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Case Report

A CASE OF GUILLAIN BARRE WITH HYPERREFLEXIA

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ABSTRACT
Guillain-Barré syndrome (GBS) is an acquired acute autoimmune polyradiculoneuropathy, areflexia and flaccid weakness are essential for diagnosis.16 year old boy presenting with acute onset flaccid quadriparesis after a brief febrile illness about a month back without higher function, sensory, autonomic and any bulbar symptom. Peculiarity of the case being hyperreflexia in all four limbs.

***Key Words**: GBS: Guillain Barre Syndrome, Hyperreflexia*

INTRODUCTION
GBS is an acquired immune polyradiculoneuropathy consisting of aidp, amsan, aman, mfs which is marked by weakness and areflexia. Diagnostic criteria include areflexia along with flaccid weakness. Here is a case of polyradiculoneuropathy with areflexia.

CASES
A 16 year old school boy presents with complaints of acute onset flaccid weakness in all four limbs 2 weeks back without any significant complaint suggestive of sensory function, bladder bowel and autonomic function or any bulbar problems. Past history reveals a history of fever lasting for 3 days a month preceding the illness without any complaint of any episode of diarrhea or respiratory infection. He was almost bed ridden and neither being able to move his limbs not able to lift his head from the bed or move from side to side following the event. He was admitted in some local hospital in his village where he found his power in limbs improving roughly after the first week. Detailed neurological examination revealed a normal higher mental function without any cranial nerve affection or neck rigidity. Power in upper limbs were 4-/5 both proximal group and distal group of muscles, and 4+/5 in his lower limbs equally again in proximal and distal groups. Normal sensory examination, no cerebellar deficit was found. However deep tendon reflexes in his upper limbs and lower limbs were exaggerated 3+ (biceps, triceps, supinator, knee, ankle). All superficial reflexes were normal, plantar reflexes were unresponsive on both sides.He was having a single breath count of 30 or more and his vitals were stable and normal.Cardiovascular, respiratory and gasterointestinal examination did not reveal anything significant. In relation to the quadriparesis with hyperreflexia in a normal sensorium person, a MRI of cervical spine was sought which was normal with no significant cord compression effects. Routine counts and biochemistry with electrolytes did not reveal any abnormality. Csf examination revealed albuminocytological dissociation with cell count of 3/cumm in csf and protein of 77mg/dl. Csf glucose was 71 mg/dl with serum glucose 106mg/dl.

Emg-Ncs study of all four limbs revealed decreased nerve conduction velocities in upper and lower limbs along with conduction block in bilateral tibial nerve signifying an acquired demyelinating type of neuropathy.sensory testing was however normal .f wave in upper limb were lost and in lower limbs showed impersistence and chronodispersion signifying demyelination of motor radicles as well diminished amplitude was also noted in few motor nerves signifying an axonal involvement. Thus a diagnosis of acquired predominantly demyelinating (with axonal) type of motor polyradiculoneuropathy was made.

DISCUSSION

Guillain-Barré syndrome (GBS) is an acute, frequently severe, and fulminant polyradiculoneuropathy that is autoimmune in nature.

Subtypes of Guillain-Barré Syndrome (GBS) :

Subtype	Features	Electrodiagnosis	Pathology
Acute inflammatory demyelinating polyneuropathy (AIDP)	Adults affected more than children; 90% of cases in western world; recovery rapid; anti-GM1 antibodies (<50%)	Demyelinating	First attack on Schwann cell surface; widespread myelin damage, macrophage activation, and lymphocytic infiltration; variable secondary axonal damage
Acute motor axonal neuropathy (AMAN)	Children and young adults; prevalent in China and Mexico; may be seasonal; recovery rapid; anti-GD1a antibodies	Axonal	First attack at motor nodes of Ranvier; macrophage activation, few lymphocytes, frequent periaxonal macrophages; extent of axonal damage highly variable
Acute motor sensory axonal neuropathy (AMSAN)	Mostly adults; uncommon; recovery slow, often incomplete; closely related to AMAN	Axonal	Same as AMAN, but also affects sensory nerves and roots; axonal damage usually severe
M. Fisher syndrome (MFS)	Adults and children; uncommon; ophthalmoplegia, ataxia, and areflexia; anti-GQ1b antibodies (90%)	Demyelinating	Few cases examined; resembles AIDP

Diagnostic Features of Acute Inflammatory Demyelinating Polyneuropathy (AIDP)

I. Required for Diagnosis

1. Progressive weakness of variable degree from mild paresis to complete paralysis
2. Generalized hypo- or areflexia

II. Supportive of Diagnosis

1. Clinical Features

a. Symptom progression: Motor weakness rapidly progresses initially but ceases by 4 weeks. Nadir attained by 2 weeks in 50%, 3 weeks 80%, and 90% by 4 weeks.

b. Demonstration of relative limb symmetry regarding paresis.

c. Mild to moderate sensory signs.

d. Frequent cranial nerve involvement: Facial (cranial nerve VII) 50% and typically bilateral but asymmetric; occasional involvement of cranial nerves XII, X, and occasionally III, IV, and VI as well as XI.

e. Recovery typically begins 2–4 weeks following plateau phase.

f. Autonomic dysfunction can include tachycardia, other arrhythmias, postural hypotension, hypertension and other vasomotor symptoms.

g. A preceding gastrointestinal illness (e.g., diarrhea) or upper respiratory tract infection is common.

Cerebrospinal Fluid Features Supporting Diagnosis

a. Elevated or serial elevation of CSF protein.

b. CSF cell counts are <10 mononuclear cell/mm3.

3. Electrodiagnostic Medicine Findings Supportive of Diagnosis

a. 80% of patients have evidence of NCV slowing/conduction block at some time during disease process.

b. Patchy reduction in NCV attaining values less than 60% of normal.

c. Distal motor latency increase may reach 3 times normal values.

d. F-waves indicate proximal NCV slowing.

e. About 15–20% of patients have normal NCV findings.

f. No abnormalities on nerve conduction studies may be seen for several weeks.

III. Findings Reducing Possibility of Diagnosis

1. Asymmetric weakness
2. Failure of bowel/bladder symptoms to resolve
3. Severe bowel/bladder dysfunction at initiation of disease
4. Greater than 50 mononuclear cells/mm3 in CSF
5. Well-demarcated sensory level

IV. Exclusionary Criteria

1. Diagnosis of other causes of acute neuromuscular weakness (e.g., myasthenia gravis, botulism, poliomyelitis, toxic neuropathy).
3. Abnormal CSF cytology suggesting carcinomatous invasion of the nerve roots

Thus GBS is a group of syndromes with several distinctive subtypes classified on a pathologic basis into demyelinating and axonal forms. Axonal GBS has been classified further into 2 groups: AMAN and acute motor and sensory axonal neuropathy (Gupta *et al.,* 2008). Although hyporeflexia or areflexia is the hallmark of GBS, normal reflexes or hyperreflexia is not a finding inconsistent with the diagnosis of GBS. The variants most commonly reported to be associated with retained or brisk reflexes are AMAN, acute motor conduction block neuropathy, and acute facial diplegia with brisk reflexes (Kuwabara *et al.,* 1999; Susuki *et al.,* 2004; Capasso *et al.,* 2003).

The incidence of hyperreflexia in AMAN is reported to be between 33% and 48% (Kuwabara *et al.,* 1999 and McKhann *et al.,* 1993).

CSF analysis shows albuminocytologic dissociation in most cases. Almost all of them have IgG anti-GM1 ganglioside antibodies although anti-*C. jejuni* antibodies are frequently negative (Kuwabara *et al.,* 1999). Antibody testing is not freely available in developing countries, such as India, which makes the diagnosis more difficult.

Although preservation of reflexes may simply be due to sparing of the sensory afferent pathway, the occurrence of brisk reflexes suggests a central mechanism. Dysfunction of inhibitory systems in the spinal interneurons has been proposed (Kuwabara *et al.,*1999). In these cases, distal conduction disturbance, not axonal degeneration, produces low motor responses on nerve conduction studies, termed as reversible conduction failure or acute motor conduction block neuropathy (McKhann *et al.,* 1993). The presumed mechanism producing reversible conduction block is impaired physiologic conduction at the nodes of Ranvier.

So uniqueness of this case is it is a demyelinating type of motor polyradiculoneuropathy with hyperreflexia unlike previously reported as either aman or variants.

The most common differential diagnosis presenting in a similar manner with acute progressive weakness and brisk reflexes is a high cervical myelopathy.

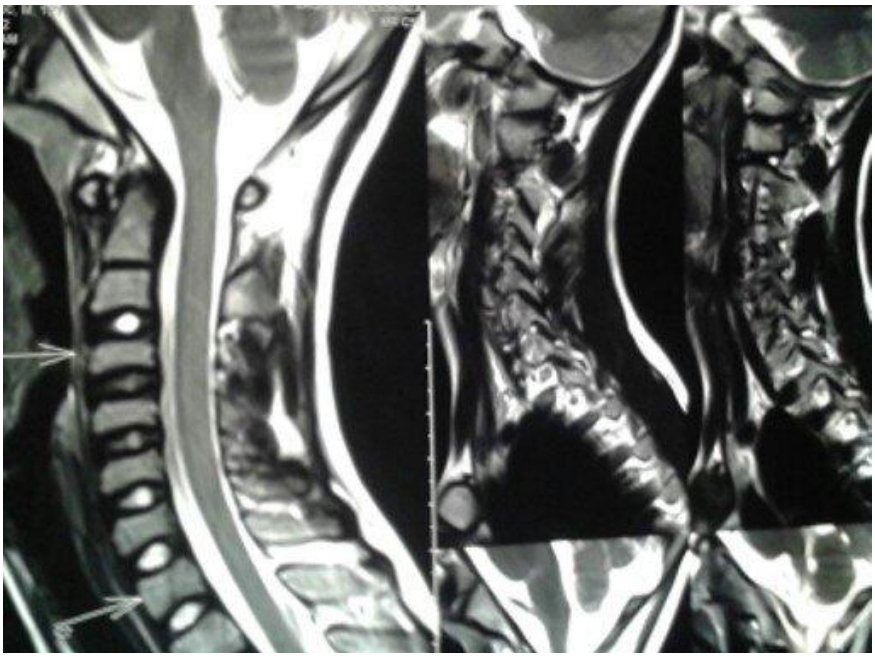

Figure 1: MRI cervical spine showing no feature of myelopathy

Therefore, GBS should be considered in patients with acute pure motor quadriparesis with normal or brisk reflexes even though as shown in the criteria for AIDP hypo or areflexia is a diagnostic requirement. This case report is to impress upon treating physicians and neurologists in training that a normo/hyperreflexic variant of GBS albeit rare, should not be missed in a given clinical setting.

CONCLUSION

Even though arflexia or hyporeflexia is expected as a requirement for diagnosis of GBS variants like this case with hyperreflexia is possible and must not be missed.

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