CASE REPORTS

ACUTE MOTOR AXONAL NEUROPATHY VARIANT OF GUILLAIN BARRE SYNDROME PRESENTING WITH BRISK KNEE JERKS

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ABSTRACT

The cardinal clinical presentation of Guillain Barre Syndrome (GBS) is acute motor weakness accompanied by absent or depressed deep tendon reflexes (DTRs). DTRs may be occasionally preserved in acute motor axonal neuropathy (AMAN) variant of GBS. We report the case of a 40 year old male suffering from GBS who presented with brisk knee jerks at the time of presentation. He was treated with plasmapheresis to which he responded well. His Electromyography and Nerve Conduction Studies (EMG/NCS) confirmed AMAN.

Keywords: Guillain-barre syndrome (GBS), Acute motor axonal neuropathy (AMAN), Electromyography and nerve conduction studies (EMG/NCS).

INTRODUCTION

Guillain-Barre syndrome (GBS) is an acute immune mediated polyneuropathy presenting as a rapidly progressive symmetric motor paralysis accompanied by absent or depressed deep tendon reflexes (DTRs). The syndrome has been classified into demyelinating and axonal forms on the basis of electromyography and nerve conduction studies (EMG/NCS). The most common variant is acute inflammatory demyelinating polyneuropathy (AIDP). The axonal variants include acute motor axonal neuropathy (AMAN) and acute motor sensory axonal neuropathy (AMSAN)[1].

The most important investigations in GBS are EMG/NCS and cerebrospinal fluid (CSF) examination. CSF characteristically contains a high protein concentration with normal cell content. EMG/NCS abnormalities may be minimal and CSF may be normal in the first week. If the diagnosis is strongly suspected, treatment should be initiated without waiting for characteristic EMG/NCS and CSF findings.

GBS is the most frequent, rapidly evolving and potentially fatal acute generalized poly-neuropathy. Patients who are able to walk without support are observed closely and monitored for progression of weakness. If the patient becomes unable to walk unaided, has reduction in vital capacity or shows signs of oropharyngeal weakness then plasma exchange or intravenous immunoglobulins (IVIG) should be instituted without delay[2].

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CASE REPORT

A 40 years old male patient was admitted to Combined Military Hospital (CMH) Lahore on 28 Jan 2015 with 2 days history of progressive weakness. He had no history of fever, headache, pain in neck or loss of control of sphincters. His vital signs were stable and general physical examination revealed no abnormality. His neurologic examination revealed power graded as 3 ± 5 in all four limbs. His knee jerks were brisk (+++) bilaterally but rest of his DTRs were normal with bilateral flexor plantar response. There was no sensory deficit. Rest of his neurological examination was unremarkable. A provisional diagnosis of GBS was made and investigations including hematologic values, blood chemistry, EMG/NCS and magnetic resonance imaging (MRI) of cervical spine were ordered.

He was kept under close observation and was started subcutaneous low molecular weight

heparin (Clexane) in addition to physiotherapy. His weakness progressed and on the 5th day of admission his power reduced to 2/5 and all DTRs were lost. His investigations, except for EMG/NCS which was pending, revealed no abnormality. His plasmapheresis was started on alternate days. His 5th session of plasmapheresis had to be cancelled and dual lumen central catheter removed when he developed high grade fever with rigors. He was treated with parenteral antibiotics to which he responded well. His EMG/NCS on 15th day of admission suggested Acute motor axonal neuropathy (AMAN) variant of GBS. On the 18th day of admission his power was 2/5 in the upper limbs and 3/5 in the lower limbs and then 10 days later it further improved to 3/5 in upper limbs and 4/5 in lower limbs. At the end of 5th week of admission the patient was walking without support. He was discharged on 16 Mar 2015 (7th week of admission).

DISCUSSION

AMAN was first recognized in 1986, mainly occurring in China and Japan. Most cases are preceded by Campylobacter jejunienteritis. The clinical presentation of AMAN is similar to AIDP except that DTRs are occasionally preserved in AMAN.

AMAN has been strongly associated with antibodies to gangliosides GM1 and GD1a in axons of peripheral nerves[3]. These anti-ganglioside antibodies may be induced by campylobacter jejuni and other infections and may mediate axonal damage. The frequency of AMAN is not known in Pakistan. However, George et al from South India have reported that 14% of their GBS cases were AMAN variant. One of their patients had hyperreflexia while another had hyporeflexia with extensor plantar response[4]. Although hyporeflexia or areflexia is necessary for diagnosis of GBS, hyperreflexia does not exclude a GBS variant. DTRs may be preserved throughout the disease course in AMAN and have been considered an indicator of rapid clinical recovery[5].

This case report emphasizes the importance of considering a GBS variant in patients presenting with acute progressive motor weakness with preserved or hyperactive DTRs. Early institution of plasma exchange or IVIG can prevent further progression of the disease resulting in good prognosis.

CONFLICT OF INTEREST

This study has no conflict of interest to declare by any author.

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