Case study

Hyperacute Gullain 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. We present a case of 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:

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. [1]

The classic presentation of Guillain-Barré syndrome (GBS) is an acute inflammatory polyradiculoneuropathy manifesting as ascending, areflexic motor weakness with or without autonomic and cranial nerve palsy. Apart from the classic form other variants of GBS exist.
The diagnostic criteria of GBS require the development of progressive weakness from onset to nadir within 4 weeks. According to one study 75% of patients reached their nadir within 2 weeks, and 92% of those reach maximum incapacitation within 3 weeks.
Another study by Chio A et al; showed that the mean time from onset of symptoms to nadir was 9.7 (8 6.9) days (median time, 7 days) [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 7.8 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 24to 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 Gullain Barre Syndrome (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/mm 3, Hb-13 gram%, Kidney function test and liver function tests were within normal limits. 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.
Lumbar puncture revealed clear fluid with normal pressure. CSF protein - 288 mg/l, normal glucose and 4 lymphocytes /mm ^{3} . 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 gms 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.

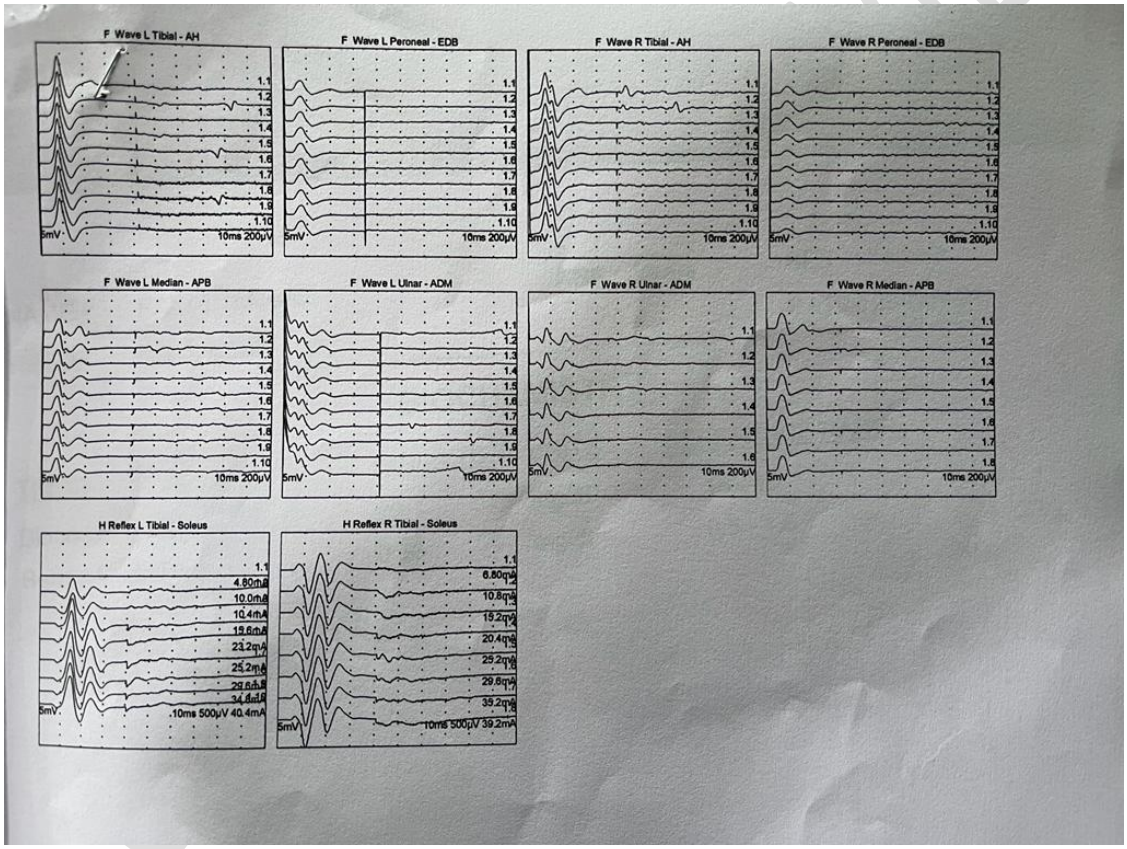
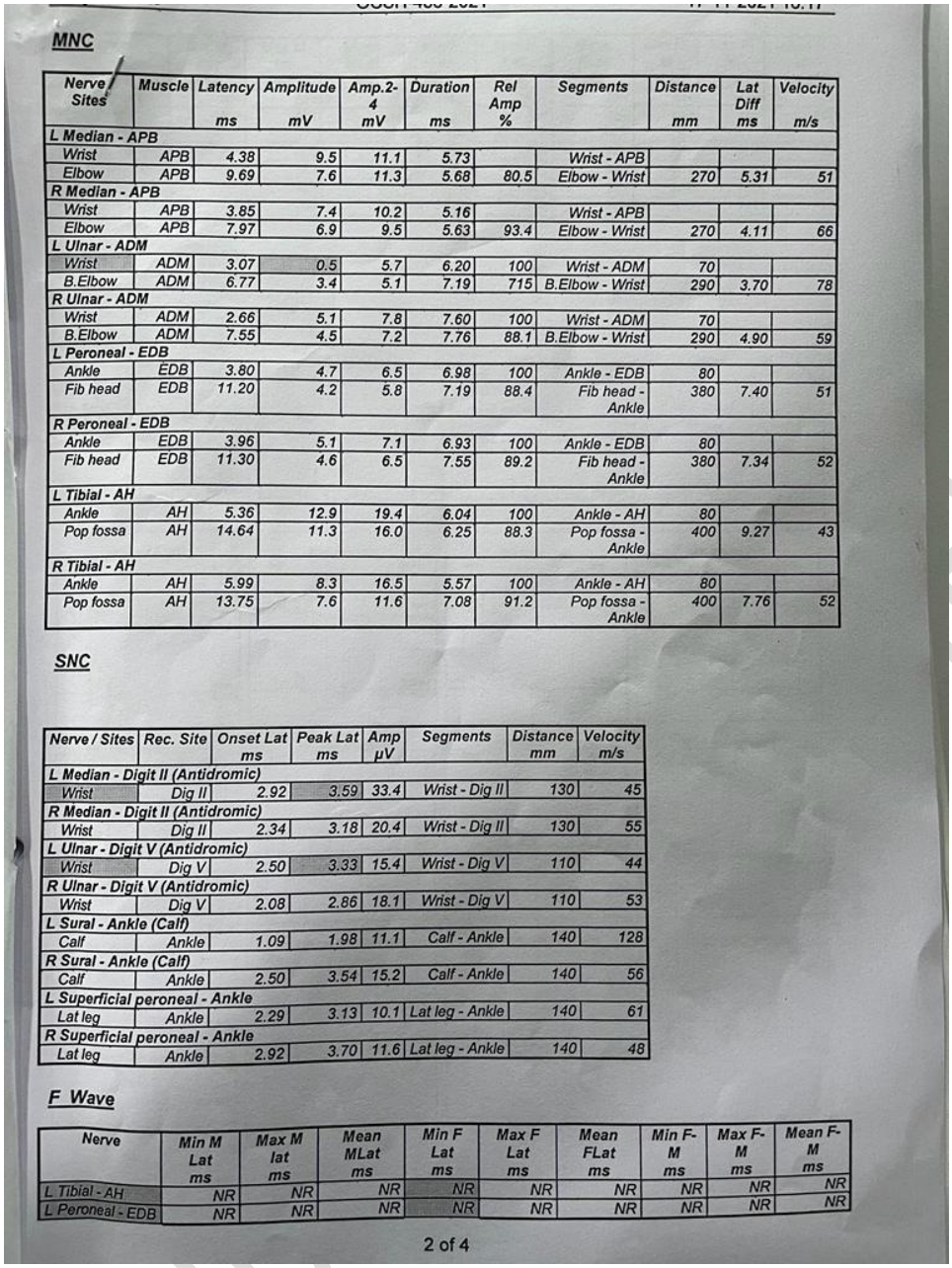


Figure-1: F wave latencies in various nerves



MNC

Nerve / Sites	Muscle	Latency ms	Amplitude mV	Amp.2-4 mV	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	6.20	100	Wrist - ADM	70		
B.Elbow	ADM	6.77	3.4	5.1	7.19	715	B.Elbow - Wrist	290	3.70	78
R Ulnar - ADM										
Wrist	ADM	2.66	5.1	7.8	7.60	100	Wrist - ADM	70		
B.Elbow	ADM	7.55	4.5	7.2	7.76	88.1	B.Elbow - Wrist	290	4.90	59
L Peroneal - EDB										
Ankle	EDB	3.80	4.7	6.5	6.98	100	Ankle - EDB	80		
Fib head	EDB	11.20	4.2	5.8	7.19	88.4	Fib head - Ankle	380	7.40	51
R Peroneal - EDB										
Ankle	EDB	3.96	5.1	7.1	6.93	100	Ankle - EDB	80		
Fib head	EDB	11.30	4.6	6.5	7.55	89.2	Fib head - Ankle	380	7.34	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	16.0	6.25	88.3	Pop fossa - Ankle	400	9.27	43
R Tibial - AH										
Ankle	AH	5.99	8.3	16.5	5.57	100	Ankle - AH	80		
Pop fossa	AH	13.75	7.6	11.6	7.08	91.2	Pop fossa - Ankle	400	7.76	52

SNC

Nerve / Sites	Rec. Site	Onset Lat ms	Peak Lat ms	Amp µV	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	2.50	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	2.50	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	3.70	11.6	Lat leg - Ankle	140	48

F Wave

Nerve	Min M Lat ms	Max M lat ms	Mean MLat ms	Min F Lat ms	Max F Lat ms	Mean FLat 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

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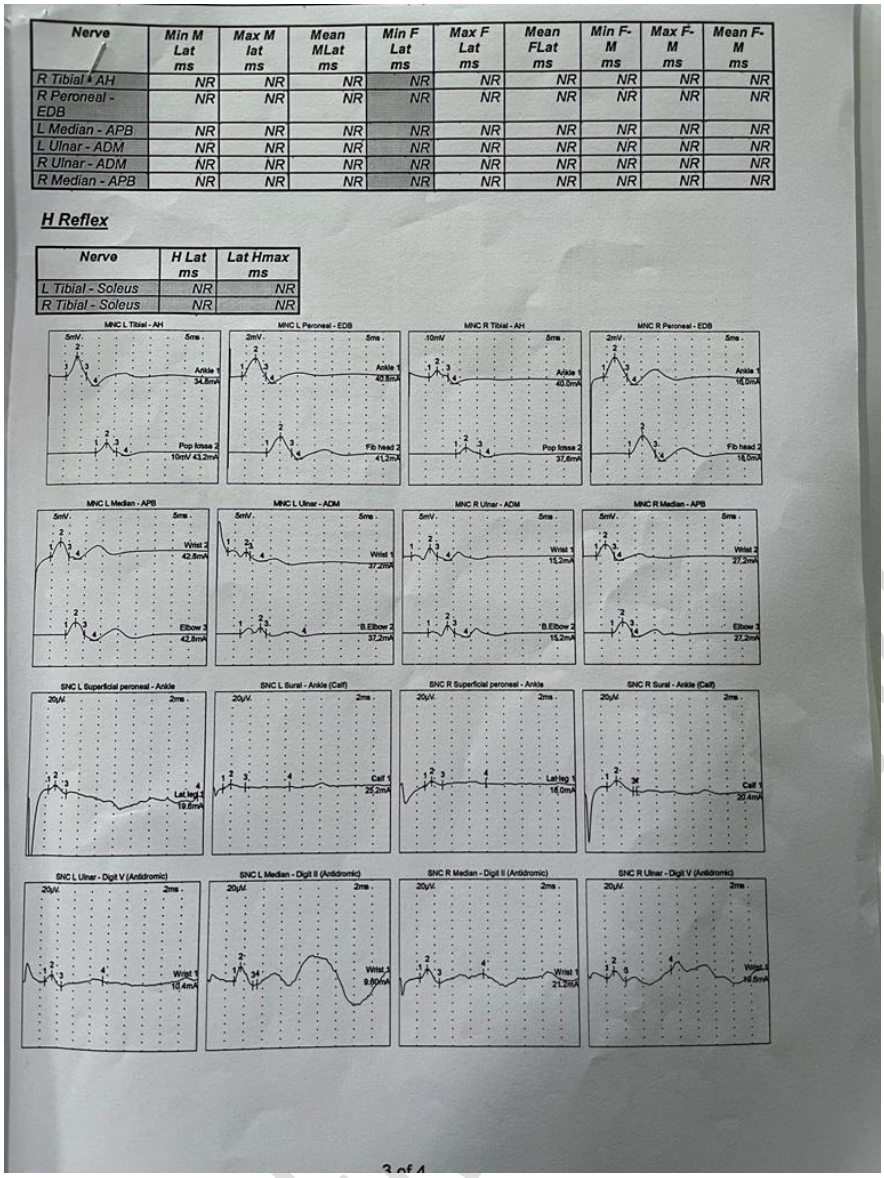
Figure-2: Showing the motor and sensory nerve conductions in various nerves



Nerve	Min M Lat ms	Max M lat ms	Mean MLat ms	Min F Lat ms	Max F Lat ms	Mean FLat 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 - EDB	NR	NR	NR	NR	NR	NR	NR	NR	NR
L Median - APB	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 - APB	NR	NR	NR	NR	NR	NR	NR	NR	NR

H Reflex

Nerve	H Lat ms	Lat Hmax ms
L Tibial - Soleus	NR	NR
R Tibial - Soleus	NR	NR

Figure 3: Showing the H-reflex pattens

Discussion:

'Hyperacute' GBS is an extremely rare condition. Steiner I; etal 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 patient 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. [4] Several studies had shown that a shorter interval to nadir suggest and 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 best effective when started within 2 weeks of onset of weakness. 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.

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