

Guillain-Barre Syndrome Presenting with Hyperreflexia – A Rare Case Report.

N. S. Neki¹, Gural Singh Sachdeva², Chandandeep³, Husanpal³, Amandeep singh³, Harpreet Kaur³, Parminderpal Singh⁴

¹Professor, Department of Medicine, Govt. Medical College/ Rajindra Hospital, Patiala, 147001, Punjab, India

²Assistant Prof, Department of Medicine, Govt. Medical College/ Rajindra Hospital, Patiala, 147001, Punjab, India

³Junior Resident, Department of Medicine, Govt. Medical College/ Rajindra Hospital, Patiala, 147001, Punjab, India

⁴Senior Resident, Department of Medicine, Govt. Medical College/ Rajindra Hospital, Patiala, 147001, Punjab, India

ABSTRACT

Guillain-Barre syndrome (GBS) is an acquired autoimmune polyradiculopathy. Areflexia and progressive flaccid weakness are essential for its diagnosis. Here we report a case of 25 year old female presenting with acute onset flaccid quadriplegia. The diagnosis of GBS was made on history and clinical findings supported by cerebro-spinal fluid (CSF) analysis and nerve conduction studies (NCS). The hallmark of this case was the presence of hyperreflexia in all four limbs without involvement of higher functions, sensory, autonomic and bulbar dysfunction. To diagnose this rare entity, a high index of suspicion is needed.

Keywords: Guillain-Barre Syndrome; Hyperreflexia; Cerebrospinal fluid; Nerve conduction studies.

INTRODUCTION

GBS is an acquired acute inflammatory demyelinating polyradiculopathy (AIDP) characterized by areflexia, flaccid paralysis and albuminocytological dissociation reflected in CSF analysis.^[1] Reflex preservation and hyperreflexia can occur in the axonal variant of GBS in Japanese, Chinese and European populations, but it is rare in India.^[2] We report a case of polyradiculopathy with a reflexia with review of literature.

Name & Address of Corresponding Author

Dr. N. S. Neki,

Professor, Dept of Medicine,

Govt. Medical College/ Rajindra Hospital, Patiala, 147001, Punjab, India

E-mail: drneki123@gmail.com

CASE REPORT

A 25 year old female, non-diabetic and nonalcoholic, presented with acute onset flaccid weakness affecting all four limbs for the last five days. There was no history suggestive of sensory, bladder/ bowel, autonomic or bulbar dysfunction. Past history revealed a history of diarrhoea lasting for two days, which was not accompanied by fever, abdominal pain or respiratory infection. On neurological examination, higher mental functions and cranial nerves were normal with no neck rigidity. The Patient was almost bed ridden and could not move her limbs and lift head from the bed. Power in the upper limbs involving both proximal and distal group of muscles was 3/5 and in lower limbs, involving both proximal and distal group of muscles was 2/5. Sensory, autonomic & cerebellar examination was normal. All superficial reflexes were normal while plantar reflex was nonresponsive

bilaterally. However, deep tendon reflexes in the upper & lower limbs (biceps, triceps, supinator, knee & ankle) were exaggerated 3+ throughout the course of illness. Vitals were stable. Examination of the cardiovascular, respiratory and gastrointestinal systems was noncontributory. Laboratory profile, including hemogram, serum biochemistry, electrolytes and creatinine kilns along with vasculitis work-up were within the normal limits. CSF examination revealed protein 90 mg/dl, sugar 70 mg/dl (plasma glucose 120 mg/dl), cell count of 3/mm³ suggestive of albumin cytological dissociation. Magnetic resonance imaging (MRI) of the cervical spine showed no evidence of compressive myelopathy. Nerve conduction studies showed decreased nerve conduction velocities in all four limbs along with conduction block in the bilateral tibial nerve suggestive of acquired demyelinating type neuropathy while sensory testing was normal. F wave was lost in upper limbs while in lower limbs there was impression of impersistence and chrono-dispersion signifying demyelinating of motor radicles. Few motor nerves also revealed evidence of diminished amplitude suggestive of axonal involvement. Based on history, clinical examination and nerve conduction studies, a diagnosis of acquired predominantly demyelinating (with axonal) type of motor polyradiculoneuropathy was made. She was treated with intravenous immunoglobulin 400/kg/day for 5 days. She showed improvement and was discharged after 2 weeks with power in upper limbs 4/5 and lower limbs 3/5 both proximally and distally with evidence of ulcers on the back.

DISCUSSION

GBS is a group of syndromes, classified on the pathologic basis into demyelinating and axonal forms. Axonal GBS is further classified into 2

groups: acute motor axonal neuropathy (AMAN) and acute motor and sensory axonal neuropathy (AMSAN).^[1,3] The subtypes/ forms most commonly associated with preserved or brisk reflexes are AMAN, acute motor conduction block neuropathy and acute flaccid diplegia with brisk reflexes.^[4,5] Although hyporeflexia or areflexia is the hallmark of GBS, but preserved reflexes or hyperreflexia can rarely be found in GBS.^[2] The incidence of hyperreflexia in AMAN varies from 33% to 48% in various studies.^[6,7] Although preservation of reflexes may occur due to sparing of sensory afferent pathways, yet the central mechanism could explain the occurrence of hyperreflexia. Dysfunction of the inhibitory systems in the spinal inter neurons has been postulated.^[6] In these cases, distal conduction disturbance instead of axonal degeneration may cause low motor response on nerve conduction studies, labelled as reversible conduction failure or acute motor conduction block neuropathy.^[7] It has been purposed that reversible conduction block may occur due to impaired physiologic conduction at nodes of Ranvier.^[5] Preserved reflexes or hyperreflexia seen in GBS is usually associated with antecedent C. Jejuni infection in majority patients reporting a history of diarrhoea. These cases are usually clinically mild and bulbar, respiratory involvement is uncommon. Our patient had a past history of diarrhoea without involvement of bulbar and autonomic system. Almost all patients have IgG anti GM₁ ganglioside antibodies but no anti C jejuni antibodies.^[6] However, antibody testing is not freely available in India & this makes the diagnosis more difficult. CSF examination reveals an albuminocytological dissociation in most cases as documented in our case. The differential diagnosis includes high cervical myelopathy. So GBS (especially axonal form) must be thought of as a strongest possibility in patients presenting with acute pure motor quadriparesis even with normal or brisk reflexes particularly with a history of diarrhoea.

CONCLUSION

Although areflexia or hyporeflexia and progressive flaccid weakness are considered essential for the diagnosis of GBS, but the aim of reporting this case is to make the treating physicians & neurologists aware that normo/ hyperreflexic variant of GBS which is rare clinical entity, must be kept in mind in an appropriate clinical setting.

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