

Hyperacute Guillain Barre Syndrome (GBS); The Catastrophic Variant-A Rare Case Report

Abstract:

Guillain-Barré syndrome (GBS) also known as acute demyelinating polyradiculoneuropathy (AIDP) is an immunologically mediated rare neurological disorder. The basic pathogenic mechanism is regulated by molecular mimicry. Usually there is a history of preceding infection which occurs some weeks before the attack. The infections are gastroenteritis or upper respiratory. The clinical spectrum of ranges from mild weakness to devastating paralysis including respiratory failure. Majority of the cases recover but a few continue to have residual neurodeficit. The usual clinical course of GBS from the starting of weakness to development of maximum neurologic progression usually progresses over 4 weeks. Hyperacute GBS is a term used when the progression of weakness occurs within hours to days to maximum neurologic impairment. In this case report we present a 28 year old female who developed rapidly progressive, areflexic quadriparesis with respiratory muscle involvement requiring mechanical ventilatory support within nine hours. Clinical , laboratory and nerve conduction studies suggested a diagnosis of GBS.

Key Words: molecular mimicry, gastroenteritis, neurodeficit, quadriparesis, areflexic

Introduction:

The classic presentation of Guillain–Barré syndrome (GBS) is acute flaccid paralysis which is characteristic of symmetrical limb weakness and diminished or absent reflexes with a maximal severity in 4 weeks. Sensory abnormalities such as paraesthesias or numbness begin distally in general and follow a symmetrical pattern. Acute inflammatory demyelinating polyneuropathy (AIDP) and acute motor axonal neuropathy (AMAN) are the two most prevalent subtypes of GBS. Miller Fisher syndrome (MFS), which comprises

ophthalmoplegia, ataxia, and areflexia, is unusual subtype. GBS has a wide range of clinical manifestations, severity, and consequences. Post infectious GBS leads to production of antibodies causing an immunological reaction by reacting with gangliosides on membranes of neuron leading to nerve injury or conduction blockage. The subtype and clinical course of GBS are mainly governed by the kind of prior infection and the specificity of antiganglioside antibodies. GBS is currently managing with intravenous immunoglobulin (IVIg) and plasma exchange, both of which have been shown to be efficacious.[1]

Guillain-Barré syndrome has no age or sex predilection. Virtually anyone can be affected. The estimated Incidence of GBS per year is one in 100,000. The diagnostic criteria of GBS include progressive weakness from onset to nadir within 4 weeks. According to one study within 14 days, 75% patients reached their nadir, and 92% of those reach maximum incapacitation within 3 weeks.

Another study by Chio A et al; showed that the mean time was 9.7 days (median time, 7 days) from onset of symptoms to nadir[2]. In a study from Japan [3], the median time to nadir was 18.0 to 10.4 days for AIDP and 11.5 to 8.7 days for the acute motor axonal neuropathy (AMAN) variant. Hyperacute GBS is usually described when the onset of motor weakness reaches to its maximum within 24 to 48 hours. [4]

Case Report:

A 28-year-old female was referred to this hospital in critical care ambulance intubated on mechanical ventilatory support with a provisional diagnosis of GBS with respiratory paralysis. The patient was immediately shifted to medicine intensive care unit. Mechanical ventilation was continued. She was put on volume control mode. With a FiO₂ of 90% she was maintaining 94% saturation.

As per the history narrated by relatives, on the fateful morning around 6 AM she went for habitual cycling. After 20 minutes she felt tingling sensations in both lower limbs, she felt it was non specific and continued her exercise. In next 10 minutes the tingling sensation progressed upwards and involved the thighs and hip. At this point she became anxious and returned back home. In the next 4 hours she felt weakness in both lower limbs, such that she had difficulty in wearing and holding chappals and getting up from squatting position. She immediately informed the relatives and then was taken to the local practitioner. There was history of diarrhoea 10 days back which lasted for 2 days. There the attending doctor referred

her to a nursing home. In the Nursing home the physician advised MRI of brain and spine. By that time the weakness had progressed to both upper limbs. No sooner did she come out of the MRI machine, than she started having difficulty in breathing. An examination done there revealed tachypnoea, tachycardia with an SpO₂ of 86% while breathing ambient air. Her blood pressure was 178/102 mm of Hg. CVS and RS examination was normal.

She was immediately intubated and was put on mini mechanical ventilator of critical care/ cardiac ambulance and was referred to this hospital. The total duration of symptoms starting from tingling sensations and weakness with development of respiratory paralysis was 9 hours.

Examination in this hospital ICU revealed; Pulse -142/minute, regular, Blood pressure-156/98 mm of Hg. CNS examination revealed normal cognition, hypotonia in all 4 limbs with power grade 2/5 in 4 limbs on MRC scale. There was generalised areflexia. There were no obvious cranial nerve palsy. There was no objective sensory loss on examination.

Investigations: CBC- 4,300/mm³, Hb-13 gram%, KFT & LFT were normal. Serum Potassium-4.3 mEq/L, serum magnesium 2.7 mEq/L. Urine for porphobilinogen was negative. The MRI of brain and spine done earlier was normal.

CSF analysis revealed protein - 288 mg/l, glucose within normal range and 4 lymphocytes /mm³. A nerve conduction study showed delayed distal latencies over bilateral median and tibial nerves with decreased compound muscle action potentials (CAMP). The F latencies were not elicitable on all 4 limbs. The H response were not elicitable in bilateral tibial nerves. The sensory conduction parameters showed normal sensory nerve action potential (SNAP) values over bilateral upper and lower limb nerves. (Figure 1-3) Anti-ganglioside antibody analysis of the serum and CSF revealed high levels of anti-GQ1b.

A diagnosis of Hyperacute GBS was entertained and treatment was started with intra venous immunoglobulin (IVIg) at a dose of 2 grams per day for 5 days. She required tracheostomy after 7 days. In the subsequent 2 weeks her condition improved she was weaned off from ventilator and was transferred to general ward with power of 3/5 in upper limbs and 4/5 in lower limbs.

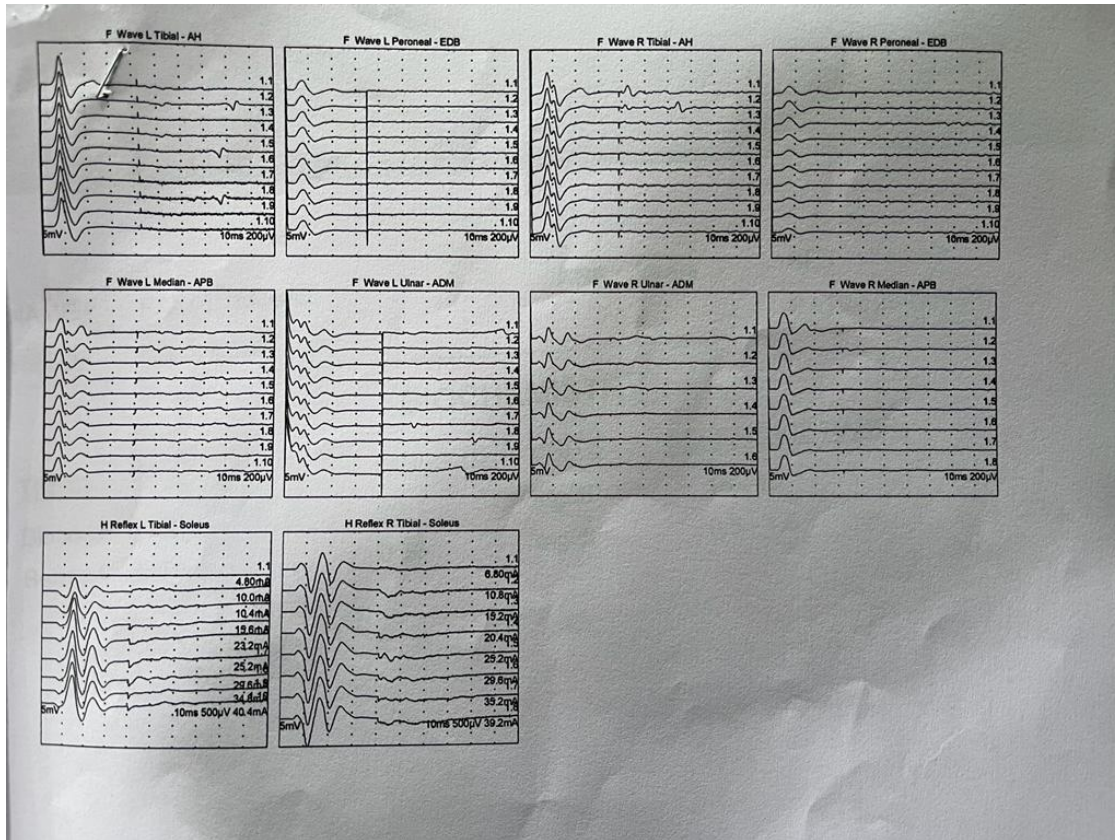


Figure-1: F wave latencies in various nerves

Table 1: MNC in various nerves:-

Nerve sites	Muscle	Latency ms	Amplitude mV	Amp. 2-4mV	Duration ms	Rel Amp %	Segments	Distance mm	Lat Diff ms	Velocity m/s
L Median-APB										
Wrist	APB	4.38	9.5	11.1	5.73		Wrist APB			
Elbow	APB	9.69	7.6	11.3	5.68	80.5	Elbow Wrist	270	5.31	51
R Median-APB										
Wrist	APB	3.85	7.4	10.2	5.16		Wrist APB			
Elbow	APB	7.97	6.9	9.5	5.63	93.4	Elbow Wrist	270	4.11	66

L- Ulnar ADM										
Wrist	ADM	3.07	0.5	5.7	6.2	100	Wrist ADM	70		
B. Elbow	ADM	6.77	3.4	5.1	7.19	715	B Elbow Wrist	290	3.7	78
R- Ulnar ADM										
Wrist	ADM	2.66	5.1	7.8	7.6	100	Wrist ADM	70		
B. Elbow	ADM	7.55	4.5	7.2	7.76	88.1	B Elbow Wrist	290	4.9	59
L- Peroneal EDB										
Ankle	EDB	3.8	4.7	6.5	6.98	100	Ankle EDB	80		
Fib Head	EDB	11.2	4.2	5.8	7.19	88.4	Fib Head Ankle	380	7.4	51
R-Peroneal EDB										
Ankle	EDB	3.96	5.1	7.1	6.93	100	Ankle EDB	80		
Fib Head	EDB	11.3	4.6	6.5	7.55	89.2	Fib Head Ankle	380	7.3 4	52
L tibial AH										
Ankle	AH	5.36	12.9	19.4	6.04	100	Ankle AH	80		
Pop Fossa	AH	14.64	11.3	16	6.25	88.3	Pop Fossa Ankle	400	9.2 7	43
R-Tibial AH										
Ankle	AH	5.99	8.3	16.5	5.57	100	Ankle AH	80		

Pop Fossa a	AH	13.75	7.6	11.6	7.08	91.2	Pop Fossa Ankle	400	7.7 6	52
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Table 2: SNC in various nerves:-

Nerve/ Sites	Rec. Site	Onset Lat ms	Peak Lat ms	Amp	Segments	Distance mm	Velocity m/s
L Median- Digit II (Antidromic)							
Wrist	Dig II	2.92	3.59	33.4	Wrist Dig II	130	45
R Median- Digit II (Antidromic)							
Wrist	Dig II	2.34	3.18	20.4	Wrist Dig II	130	55
L-Ulnar Digit V (Antidromic)							
Wrist	Dig V	2.5	3.33	15.4	Wrist Dig V	110	44
R-Ulnar- Digit V (Antidromic)							
Wrist	Dig V	2.08	2.86	18.1	Wrist Dig V	110	53
L-Sural Ankle (Calf)							
Calf	Ankle	1.09	1.98	11.1	Calf Ankle	140	128
R-Sural Ankle (Calf)							
Calf	Ankle	2.5	3.54	15.2	Calf Ankle	140	56
L-Superficial peroneal Ankle							
Lat Leg	Ankle	2.29	3.13	10.1	Lat Leg Ankle	140	61
R-Superficial Peroneal Ankle							
Lat Leg	Ankle	2.92	3.7	11.6	Lat Leg Ankle	140	48

Table 3: F Wave in various nerves:-

Nerve	Min M Lat ms	Max M Lat ms	Mean M Lat ms	Min F Lat ms	Max Lat ms	Mean F Lat ms	Min F- M ms	Max F-M ms	Mean F-M ms
L Tibial Ah	NR	NR	NR	NR	NR	NR	NR	NR	NR
L peroneal EDB	NR	NR	NR	NR	NR	NR	NR	NR	NR

Table Showing the motor and sensory nerve conductions in various nerves

Nerve	Min M Lat ms	Max M Lat ms	Mean M Lat ms	Min F Lat ms	Max F Lat ms	Mean F Lat ms	Min F M ms	Max F M ms	Mean F M ms
R Tibial AH	NR	NR	NR	NR	NR	NR	NR	NR	NR
R Peroneal ADB	NR	NR	NR	NR	NR	NR	NR	NR	NR
L median ABP	NR	NR	NR	NR	NR	NR	NR	NR	NR
L Ulnar ADM	NR	NR	NR	NR	NR	NR	NR	NR	NR
R Ulnar ADM	NR	NR	NR	NR	NR	NR	NR	NR	NR
R Median ABP	NR	NR	NR	NR	NR	NR	NR	NR	NR

H Reflex:-

Nerve	H Lat ms	Lat H max ms
L Tibial Soleus	NR	NR
R Tibial Soleus	NR	NR

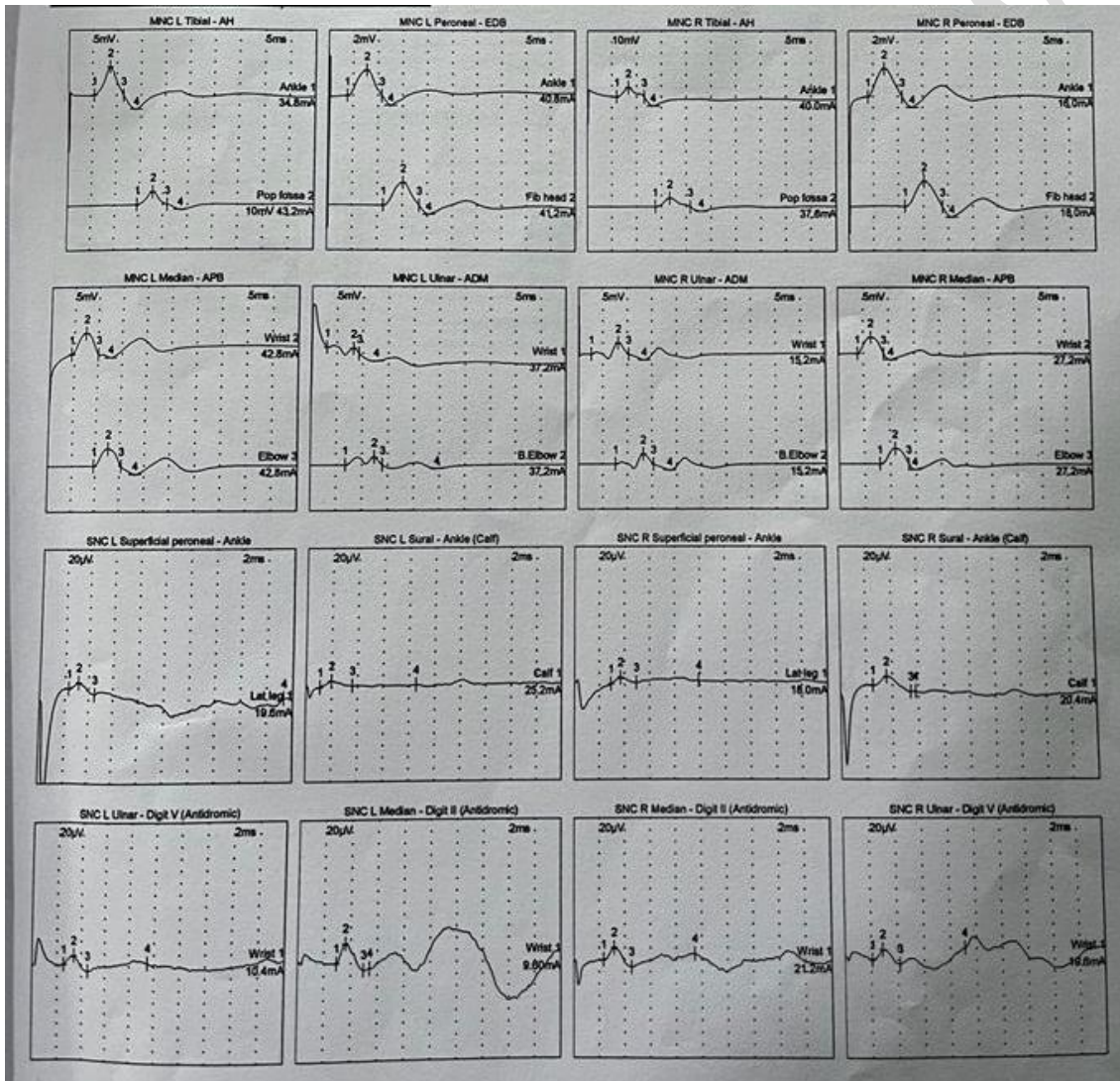


Figure 2: Showing the H-reflex patterns

Discussion:

In spite of wide variety of clinical presentations in GBS, patients often present with weakness and sensory symptoms in the legs that progress to the arms and cranial muscles. The patient's clinical history as well as neurological, electrophysiological, and cerebrospinal fluid (CSF) investigations are used to make the diagnosis of GBS[5]. Other illnesses with a clinical presentation similar to GBS must be sought out. Electrophysiological investigations show that the PNS is impaired and can differentiate between GBS subtypes. The disease can progress rapidly, and most GBS patients attain their maximal impairment within two weeks.[6] GBS causes respiratory failure in around 20% of patients, necessitating mechanical ventilation. Involvement of autonomic nervous system leads to arrhythmias and hemodynamic instability.[7]

In patients with rapidly ascending weakness without CNS involvement, distal paraesthesias, or sensory loss, diminished or absent reflexes, dysautonomia, radicular pain, increased protein level in CSF, sensorimotor neuropathy in electrophysiological studies; GBS should be considered as a diagnosis.

Intravenous immunoglobulin (0.4 g/kg for 5 days) and plasmapheresis are both effective GBS therapies.[8]

'Hyperacute' GBS is an extremely rare condition. Steiner I; et al reported a series of 5 cases of hyperacute GBS. The duration between the onset of symptoms to development of quadriplegia ranged from 20-36 hours. Three patients had respiratory paralysis. Two patients had history of upper respiratory tract infections, two patients had history of gastroenteritis within the previous 10 days. Only one patient had abnormal CSF findings in form of albumin-cytological dissociation. Four patients received IVIg Therapy and one patient was treated with plasma exchange therapy. Three of those five patients recovered fully. Several studies had shown that a shorter interval to nadir suggest an adverse prognostic outcome.

[2,3] In some studies it was noted that axonal involvement was associated with poor prognosis. [2,5,6] History of previous infection and younger age had a favourable outcome according to a study. [3]

In our case the onset of symptoms to nadir was only nine hours, still our patient showed a favourable outcome . This report should draw attention to an unusually rapid progression of GBS. Other possible neurologic entities like acute botulism, myasthenic crisis, periodic paralysis, brainstem stroke, neuroparalytic snake bite should be also entertained and ruled out

Immunomodulatory therapy are the cornerstone treatment for GBS. It should be started when the patient is unable to walk a distance of approximately 10 meters independently, or when there is rapid progression of symptoms. The two recommended immunomodulatory therapies are ; intravenous immunoglobulin (IVIg) and plasma exchange (PE). IVIg therapy is efficacious if initiated within 14 days of symptom onset. Plasma exchange therapy has a window of 4 weeks. [7,8,9] As far as efficacy is concerned both therapies are equally effective in treatment.[10]

Conclusion:

Hyperacute GBS is an aggressive form of demyelinating polyradiculoneuropathy where onset of symptoms to maximum neurodeficit occur in 48 hours or less. Other possible disorders that present similarly should be ruled out first. Early progression of weakness is also a heralding sign of respiratory paralysis and need of mechanical ventilation. Immunomodulatory therapy should be initiated at the earliest.

Ethical Approval:

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

Consent

As per international standard or university standard, patient's written consent has been collected and preserved by the author(s).

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