CASE REPORT

An Acute Pharyngeal-Cervical-Brachial (PCB) Variant of Guillain-Barre Syndrome Presenting with Isolated Bulbar Palsy

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Summary

Acute Guillain-Barre syndrome (GBS) is characterized by an acute onset of limb weakness and areflexia. There are a few rare variants that have been described and one of them is the pharyngeal-cervical-brachial (PCB) variant (oropharynx, neck, and proximal upper limb muscles). However, in this patient, the only presentation was bulbar involvement with fast recovery within days. This is likely to be the milder form of PCB that has rarely been described before. A 19-year-old Malay lady presented with progressive dysphagia associated with nasal voice for one week duration. There was no limb weakness. Examination showed generalized areflexia. Pharyngeal and palatal muscles were markedly weak. Cerebrospinal fluid (CSF) examination showed raised protein level. Nerve conduction studies revealed generalized demyelinating motor polyneuropathy consistent with GBS. The patient fully recovered within three days and was discharged well.

Key Words: Guillain-Barre syndrome, Pharyngeal-cervical-brachial variant

Introduction

The pharyngeal-cervical-brachial variant of GBS is a rare condition and only a few cases have been reported[1]. This variant does not cause generalized limb weakness as in the acute inflammatory demyelinating polyradiculoneuropathy (AIDP) type, which is the commoner form of GBS. It usually affects the oropharyngeal muscles especially the bulbar muscles, neck and proximal upper limb muscles. Raised protein CSF and demyelinating features on nerve conduction studies are the other features to support the diagnosis. Several reports have shown that the PCB variant is associated with the anti-GT1a IgG antibody[2]. This probably explains the immune mechanism that is involved in the pathogenesis of this disease

Case Report

A 19-year-old lady developed a 1-week history of progressive dysphagia mainly to fluids. She also noticed that her voice had slowly become nasal. She did not experience any limb weakness, unsteadiness, facial asymmetry or any antecedent infection. The patient's previous medical history was unremarkable.

On admission to our institution, a neurological examination showed marked bulbar palsy involving the pharyngeal and palatal muscles. There was no facial weakness and neck flexion was strong. Limb muscle power was generally normal; grade 5/5. Deep tendon reflexes were generally absent, and plantar responses were down-going bilaterally. There was no sensory deficit. Her blood pressure was 130/80 mmHg, and pulse rate 88/minute (regular). She was afebrile and

This article was accepted: 27 August 2005

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not in respiratory distress. Examination of the lungs, abdomen and heart was normal.

Her serum potassium was 4.3 mmol/l, sodium 138 mmol/l, urea 4.0 mmol/l, random glucose 6.5 mmol/l and creatinine 67 umol/l. The haemoglobin was 11.2g%, white cell count $8.3x10^9$/l and platelet count $326x10^9$/l. The liver function test was normal. The chest radiograph was also normal. Cerebrospinal fluid (CSF) examination revealed no white or red blood cells. The protein level was high at 673 mg/l and glucose content normal (3.9 mmol/l; blood glucose 5.1 mmol/l). An urgent brain magnetic resonance imaging (MRI) was normal. Nerve conduction study was done on the third day of admission. The nerve conduction studies involve measurement of motor conduction velocities (MCV), distal motor latencies (DML) and f-wave latencies from the median, ulnar, common peroneal, and posterior tibial nerves. Compound motor action potentials (CMAP) are recorded with surface electrodes following supramaximal stimulation of the nerve at two sites. Sensory nerve action potentials (SNAP) are recorded from median, ulnar and sural nerves. Needle examination was not performed in this patient. The DML of all motor nerves examined were prolonged (Table I). The CMAP and the f waves of all the nerves studied were normal. The sensory studies were also normal. This is suggestive of demyelinating motor polyneuropathy consistent with Guillain-Barre syndrome. A bedside simple spirometry was also done and normal.

A diagnosis of acute GBS was made based on clinical, CSF and electrophysiological findings. She failed her swallowing test (coughing reflex after swallowing fluids) and a nasogastric tube was inserted. The patient started to show marked improvement while in the ward and the nasogastric tube was removed on day 3 of admission. By day 5, she was able to swallow fluids and solids without any problems and had almost returned to normal. Her reflexes also had returned to normal and she was discharged the next day. No treatment was instituted.

Discussion

The pharyngeal-cervical-brachial (PCB) variant of GBS was first reported by Roper *et al.* in 1986[1]. The illness ended without affecting the power of the legs. He called this condition PCB and considered it a regional variant of GBS. The above case presented with only bulbar muscle involvement and improved markedly within a week. This should be regarded as having a milder manifestation of the same immunopathogenetic process as in PCB. Usually PCB presents as severe bulbar palsy, and slowly progresses to facial palsy, ophthalmoplegia, and weakness of the neck flexors and proximal muscles of the upper limbs[3]. The other criteria for diagnosis of PCB variant are similar to acute GBS and include areflexia, raised CSF protein, and demyelinating disease on nerve conduction studies. Although the patient did not have any prior symptoms to suggest earlier infection, this has been described in many cases of GBS and is not a major criterion in diagnosing GBS. As bulbar palsy can be a main symptom in many neurological disorders, it is important to determine that other differential diagnoses have been excluded. Lesions affecting the brainstem may present with isolated bulbar palsy. However, MRI of the brain was done and showed no abnormality. Lesions involving the motor neurons and neuromuscular junction do not behave like the patient's presentation and therefore are unlikely. The detection

Table I: The motor nerves showing generalized prolonged DMLs

	DML	Normal range
Right median nerve	4.35ms	<3.5ms
Left median nerve	4.26ms	<3.5ms
Right ulnar nerve	3.06ms	<3.0ms
Left ulnar nerve	3.63ms	<3.0ms
Right common peroneal nerve	6.63ms	<6.5ms
Left common peroneal nerve	6.66ms	<6.5ms
Right posterior tibial nerve	8.34ms	<5.8ms
Left posterior tibial nerve	8.67ms	<5.8 ms

of anti-ganglioside antibodies has also been shown to help in the diagnosis of the PCB variant. Serum anti-GT1a IgG antibody has been detected in GBS with bulbar palsy but in most cases, the bulbar palsy was accompanied by other muscle involvement. So far, there has been only one case report showing isolated acute bulbar palsy due to the PCB variant[2]. The case was associated with anti GT1a IgG antibody but the nerve conduction studies were normal. The antibody test is not readily available in the country and unfortunately, was not done in this patient. As the patient improved markedly after admission, no pharmacological treatment was given. Furthermore, there was no limb weakness and the respiratory muscles were not compromised. Nasogastric tube was inserted as he regurgitated during the swallow test. It is important not to give oral feeding during this period as this may cause aspiration pneumonia. The swallow test was performed periodically to assess the progress of the dysphagia. Fortunately she recovered early and the tube was removed. For the severe form of the PCB variant, an alternative option is insertion of a percutaneous endoscopic gastrostomy (PEG) tube.

Acknowledgements

We would like to thank the Dean of the Medical Faculty Universiti Kebangsaan Malaysia (UKM) for allowing us to publish this paper.

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