ORIGINAL RESEARCH PAPER

Medicine

KEY WORDS:

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

Dr. Chitralekha Vora Associate Professor, Department of Medicine, B.J. Medical College, Civil Hospital, Ahmedabad

Dr. Harshad Chovatiya* 3rd Year Resident, Department of Medicine, B.J. Medical College, Civil Hospital, Ahmedabad *Corresponding Author

Dr. Suresh Bishnoi 3rd Year Resident, Department of Medicine, B.J. Medical College, Civil Hospital, Ahmedabad

Dr. Kinjal Patel 3rd Year Resident, Department of Medicine, B.J. Medical College, Civil Hospital, Ahmedabad

Dr. Saurabh Biswas 2nd Year Resident, Department of Medicine, B.J. Medical College, Civil Hospital, Ahmedabad

Dr. Vishal Chauhan 2nd Year Resident, Department of Medicine, B.J. Medical College, Civil Hospital, Ahmedabad

Dr. Minesh Chaudhary 2nd Year Resident, Department of Medicine, B.J. Medical College, Civil Hospital, Ahmedabad

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].

T he 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 likely hood 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]

Gullian 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 Gullian 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 3rd 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

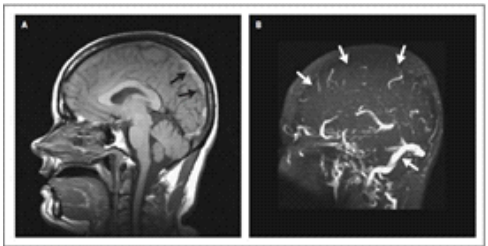
	Rt	Lt
UL	0	0
LL	5	5

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



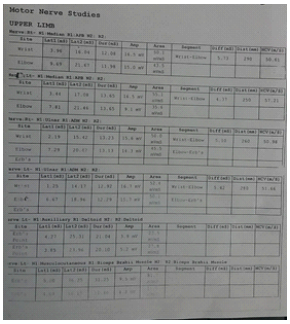
MRI of Sinus Thrombosis.
In Panel A, a T_1-weighted MRI scan obtained with the spin–echo technique provides a sagittal view of a hyperintense signal in the thrombosed superior sagittal sinus (arrows). In Panel B, a magnetic resonance venogram obtained without the administration of contrast material reveals the absence of a signal in the superior sagittal sinus (upper arrows) and a normal flow signal in the transverse and sigmoid sinuses (lower arrow) as well as in a number of veins.

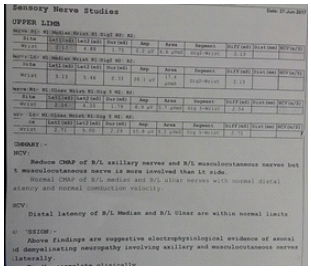
MRI cevical spine with whole spine screening was normal, done to rule out central causes.

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 demylinating neuropathy involving axillary and musculocutaneous nerve bilaterally.


Motor Nerve Studies
UPPER LIMB


Sensory Nerve Studies
UPPER LIMB
SUMMARY:-
NCV:
Reduce CMAP of B/L axillary nerves and B/L musculocutaneous nerves but t musculocutaneous nerve is more involved than Lt side.
Normal CMAP of B/L median and B/L ulnar nerves with normal distal atency and normal conduction velocity.
NCV:
Distal latency of B/L Median and B/L Ulnar are within normal limits
CESSION:-
Above findings are suggestive electrophysiological evidence of axonal id demyelinating neuropathy involving axillary and musculocutaneous nerves laterally

Treatment:
Plasmapheresis and limb physiotherapy was started. After 5 cycles of plasmapheresis weakness improved and patient discharged on oral anticoagulats 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 demylinating neuropathy involving axillary and musculocutaneous nerve b/l and it was improved after plasmapheresis and physiotherapy s/o of AIDP with rare association of CSVT.

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. Jejuni(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.

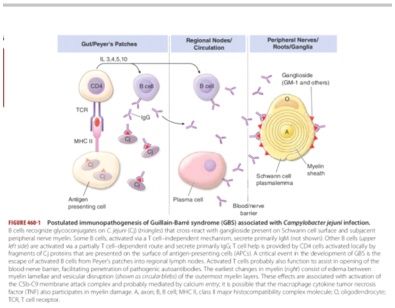


FIGURE 668-1 Postulated immunopathogenesis of Guillain Barré syndrome (GBS) associated with Campylobacter jejuni infection.

[8]
CSF: Elevated protein (1-10 gm/l or100-1000ug/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. Plasmapharesis 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.

References:
[1] Hughes RA, Cornblath DR. Guillain-Barré syndrome. Lancet. 2005 Nov;366 (9497):1653–66.

[2] AK Meena 29th Chapter, Peripheral Neuropathy, API Text Book of Medicine, 9th edition Vol. II Jaypee Brothers Med Publishers, (Pg No. 1533)
[3] Killic R, Ozdek A, Felekl S, Safak MA, Samim E : A case presentation of Bilateral Simultaneous Bell's Palsy. Americal Journal of Otolaryngology 2003, 24 (4) : 271 – 273.
[4] Micheal Donoghy ; Disorders of Peripheral nerves Chapter 18th ; Acute Idiopathic Polyneuritis 18.25; Brain's Disease of the Nervous System 10th Edition ; Published in the United States of Oxford University Press Inc., New York (Pg no. 599 – 601).
[5] Maurice Vitor, M.D., & Allan H Ropper, M.D., ; Chapter 46, Diseases of Peripheral Nerves ; Adams & Victors, Principles of Neurology 7th Edition; Mc. Graw - Hill Medical Publishing division. (Pg No.1381-1382).
[6] Anthony A. Amato, Richard J. Barohn 384th Chapter Peripheral Neuropathy, Harrison Text Book of Medicine 18th Edition, Vol II Graw Hill Publishers, 2012, Pg 3448.
[7] Stephan L. Hauser, Anthony A. Amato Guillain barre syndrome and other immune mediated neuropathies chapter no 460-Harrission's (19th edition) principal of internal medicine Page no2694.
[8] Stephan L. Hauser, Anthony A. Amato Guillain barre syndrome and other immune mediated neuropathies chapter no 460-Harrission's (19th edition) principal of internal medicine Page no2696 figure no 460-1.