Hyperreflexic Guillain-Barré Syndrome: A Case Report

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ABSTRACT

Guillain-Barré syndrome (GBS) is an acquired acute autoimmune polyradiculoneuropathy. Progressive motor weakness and areflexia are considered two essential features to diagnose GBS. But sometimes in axonal variants of GBS, there have been several descriptions of hyperreflexia. Although not common in South Asia, there have been several reports of hyperreflexic GBS. A 12-year-old girl presented with progressive lower limb weakness and was diagnosed as a case of hyperreflexic GBS. GBS is a common disease in Bangladesh but there have been no reports of the hyperreflexic variety.

Keywords: Guillain-Barré syndrome; Autoimmune polyradiculoneuropathy; Hyperreflexia

Conflict of Interest: None declared

This article has been peer reviewed.

Article Submitted on: 30th March 2014

Article Accepted on: 20th July 2014

Funding Sources: None declared

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Cite this Article: Shafi AM, Rahman S. Hyperreflexic Gullian Barre syndrome: a case report. J Pioneer Med Sci. 2014; 5(1):13-14

INTRODUCTION

Guillain-Barré syndrome (GBS) presents with acute paralysis in 70% of patients, usually 1–4 weeks following a respiratory infection or diarrhea (particularly due to *Campylobacter*) [1]. Patho-physiologically, cell-mediated inflammatory response is directed at the myelin sheath of spinal roots, peripheral and extra-axial cranial nerves, triggered by molecular mimicry between epitopes found in the cell walls of some microorganisms and gangliosides in the Schwann cell and axonal membranes [1]. The two features essential for the diagnosis of GBS are progressive motor weakness and areflexia. There are two distinctive pathologic subtypes of GBS: demyelinating and axonal. Recently, there have been several reports of reflex preservation and hyperreflexia in axonal GBS in Chinese, Japanese, and European populations [2, 3].

CASE REPORT

A 12-year-old girl was admitted with sudden onset of progressive weakness of lower limbs for 10 days. She complained of difficulty in getting up from squatting position and climbing stairs. There were no cranial nerve symptoms, sensory symptoms or bladder symptoms. There was no muscle pain. There was no preceding fever, diarrhea, vaccination or drug intake. On examination, she was afebrile, with flaccid lower limbs, weakness of proximal muscles of lower limb (1/5, Medical Research Council grading) and examination of upper limb was normal. Deep tendon reflexes of lower limb were brisk. Plantar reflexes were flexor bilaterally. Sensory system was normal. There were no abnormal cerebellar signs. Her cognitive function was intact. She needed one-person support to walk. Laboratory investigations showed normal total and differential leukocyte counts, erythrocyte sedimentation rate, creatine kinase and electrolytes. Vasculitis work-up was negative. Cerebrospinal fluid examination showed increased protein with no cells. Magnetic resonance imaging of the brain and spine was normal and thus excluded compressive myelopathy. Nerve conduction study was suggestive of pure motor axonopathic variant of GBS (acute motor axonal neuropathy). She was prescribed methylprednisolone with improvement in her symptoms. We could not perform immunological tests such as anti GM1 and anti GQ1b antibodies as these tests were not available locally.

DISCUSSION

GBS may be described as a collection of clinical syndromes that manifests as an acute inflammatory polyradiculoneuropathy with weakness and diminished reflexes. Acute inflammatory demyelinating polyradiculopathy is the most widely recognized form of GBS in Western countries, but the variants known as acute motor axonal neuropathy and acute motor-sensory axonal neuropathy also are well recogni-

-zed. On a pathologic basis GBS is divided into demyelinating and axonal forms. Axonal GBS is further classified into 2 groups: acute motor axonal neuropathy and acute motor and sensory axonal neuropathy [5]. The variants most commonly associated with retained or brisk reflexes are acute motor axonal neuropathy, acute motor conduction block neuropathy, and acute facial diplegia with brisk reflexes [5, 6]. Although in general, acute motor axonal neuropathy has been associated with extensive axonal loss and poor outcome, this subgroup with reversible conduction failure recovers rapidly [2-4].

Hyperreflexia seen in GBS has a common association with antecedent *C. jejuni* infection. The occurrence of brisk reflexes suggests a central mechanism although dysfunction of inhibitory signals in the spinal interneurons has also been proposed [4]. The severities of these GBS cases are usually mild and patients very rarely develop respiratory depression. In most cases, albuminocytologic dissociation is seen on cerebrospinal fluid examination. Almost all patients have IgG anti-GM1 ganglioside antibodies although anti-C. Jejuni antibodies are frequently negative [2]. Antibody testing is not freely available in developing countries, which makes the diagnosis challenging.

CONCLUSION

We suggest that GBS (axonal form) should not be excluded in patients with acute pure motor paraparesis or quadriparesis with normal or brisk reflexes, especially while there is a past history of antecedent gastroenteritis.

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