CASE REPORTS

PHARYNGEAL CERVICAL BRACHIAL VARIANT OF GUILLAIN BARRE SYNDROME REQUIRING PROLONGED VENTILATORY SUPPORT

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ABSTRACT

Patients with pharyngeal cervical brachial (PCB) variant of Guillain Barre Syndrome (GBS) typically present with rapid progression of oropharyngeal and cervicobrachial weakness. We report case of a 25 year old male soldier suffering from PCB who presented with brachiopharyngeal weakness which progressed to involve his lower limbs. He was intubated and mechanically ventilated and was treated with plasmapheresis. He made good recovery after remaining on ventilatory support for a long time. His electromyography and nerve conduction studies (EMG/NCS) were consistent with axonal polyneuropathy.

Keywords: Electromyography and Nerve Conduction Studies (EMG/NCS), Guillain Barre Syndrome (GBS), Pharyngeal cervical brachial (PCB).

INTRODUCTION

Guillain Barre Syndrome (GBS) is an immune mediated polyneuropathy presenting with rapidly progressive generalized motor weakness. In a typical case the paralysis ascends from legs to arms and then to cranial nerves. In some patients GBS may appear in abortive forms (table)[1]. Pharyngeal cervical brachial (PCB)variant of GBS represents a focal form of acute motor axonal neuropathy (AMAN).

In PCB, pharyngeal, cervical and brachial muscles may be affected first or constitute the entire illness. Incomplete forms of PCB may also occur. Recently, Kim et al have described another regional variant of GBS i.e. acute bulbar palsy (ABP) with no limb weakness, differentiating it from the Miller Fisher syndrome (MFS) and PCB[2].

Diagnosis of PCB can be made on the basis of history and neurologic examination. The differential diagnosis includes myasthenia gravis, brainstem lesions, diphtheria and botulism. Magnetic resonance imaging (MRI) of brain may exclude brainstem lesions. Cerebrospinal fluid (CSF) albuminocytological dissociation, electromyography and nerve conduction studies (EMG/NCS) and anti-GT1a antibodies support the diagnosis of PCB. In early disease anormal CSF or EMG/NCS do not exclude PCB. PCB is treated with plasma exchange or intravenous immunoglobulins (IVIG).

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CASE REPORT

A 25 year old male soldier was transferred to Combined Military Hospital (CMH) Lahore, from CMH Sialkot on 10 Dec 2015 with 2 days history of progressive weakness of upper limbs followed by pharyngeal weakness. His power in upper limbs was graded as 3/5 and lower limbs 5/5 with bilateral flexor plantar response. He was diagnosed as a case of GBS variant and shifted to Intensive Care Unit (ICU), intubated and mechanically ventilated. His routine investigations including hematologic values, blood chemistry and cerebrospinal fluid examination were normal. He was started plasmapheresis on alternate days but his weakness continued to increase. On the 9th day of admission tracheostomy was done. At the end of last session (6th) of plasmapheresis, his power in upper limbs was 0/5 and lower limbs 1/5 and he had right facial palsy. Then his power started improving. On 20th day of admission his power was 1/5 in upper limbs and 2/5 in lower limbs.

During this period he developed repeated chest and tracheostomy site infections which were treated with antibiotics. Weaning off trials from ventilator were started and eventually he was off ventilator completely after more than 5½ months (170 days) on 29 May 2016. Physiotherapy was continued and he started walking with support. His EMG/NCS on 16 June 2016 revealed chronic axonal motor neuropathy involving all limbs; upper > lower. He was transferred back to CMH Sialkot on 16 July 2016 when he was fully ambulatory, having only mild weakness of upper limbs (power 4+/5).

Table: Regional Variants of GBS.

1.	Pharyngealcervicalbrachial weakness.
2.	Oculopharyngeal weakness.
3.	Ophthalmoplegia with GQ1b autoantibodies.
4.	Fisher syndrome (Ophthalmoplegia, ataxia, and areflexia.)
5.	Bilateral facial or abducens weakness with distal paresthesias.
6.	Predominantlyparaparesis.

DISCUSSION

PCB, defined as a rapidly progressive cervicobrachial and oropharyngeal weakness associated with areflexia in upper limbs, was first described by Ropper in 1986[3]. There was serological evidence of campylobacter jejuni infection in 31% of patients.

Some PCB patients may have additional features of ophthalmoplegia and ataxia which are characteristic of MFS and Bickerstaff Brainstem Encephalitis (BBE). Nagashima et al defined 'pure' PCB as rapidly progressive oropharyngeal and cervicobrachial weakness associated with hyporeflexia or areflexia in the absence of leg weakness or ophthalmoplegia. Patients with PCB having ophthalmoplegia and ataxia were defined as having PCB overlap with MFS; patients having PCB with ataxia ophthalmoplegia and altered consciousness as having PCB overlap with BBE; and PCB with leg weakness as having PCB overlap with GBS[4]. However, as PCB represents a focal form of AMAN, PCB overlap with GBS may be considered as a fulminant form of PCB[5].

GBS variants may be associated with autoantibodies against specific neuronal gangliosides i.e. AMAN with antibodies to GM1 and GD1a; MFS and BBE with antibodies to GQ1b and PCB with antibodies to GT1a. In a study of 100 patients suffering from PCB, 50% had anti-GT1a antibodies and 25% had antibodies against GM1 or GD1a[4].

The diagnosis of PCB may be very challenging in early disease. This case report emphasizes the importance of early diagnosis of PCB and assessment of bulbar functions and respiratory effort to pass a nasogastric tube for feeding and to provide ventilatory support without delay. Early institution of plasma exchange or IVIG can result in a better outcome.

CONFLICT OF INTEREST

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

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