



**ORIGINAL RESEARCH PAPER**

**Medicine**

**A RARE AND ATYPICAL PRESENTATION OF AIDP WITH CORTICAL VENOUS SINUS THROMBOSIS**

**KEY WORDS:**

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**Introduction:**

Guillain-Barre syndrome (GBS) was first reported by Landry in 1859 and later detailed by Guillain, Barré and Strohl, in 1916. The disease has become well-known internationally under the name of GuillainBarré Syndrome[1].

The Guillian - Barre Syndrome is one of the commonest forms of polyneuropathy. The reported incidence rates for GBS are 1-2 per 1 00,000 population. The lifetime likelihood of any individual acquiring GBS is 1 in 1000. Available Indian literature indicates a peak incidence between June, July and September - October. In the Western Countries GBS is common in the 5th decade, but in India it occurs more commonly in younger age. GBS is equally common in men and women and can occur at any age. There is a male preponderance among the hospitalized population [2]

Guillain Barre Syndrome also known as an Acute Inflammatory Demyelinating Polyneuropathy (AIDP) is an acute demyelinating polyradiculopathy of uncertain etiology which may present with facial nerve involvement in 27-50% of cases, often bilaterally [3]. Over half of Guillain Barre syndrome patients experience symptoms of viral respiratory or gastrointestinal infections during the 1-3 weeks prior to the onset of neurological symptoms. Clinical criteria, spinal fluid protein elevation, and nerve conduction abnormalities are the mainstay of diagnosis [4]

**Case report:**

A 50 year old male patient presented with c/o headache since 5 days. On neuroimaging patient diagnosed as cortical venous sinus thrombosis for which anticoagulant therapy was started. On 3<sup>rd</sup> day of treatment he developed left sided upper limb weakness and after 2 days he also developed right sided upper limb weakness without sensory disturbance and areflexic motor paralysis.

**Past h/o :**

K/c/o hypertension since 5 years was on antihypertensive therapy  
No p/h/o of convulsion/DM/TB/jaundice/BT/any major surgery

**Personal history:**

Vegetarian, sleep pattern normal,  
Bladder and bowel habits not altered  
No addiction

**Family h/o:** Not significant

**General examination:**

Pt is well built and fairly nourished  
Temp. -normal  
Pulse - 82/min regular  
BP - 128/70 mm hg  
No evidence of pallor/icterus/clubbing /cyanosis /lymphadenopathy/edema.

**Systemic examination:**

CVS, RS, GIT system examination were unremarkable.

**CNS Examination:**

Conscious and well oriented in time, place and person.

**Tone**

	Rt	Lt
UL	↓	↓
LL	N	N

**Power**

	Rt	Lt
UL	0	0
LL	5	5

**Reflex:**

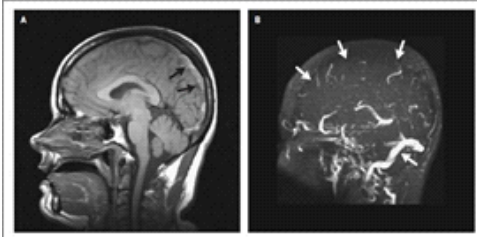
Planter: B/L dorsiflexion  
Pupils-B/L RTL

	K	A	T	S	B
Rt	Ab	Ab	Ab	Ab	Ab
Lt	Ab	Ab	Ab	Ab	Ab

**Investigations:**

CBC, PT/INR, ELECTROLYTES, RFT, LFT, ECG ,Chest x ray were within normal limits.  
CPK total -normal  
Homocystine -elevated (60mcmol/l)  
Vit b12 -430 pg/ml  
CSF r/m- WNL

MRI brain-cortical venous sinus thrombosis



**MRI of Sinus Thrombosis.**  
In Panel A, a T<sub>2</sub>-weighted MRI scan obtained with the spin-echo technique provides a sagittal view of a hyperintense signal in the thrombosed superior sagittal sinus (arrow). In Panel B, a magnetic resonance venogram obtained without the administration of contrast material reveals the absence of a signal in the superior sagittal sinus (upper arrow) and a normal flow signal in the transverse and sigmoid sinuses (lower arrow) as well as in a number of veins.

MRI cervical spine with whole spine screening was normal, done to rule out central causes.

MRI b/l brachial plexus was normal, done to r/o plexopathy.

CECT thorax and abdomen was normal done to r/o paraneoplastic syndrome.

**NCV study: Axonal demyelinating neuropathy involving axillary and musculocutaneous nerve bilaterally.**

Motor Nerve Studies											
UPPER LIMB											
Nerve	Conduction Velocity (m/s)	Amplitude (mV)	Area (mV.s)	Latency (ms)	Conduction Time (ms)	Distal Latency (ms)	Conduction Velocity (m/s)	Amplitude (mV)	Area (mV.s)	Latency (ms)	Conduction Time (ms)
Right Median Nerve (50 Hz)	54.00	14.00	1.00	3.00	10.00	1.00	54.00	14.00	1.00	3.00	10.00
Left Median Nerve (50 Hz)	52.00	12.00	0.80	3.00	10.00	1.00	52.00	12.00	0.80	3.00	10.00
Right Ulnar Nerve (50 Hz)	56.00	16.00	1.20	3.00	10.00	1.00	56.00	16.00	1.20	3.00	10.00
Left Ulnar Nerve (50 Hz)	54.00	14.00	1.00	3.00	10.00	1.00	54.00	14.00	1.00	3.00	10.00
Right Radial Nerve (50 Hz)	58.00	18.00	1.40	3.00	10.00	1.00	58.00	18.00	1.40	3.00	10.00
Left Radial Nerve (50 Hz)	56.00	16.00	1.20	3.00	10.00	1.00	56.00	16.00	1.20	3.00	10.00
Right Peroneal Nerve (50 Hz)	50.00	10.00	0.60	3.00	10.00	1.00	50.00	10.00	0.60	3.00	10.00
Left Peroneal Nerve (50 Hz)	48.00	8.00	0.40	3.00	10.00	1.00	48.00	8.00	0.40	3.00	10.00

Sensory Nerve Studies											
UPPER LIMB											
Nerve	Conduction Velocity (m/s)	Amplitude (mV)	Area (mV.s)	Latency (ms)	Conduction Time (ms)	Distal Latency (ms)	Conduction Velocity (m/s)	Amplitude (mV)	Area (mV.s)	Latency (ms)	Conduction Time (ms)
Right Median Nerve (50 Hz)	50.00	10.00	0.60	3.00	10.00	1.00	50.00	10.00	0.60	3.00	10.00
Left Median Nerve (50 Hz)	48.00	8.00	0.40	3.00	10.00	1.00	48.00	8.00	0.40	3.00	10.00
Right Ulnar Nerve (50 Hz)	52.00	12.00	0.80	3.00	10.00	1.00	52.00	12.00	0.80	3.00	10.00
Left Ulnar Nerve (50 Hz)	50.00	10.00	0.60	3.00	10.00	1.00	50.00	10.00	0.60	3.00	10.00
Right Radial Nerve (50 Hz)	54.00	14.00	1.00	3.00	10.00	1.00	54.00	14.00	1.00	3.00	10.00
Left Radial Nerve (50 Hz)	52.00	12.00	0.80	3.00	10.00	1.00	52.00	12.00	0.80	3.00	10.00
Right Peroneal Nerve (50 Hz)	48.00	8.00	0.40	3.00	10.00	1.00	48.00	8.00	0.40	3.00	10.00
Left Peroneal Nerve (50 Hz)	46.00	6.00	0.20	3.00	10.00	1.00	46.00	6.00	0.20	3.00	10.00

**DISCUSSION:**  
Above findings are suggestive electrophysiological evidence of axonal demyelinating neuropathy involving axillary and musculocutaneous nerve bilaterally.

**Treatment:**

Plasmapheresis and limb physiotherapy was started. After 5 cycles of plasmapheresis weakness improved and patient discharged on oral anticoagulants for CVST.

**Impression:**

Typically GBS involves B/L lower limb followed by B/L upper limb (ascending paralysis), in our case there was involvement of B/L upper limb without lower limb involvement and without sensory disturbance, thus an atypical presentation of AIDP. NCV study s/o axonal demyelinating neuropathy involving axillary and musculocutaneous nerve b/l and it was improved after plasmapheresis and physiotherapy s/o of AIDP with rare association of CVST.

**Discussion:**

AIDP is acute frequently severe and fulminant polyradiculoneuropathy which is rapidly evolving areflexic ascending paralysis with or without sensory disturbance is most common subtype of GBS (90%). Adults are more affected than children. Weakness typically evolved over hour to few days and frequently accompanied by tingling & diasthesis & it recover rapidly.

The usual pattern is an ascending paralysis that may be first noticed as rubbery leg.[7]

Maximal weakness generally develops within 12-14 days of the onset of neurological symptoms. Although cessation of symptom progression within 4 weeks is often regarded as a necessary criterion for the diagnosis of Guillain – Barre Syndrome (Asbury and Cornblath 1990).

Tendon reflexes are usually lost early in the disease. Total areflexia occurs in over 80 per cent of patients at some stage of the illness. Approximately half the patients develop cranial – nerve palsies, usually in the wake of severe ascending limb weakness (Loffel Rossi, Mumenthaler, et al 1977 ; Winer, Hughes, and Osmond 1988). Isolated unilateral or bilateral facial palsy is the commonest cranial – nerve lesion in Guillain – Barre syndrome.

Bulbar palsy and weakness of the muscles of mastication are the next commonest cranial nerve abnormalities. Ocular palsy only occurs in about 10 per cent of patients.[4].

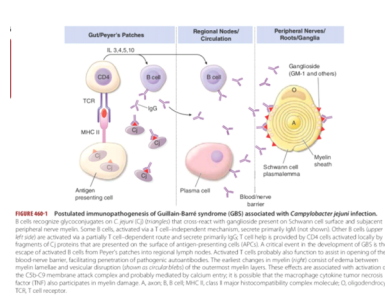
Bowel & bladder are not affected usually. Autonomic involvement is common, so wide fluctuation of blood pressure, postural hypotension & cardiac arrhythmia.

70% cases occur after 1-6 week of acute infection, usually respiratory and gastrointestinal caused by c. Jejunii(most common),CMV ,EBV, HIV, Hep-B, mycoplasmapneumonie & swine flu vaccine(rare).

**Pathogenesis:**

It is mostly autoimmune in nature results from immune response to non-self antigen (like virus vaccine) than misdirect to host nerve tissue through resemble to epitops (molecular mimicry) mostly gangliosides affected.

Most commonly antigm1 antibody is detected which first attack on Schwann cell surface and cause widespread damage. Also activate macrophage, causes lymphocytic infiltration and variable secondary axonal damage.



**FIGURE 400-1** Pathogenesis of Guillain-Barre syndrome (GBS) associated with Campylobacter jejuni infection. B cells recognize glycosylated antigens of C. jejuni (Cj) through their cross-react with ganglioside present on Schwann cell surface and adjacent peripheral nerve myelin. Some B cells, activated via a T cell-independent mechanism, secrete primarily IgM (not shown). Other B cells later differentiate and secrete a primarily T cell-dependent IgG and secrete primarily IgG. T cell help is provided by CD4 cells activated locally by fragments of Cj proteins that are presented on the surface of antigen-presenting cells (APCs). A critical event in the development of GBS is the escape of activated B cells from lymph nodes into peripheral nerve nodes. Activated T cells probably also function to assist in opening of the blood-nerve barrier, facilitating penetration of pathogenic autoantibodies. The earliest changes in myelin (right) consist of edema between myelin lamellae and axolemma disruption (shown as condensation of the myelin sheath). These effects are associated with activation of the CD4-CD8 membrane attack complex and probably mediated by calcium entry. It is possible that the macrophage-cytokine tumor necrosis factor (TNF) also participates in myelin damage. A, axon; B, B cell; MHC, class II major histocompatibility complex molecule; Cj, oligodendrocyte; TCS, T cell receptor.

**[8]**

**CSF:** Elevated protein (1-10 gm/l or 100-1000ug/dl) without accompanying pleocytosis.

**Electro diagnosis:** Nerve conduction studies are a dependable and early diagnostic indicator of GBS, and in instances with a typical clinical and EMG presentation, one can dispense with the CSF analysis. The most frequent early findings are a reduction in the amplitudes of muscle action potentials, slowed conduction velocity, or conduction block in motor nerves. Prolonged distal latencies (reflecting distal conduction block) and prolonged or absent F response (indicating affection of proximal parts of nerves) are other important diagnostic findings, all reflecting demyelination.[5].

**Treatment:**

1. Plasmapheresis 40-50 ml/kg plasmaexchange four to five times/week.
2. IV immunoglobulin five daily infusion total dose as per 2gm/kg body weight.[6]
3. Physiotherapy.

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