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Case Report

A Case of Guillan Barre Syndrome Presenting with Hyperreflexia

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ABSTRACT

Areflexia and progressive flaccid weakness are considered essential for diagnosis of Guillain-Barré syndrome (GBS) .We hereby report a case of a 41 year old male presenting with acute onset flaccid quadriparesis. Diagnosis of GBS wasmade based on history and clinical findings supported by Cerebrospinal fluid(CSF) studies and Nerve Conduction Study(NCS). The significance of the case was the presence of frank hyperreflexia in all four limbs. Although reflex preservation and hyperreflexia can be noted in axonal variant of GBS in Chinese, Japanese, and European populations, it is uncommon in India. This case report is to stress upon the fact that a hyperreflexic variant of GBS though rare, should be kept in mind in an appropriate clinical setting.

KEYWORDS: Guillain-Barré syndrome, Hyperreflexia.

INTRODUCTION

Guillain-Barré syndrome (GBS) is an acquired Acute Inflammatory Demyelinating Polyradiculoneuropathy(AIDP), which manifests as areflexic, flaccid paralysis with variable sensory disturbances, and elevated cerebrospinal fluid (CSF) protein without pleocytosis i.e. albuminocytological dissociation[1]. Reflex preservation and hyperreflexia in axonal GBS have been observed in some populations but rare in India[2]. A high index of suspicion is needed to diagnose this rare entity.

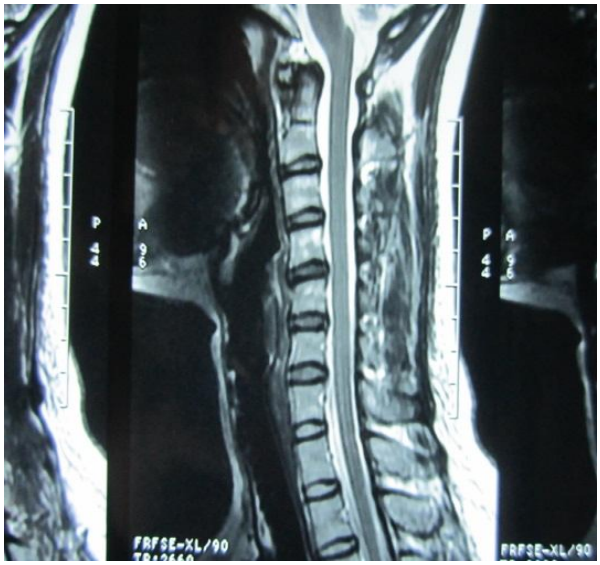
CASE REPORT

A 41 year old non-diabetic, non-alcoholic male presented with acute onset flaccid weakness in all four limbs for last 8 days. There was muscle pain involving deep muscles in the first 2 days of illness. No history suggestive of any sensory/bladder/bowel/autonomic/bulbar dysfunction. He had a history of suffering an episode of diarrhoea two months back lasting for 2 days which remitted spontaneously. Neurological examination revealed a normal higher mental function without any affected cranial nerve. All 4 limbs were flaccid, and power of the proximal muscles of the lower limb was 3/5 (Medical Research Council grading) and distal muscles was 2/5. Power in both the proximal and distal upper limb muscles, was 3/5. Deep tendon reflexes were brisk(3+) throughout the course of illness . Plantar response was nonresponsive bilaterally. All superficial reflexes were normally present.

Autonomic, sensory and cerebellar system was normal. Cardiovascular, respiratory and gastrointestinal examination was non-contributory. Investigations showed normal hematological parameters, serum biochemistry and electrolytes (Na, Ca, K, Mg, phosphate), and creatine kinase. Vasculitis work-up was negative. CSF examination was done on 10th day of illness which showed 3 cells (all lymphocytes); CSF sugar was 62 mg/dL (plasma glucose 132 mg/dL) and protein 108 mg/dL suggestive of albumino-cytological dissociation. Magnetic resonance imaging(MRI) of cervical spine revealed no features of any compressive myelopathy.

Nerve Conduction Study of all four limbs revealed decreased nerve conduction velocities in all 4 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 limbs were lost and in lower limbs showed impersistence and chronodispersion signifying demyelination of motor radicles as well. Diminished amplitude was noted in few motor nerves signifying an axonal involvement. Thus a diagnosis of acquired predominantly demyelinating (with axonal) type of motor poly radiculoneuropathy was made. He was treated with intravenous immunoglobulin (IVIg) (400 mg/kg/day) for 5 days. At discharge 3 weeks later, he had a significant improvement and upper limb power was 4/5 and lower limb power 3/5,both proximally and distally.

Figure 1: MRI Cervical spine showing no evidence of Compressive Myelopathy



DISCUSSION

GBS is broadly classified on a pathologic basis into demyelinating and axonal forms. Axonal GBS has been sub classified into: acute motor axonal neuropathy(AMAN) and acute motor and sensory axonal neuropathy(AMSAN)[1]. Although hyporeflexia or areflexia is a cardinal feature of GBS, preserved reflexes or hyperreflexia is not a finding inconsistent with GBS[2]. The subtypes most commonly associated with retained or brisk reflexes are AMAN, acute motor conduction block neuropathy, and acute facial diplegia with brisk reflexes[3]. Although preservation of reflexes may simply be due to sparing of the sensory afferent pathway, the occurrence of hyperreflexia usually denotes a central mechanism.

Dysfunction of inhibitory systems in the spinal inter neurons has also been proposed as a possible mechanism. In these cases, distal conduction disturbance instead of axonal degeneration, is the culprit here which produces low motor responses on nerve conduction studies, termed as reversible conduction failure or acute motor conduction block neuropathy. The possible mechanism producing reversible conduction block is impaired conduction at nodes of Ranvier[4]. Hyperreflexia seen in GBS is usually associated with antecedent *C. jejuni* infection with most patients having history of abdominal pain and diarrhoea. These cases are usually clinically mild and bulbar/ respiratory involvement is uncommon.

Our patient also had prior history of diarrhoea and no bulbar/ autonomic involvement. Almost majority of patients have IgG anti-GM1 ganglioside antibodies but no anti-*C. jejuni* antibodies[5].However antibody testing is not freely available in India, which makes proving this association difficult. The most common differential diagnosis presenting in this setting is a high cervical myelopathy.A proper history and examination, with NCS, CSF examination becomes essential. Therefore, GBS (specially axonal form) should be considered as a definite possibility in patients with acute pure motor quadriparesis even with normal or brisk reflexes, especially with prior history of gastroenteritis.

CONCLUSION

Although arflexia or hyporeflexia is expected in GBS, variants with hyperreflexia is possible and must not be missed in appropriate clinical settings.

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