MYELOPATHY OR ATYPICAL PRESENTATION OF GBS

Nikhil Verma[1], Deepali Kaushik[2], Deepika Uikey[3]

[1]Assistant Professor, Department of Medicine, ESIC Medical College and Hospital, Faridabad.
[2]Senior Resident, Department of Medicine, ESIC Medical College and Hospital, Faridabad.
[3]Senior Resident, Department of Dermatology, ESIC Medical College and Hospital, Faridabad.

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PRESENTATION OF CASE

A 53-year-old male presented to the Emergency Department of ESI Hospital complaining of recent onset tingling and numbness in both of his legs about, which he described as feeling sleepy. The symptoms begin to progress in the descending manner and patient started complaining of tingling sensation in both of his hands, also his symptoms begin slowly within a week following history of fever 15 days back, there was no history of trauma, diabetes mellitus, tuberculosis and hypertention. Patient was a nonalcoholic nor had he ever complain of the current symptoms. Although, most patients develop a weakness, which tends to begin in the lower extremities due to demyelination of the peripheral nerves resulting in ascending paralysis and also a loss of cranial nerve function.[8]

DIFFERENTIAL DIAGNOSES

1. Acute myelopathy (example from compression, transverse myelitis, vascular injury).
2. Chronic inflammatory demyelinating polyneuropathy.
3. Conversion disorder/hysterical paralysis.
4. Paraneoplastic neuropathy.
5. Spinal cord syndromes, particularly post infection.

CLINICAL DIAGNOSIS

The most common cause of acute muscle weakness associated with peripheral neuropathy in adults is Guillain-Barre syndrome.[1] Guillain-Barre Syndrome (GBS) is also known as polyradiculoneuritis and Chronic Inflammatory Demyelinating Polyradiculopathy (CIDP).[2] Clinical hallmarks are symmetrical flaccid muscle paresis and are flexia in the presence of an increased cerebrospinal fluid protein content and electrophysiologic studies demonstrating evolving demyelination.[3] Muscle stretch reflexes are depressed in most patients and the sensory loss is variable. Difficulty with walking, running, climbing stairs and getting up from a chair are usual early complaints. This weakness is usually symmetric and can also involve the upper extremities.[4] Causes are viral infection,[5] surgery,[6,7,8] blood transfusion[9] and mycoplasma infection.[3] Examination of this patient revealed a healthy person in otherwise good condition and afebrile. Generalised weakness of lower extremity was noted when testing the quadriceps, hamstring and soleus muscle group sensory loss was variable in both his hands and feet. Deep tendon reflexes of upper extremity was 1+ and lower extremities was absent.

PATHOLOGICAL DISCUSSION

Its aetiology is unclear, although it has been associated with both cell and humoral-mediated autoimmune mechanisms.[10] The pathogenesis of Guillain-Barre syndrome shows oedematous changes proximal to the spinal nerve root at the junction of the anterior and posterior roots. The myelin sheaths soon become irregular at about the third day. Lymphocytes appear about the ninth day and phagocytosis on the eleventh day. These findings were found in severe cases of this syndrome that led to death.[11] Blood tests showed increased WBC and MRI brain was done, which was suggestive of nonspecific ischaemic changes in supratentorial brain with age-related celebrate atrophic changes. CSF examinations was done, which was normal. Glucose was 2 and the protein were 4.1. Electrodiagnostic study of upper limb was done, which was suggestive of polyradiculoneuropathy with non-recordable F wave. Electrodiagnostic study of lower limb was also done, which was also suggestive of polyradiculoneuropathy with non-recordable F wave showing finding consistent with GB syndrome versus thoracolumbosacral radiculopathy. MRI dorsal spine with contrast was done, which was normal. MRI cervical spine with contrast was done, which revealed disc degenerative changes and MRI lumbar spine with contrast also revealed disc degenerative changes.

DISCUSSION OF MANAGEMENT

Treatment for patients with Guillain-Barre syndrome depends on whether they have mildly acute, severely acute or chronic involvement. The incidence of death in one study was 1.5% to 8% of patients.[12] Another study listed the death rate at 4%.[13] Other common complications include ventilatory failure and cardiovascular instability for which intensive care support should be utilised. Ventilatory failure is caused by involvement of airway and respiratory muscles, particularly the diaphragm. The use of corticosteroids demonstrated no benefit. The only well-investigated efficacious immunomodulatory therapy is plasmapheresis. High majority of these patients ultimately have a good functional recovery and can recover completely at about 12 months after onset.[14] Patient was given IVIG for five days

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Corresponding Author:
Dr. Nikhil Verma,
Flat No. 101, A-1 Block,
ESIC Medical College and Hospital Campus, Faridhabad-121001.
E-mail: dr.nick.ver@gmail.com
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with physiotherapy symptoms drastically improved two months later when patient visited OPD. His sensory deficit was very much improved and his leg strength had improved.

ELECTRO-DIAGNOSTIC STUDIES: (LOWER LIMBS)
Motor Nerve Conduction Study: Bilateral Common peroneal and Posterior tibial nerves showed normal CMAP amplitudes with normal distal latencies and conduction velocities.
Sensory: Bilateral Sural nerve showed normal Snap's amplitudes with normal peak latencies and conduction velocities.
F-Waves: Non recordable F-wave latencies were seen in Bilateral Common peroneal and Posterior tibial nerves.

Needle Electromyography (EMG):
No abnormal spontaneous activities were seen in Bilateral Tibialis anterior, Gastrocs and Vastus Lateralis muscles.
Prolonged duration and polyphasic MUAP'S on voluntary activity were seen in Bilateral Tibialis anterior, Gastrocs and Vastus Lateralis muslces.
Recruitment was incomplete in all tested muscles.

IMPRESSION
ENMG study is remarkable for unrecordable F-wave latencies suggestive of polyradiculopathy.
This findings is consistent with early GB syndrome versus lumbosacral radiculopathy. (For Clinical correlation)

Mri Lumbar Spine with Contrast

SEQUENCES OBTAINED USING A 1.5 TESLA SUPER CONDUCTING MRI UNIT:
High resolution FSE-T1W & FSE-T2W sections in the sagittal and axial planes.
STIR section in the sagittal plane. Contrast study has also done.

Findings:
Degenerative changes are seen in the form of marginal osteophytes, facet joint arthropathy and partial disc dessication at multiple levels appearing in the form of loss of T2W bright signal intensity.

Small focal fatty changes is seen in L1 vertebral body.

Otherwise lumbar vertebrae are normal in height and marrow signal intensity.
Posterior elements are normal. No marrow edema or ossessous destruction is seen.
Visualised intervertebral discs show variable desiccation.

L2-3: Intervertebral disc appears to be normal. No central canal or neural forminal narrowing.
L3-4: Intervartebral disc appears to be normal. No central canal or neural forminal narrowing.
L4-5: Intervertebral disc appears to be normal. No central canal or neural forminal narrowing.
L5-S1: Intervertebral disc appears to be normal. No central canal or neural forminal narrowing.

No evidence of primary lumbar canal stenosis is seen.
The lower end of the cord and conus medullaris reveals normal Signal Intensity.

No Pre/paravertebral collection is seen.

Impression: Findings reveal: Disc degenerative changes.
To be correlated clinically.

Mri Lumbar Spine with Contrast

MRI CERVICAL SPINE

MR imaging of the cervical spine was performed and high resolution T1-and T2-weighted serial sections obtained in the sagittal and axial planes using a phased-array surface coil on a 1.5 Tesla scanner.

Fingings:

There is straightening of cervical lordoitic curvature.
The vertebral bodies are normal in size, shape, alignment and signal characteristics.

There is evidence of generalized spondylotic changes in the form of marginal
Osteophytes and degenerative complete disc desiccation at multiple levels seen as reduction
of bright signal intensity on T2 W images.

No significant disc bulge/protrusion is noted at any other level.
The spinal cord is normal in size, contour and signal intensity. No intrinsic parenchymal lesion is seen.
The thecal sac is normal and CSF demonstrates normal signal intensity.
The craniovertebral junction is normal.
The cervico-medullary junction is normal.
No intraspinal mass or pre/paravertebral collection is seen.

Opinion: MR findings reveal disc degenerative changes.

To be correlated clinically.

Motor Nerve Conduction:

Nerve and site	Latency (ms)	Amplitude (μV)	Latency Difference (ms)	Distance (mm)	Conduction Velocity (m/s)
Peroneal L					
Fibula (head)	2.8	4.3	2.8		
Popliteal fossa	8.8	3.5	6.0	300	50
Tibial L					
Ankle	3.9	8.0	3.9		
Popliteal fossa	11.4	4.5	7.5	320	43
Peroneal. R					
Fibula (head)	3.3	4.7	3.3		
Popliteal fossa	10.1	3.7	6.8	300	44
Tibial. R					
Ankle	3.5	8.5	3.5		
Popliteal fossa	11.6	4.6	8.1	320	40

Sensory Nerve Conduction:

Nerve and Site	Onset Latency (ms)	Peak Latency (ms)	Amplitude (μv)	Latency Difference (ms)	Distance (mm)	Conduction Velocity (m/s)
Sural. R						
Lower leg	2.1	2.8	14	2.1	130	6
Sural. L						
Lower leg	2.2	2.7	15	2.2	130	59

F-Wave Studies

Nerve	M-Latency	F-Latency
Peroneal.L	NR	NR
Tibial.L	NR	NR
Peroneal.R	NR	NR
Tibial.R	NR	NR

ELECTRO –DIAGNOSTIC STUDIES: (LOWER LIMBS)

Motor Nerve conduction study: Bilateral Common peroneal and Posterior tibial nerves showed normal CMAP amplitudes with normal distal latencies and conduction velocities.

Sensory: Bilateral Sural nerve showed normal SNAP'S amplitudes with normal peak latencies and conduction velocities.
F- Waves: Non recordable F-wave latencies were seen in Bilateral Common peroneal and Posterior tibial nerves.

Needle Electromyography (EMG):
No abnormal spontaneous activities were seen in Bilateral Tibialis anterior, Gastrocs and Vastus Lateralis muscles.
Prolonged duration and polyphasic MUAP's on voluntary activity were seen in Bilateral Tibialis anterior, Gastrocs and Vastus Lateralis muscles.
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IMPRESSION:
ENMG Study is remarkable for unrecordable F-wave latencies suggestive of polyradiculopathy.
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(For Clinical Correlation)

MRI BRAIN PLAIN

MR imaging of the brain was done and TSE T1, TS3 T2, T2 FLAIR AND SW1 – coronal, sagittal and axial images were acquired.

Few discrete hyperintensities are seen in bilateral frontal lobe and periventricular white matter, suggestive of non-specific ischemic changes.

There is generalized prominence of the ventricular system and subarachnoid spaces, consistent with age –related cerebral atrophic changes.

Rest of the cerebral parenchyma is normal in signal intensity with maintained grey and white matter differentiation.

Bilateral basal ganglia and thalami are normal in volume and signal intensity.

The mid-brain, pons and medulla are central and appear normal in signal intensity.

Cerebellum is normal in signal intensity.

Septum is in midline.

Major intracranial flow voids preserved.

OPINION: MRI findings are suggestive of non-specific ischemic changes in supra-tentorial brain with age-related cerebral atrophic changes.

ELECTRO-DIAGNOSITC STUDIES: (UPPER LIMGS)

Motor Nerve conduction study: Bilateral Median and ulnar nerves showed decreased CMAP amplitudes with normal distal latencies and conduction velocities.

Sensory: Bilateral Median and Ulnar nerves showed normal SNAP amplitudes with normal peak latencies and conduction velocities.
F-waves: non recordable F- wave latencies were seen in Bilateral Median and Bilateral Ulnar nerves.

Needle Electromyography (EMG):
No abnormal spontaneous activities were seen in Bilateral FDL, brachioradialis, deltod muscles.

Prolonged duration and polyphasic MUAP'S on voluntary activity were seen in Bilateral
FDI, brachioradialis, deltoid muscles.
Recruitment was incomplete in all tested muscles.

IMPRESSION:
Abnormal ENMG study suggestive of axonal motor polyradiculoneuropathy.
For Clinical correlation.

FINAL DIAGNOSIS

A diagnosis of Guillain-Barre syndrome was made after utilising the criteria that strongly support the diagnosis of Guillain-Barre syndrome. Features required to rule out diagnoses other than Guillain-Barre syndrome would include- no history of hexacarbon abuse, no evidence of porphyria, no history or culture evidence of diphtheria, no history or evidence of lead intoxication, symptoms not purely sensory, no evidence of poliomyelitis, botulism, toxic neuropathy or tick paralysis diagnosis of GBS was made on the basis of history and onset of symptoms and above-mentioned reports.[1] Guillain-Barre syndrome is a neurological disorder resulting primarily in muscle paralysis that in most cases is symmetrical. Patients may have mild involvement or severe involvement, which may in a small percentage lead to death. Patients may present to a chiropractic office with symptoms of polyradiculoneuralgia, which may at first be interpreted as a radicular pain pattern of spinal origin. It is extremely important to identify and urgently refer, potential severe cases in order to have the appropriate investigations (i.e., electrodiagnostic studies, spinal tap) instituted and have the appropriate care administered. Differential diagnosis is of utmost importance.

REFERENCES

[1] Miller RG. Guillain-barre syndrome. Postgraduate Medicine 1985;77(7):57-64.
[2] Mendell JR. Chronic inflammatory demyelinating polyradiculopathy. Annual Rev Med 1993;44:211-219.
[3] Hund EF, Borel CO, Cornblath DR, et al. Intensive management and treatment of severe Guillain-Barre syndrome. Crit Care Med 1993;21(3):433-446.
[4] Mendell JR. Chronic inflammatory demyelinating polyradiculopathy. Annual Review Medicine 1993;44:211-219.
[5] Ormerod IE, Cockerell OC. Guillain-Barre syndrome after herpes zoster infection, a report of 2 cases. European Neurology 1993;33(2):136-318.
[6] Stambough JL, Quinlan JG, Swanson JD. Guillain-Barre syndrome following spinal fusion for adult scoliosis. Spine (Phila Pa 1976) 1990;15(1):45-46.
[7] Ennis JH, Bednar DA. Lumbosacral fusion in a patient with recurring Guillain-Barre syndrome and acute brachial neuritis. J Spine Disord 1992;5(2):217-218.
[8] Ginn D. Guillain-Barre syndrome. An uncommon but severe illness. Postgrad Med 1991;90(5):145-416:153-156.
[9] Nadkarni N, Lisak RP. Guillain-Barre Syndrome (GBS) with bilateral optic neuritis and central white matter disease. Neurology 1993;43(4):842-843.
[10] Murray DP. Impaired mobility: Guillain-Barre syndrome. J Neurosci Nurs 1993;25(2):100-104.
[11] Haymaker WE, Kernohan JW. The Landry-Guillain-Barre syndrome: clinicopathologic report of 50 fatal cases and a critique of the literature. Medicine (Baltimore) 1949;28(1):59-141.
[12] Ropper AH. Management of Guillain-Barre syndrome. In: Ropper AH, Kennedy SK, Zervas NT, eds. Neurological and neurosurgical intensive care. Baltimore: University Park Press 1983:p. 163.
[13] Hughes RA, Newsom-Davis JM, Perkin GD, et al. Controlled trial of prednisolone in acute polyneuropathy. Lancet 1978;2(8093):750-753.
[14] Smith GD, Hughes RA. Plasma exchange treatment and prognosis of Guillain-Barre syndrome. Quarterly J Medicine 1992;85(306):751-760.