG) and both are found to be effective[11]. The time to onset of recovery may be shortened by approximately 40 to 50% by treatment with PLEX or IVIG. For non-ambulatory adult patients with GBS who are within four weeks of symptom onset, we recommend treatment with PLEX or IVIG. When both therapies are equally available and there are no contra-indications for either, we prefer treating with IVIG because it is generally better tolerated and easier to administer than PLEX[12]. The choice between PLEX and IVIG is dependent on local availability and on patient preference, risk factors, and contra-indications[13]. Usual dosing of IVIG is 0.4 g/kg per day, given for 5 days to patients within four weeks of onset of GBS. Usual protocol for PLEX is four to six sessions over 8 to 10 days to patients within four weeks of onset of GBS. There are no trials comparing IVIG with placebo for the treatment of GBS; rather, the trials have compared IVIG with PLEX; IVIG and PLEX appear to have similar efficacy[14]. Acute-phase rehabilitation should include an individualized program of gentle strengthening involving isometric, isotonic, isokinetic, and manual-resistive and progressive-resistive exercises[15]. Rehabilitation should emphasize proper limb positioning, posture, and orthotics as well as nutrition. A device to help with communication may be necessary for patients with bulbar weakness. The long-term prognosis is favourable for most patients with GBS. Approximately 80% of patients are able to walk independently and more than half recover completely by one year[16]. However, severe motor impairments persist in more than 10%. Approximately 5 to 10% of patients with GBS have a prolonged course with several months of ventilator dependency and very delayed and incomplete recovery[17]. Mortality risk appears highest during recovery. In one study of 527 patients with GBS, the median time from symptom onset to death was 76 days, most frequently from respiratory or cardiovascular complications[18]. Approximately 3 to 7% of patients with GBS die despite intensive care[19]. Among patients who become ventilator dependent, mortality is approximately 20%. Causes of death include acute respiratory distress syndrome, sepsis, pulmonary emboli, and unexplained cardiac arrest.

Conclusion

GBS despite being an acute, life-threatening condition, may be cured if diagnosed early and prompt initiation of treatment is given. Practical conditions in developing countries may hinder this and may have

an impact on favourable outcome. After giving the treatment, the residual weakness if any, must be treated by physiotherapy regimens.

Funding : None

Conflict of Interest : None

Refrences

1 Govoni V, Granieri E — Epidemiology of the Guillain-Barré syndrome. *Curr Opin Neurol* 2001; **14(5):** 605-13.
2 Shahrizaila N, Lehmann HC, Kuwabara S — Guillain-Barré syndrome. *Lancet* 2021; **397(10280):** 1214-28. doi: 10.1016/S0140-6736(21)00517-1. Epub 2021 Feb 26.
3 Yuki N — A bacterium lipopolysaccharide that elicits Guillain-Barré syndrome has a GM1 ganglioside-like structure. *J Exp Med* 1993; **178(5):** 1771-5.
4 Fokke C — Diagnosis of Guillain-Barré syndrome and validation of Brighton criteria. *Brain* 2014; **137(Pt 1):** 33-43.
5 Sejvar JJ, Baughman AL, Wise M, Morgan OW — Population incidence of Guillain-Barré syndrome: a systematic review and meta-analysis. *Neuroepidemiology* 2011; **36:** 123.
6 Moulin DE, Hagen N, Feasby TE — Pain in Guillain-Barré syndrome. *Neurology* 1997; **48:** 328.
7 Flachenecker P — Autonomic dysfunction in Guillain-Barré syndrome and multiple sclerosis. *J Neurol* 2007; **254 Suppl 2:** II96.
8 Ropper AH, Wijdicks EFM, Truax BT— Guillain-Barré syndrome, *FA Davis* 1991. p.57.
9 Wong AH, Umapathi T, Nishimoto Y — Cytoalbuminologic dissociation in Asian patients with Guillain-Barré and Miller Fisher syndromes. *J Peripher Nerv Syst* 2015; **20:** 47.
10 Sumner AJ — The physiological basis for symptoms in Guillain-Barré syndrome. *Ann Neurol* 1981; **9 Suppl:** 28.
11 Kuwabara S — Guillain-Barrésyndrome. *Lancet* 2021; **397(10280):** 1214. Epub 2021 Feb 26.
12 Chevret S, Ac Hughes R, Annane D — Plasma exchange for Guillain-Barrésyndrome. *Cochrane Database Syst Rev* 2017; **2(2):** CD001798. doi: 10.1002/14651858.CD001798.pub3.
13 J Verboon C, A van Doorn P, Jacobs BC — Treatment dilemmas in Guillain-Barrésyndrome. *J Neurol Neurosurg Psychiatry* 2017; **88(4):** 346-52. doi: 10.1136/jnnp-2016-314862. Epub 2016 Nov 11.
14 van der Meché FG — *N Engl J Med* 1992.
15 Hughes RA — Supportive care for patients with Guillain-Barrésyndrome. *Arch Neurol* 2005.
16 Rajabally YA — Outcome and its predictors in Guillain-Barre syndrome. *J Neurol Neurosurg Psychiatry* 2012.
17 Kissel JT — Diagnosis and Management of Peripheral Nerve Disorders, Oxford University Press, New York 2001
18 van den Berg B — Mortality in Guillain-Barre syndrome. *Neurology* 2013.
19 ChiòA — Guillain-Barrésyndrome: a prospective, population-based incidence and outcome survey. *Neurology* 2003.