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An Acute Pharyngeal-Cervical-Brachial (PCB) variant of Guillain-Barre Syndrome

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Abstract

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 [involving oropharynx, neck and proximal upper limb muscles]. Many neurologists and physicians are unaware of this variant which is often misdiagnosed as myasthenia gravis, brainstem stroke or botulism. A 37 year old female patient presented with the classical characteristics including bulbar weakness, cervicobrachial weakness and are flexia in the upper limbs, with relatively preserved lower limbs. Her systemic review was unremarkable. Her physical examination was compatible with GBS. Nerve conduction studies revealed Motor Axonal Radiculoneuropathy. She received 5 day course of IVIG and showed significant improvement over next 1 week. The patient recovered well and was discharged after 12 days.

Keywords: 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'. In contrast to the ascending pattern of weakness seen in the acute inflammatory demyelinating polyradiculoneuropathy (AIDP) type, which is the commoner form of GBS, it usually affects the oropharyngeal muscles especially the bulbar, neck and proximal upper limb muscles. CSF shows raised proteins and nerve conduction study shows axonal changes and not demyelinating as is usually seen in typical GBS. There are several reports showing that the PCB variant is associated with the anti-GTla IgG antibody'. This probably explains the immune mechanism that is involved in the pathogenesis of this disease.

Case Report

A 37—year—old lady presented with a 2—day history of progressive weakness of her neck, on awakening on first day, with complaints of difficulty holding up her head straight. Next day, patient had difficulty standing up straight with complaints of difficulty in supporting her balance and as her truncal weakness progressed, she was not able to perform any activity. She also noticed weakness in her arms with difficulty in raising her arms above shoulder. She noticed that her voice had nasal intonation. She did not experience any lower limb weakness, facial asymmetry, double vision, fatigability or any antecedent infection. The patient's previous medical history was unremarkable.

On admission, neurological examination showed marked neck muscle weakness involving both the neck flexors and extensors. There was no facial weakness, extraocular movements were full with no diplopia and no fatigability. She had bulbar weakness with weak gag and cough, along with reduced palatal movement.

Her single breath count was noticed to be 24. Tone was reduced in upper limbs with proximal muscle power of MRC grade 2/5. Lower Limb muscle power was generally normal; grade 5/5. Deep tendon reflexes were absent in upper limbs, however were preserved in lower limbs[+2], and plantar responses were down- going bilaterally. There was no sensory deficit. Her blood pressure was 13/80mmHg, and pulse rate of 80/min, regular. Examination of lungs, heart and abdomen was normal.

Subsequent examination reveals nasal twang, pooling of secretions and decline in single breath count to 20. As patient failed swallowing test, a nasogastric tube was inserted. Her RNS study from facial, trapezius and ABP muscles was within normal limits. MRI Brain and spine were essentially negative. Laboratory investigations including CBC, Renal and Liver profile, TSH, CRP were within normal limits. Anti-Ach-R, and anti-Ganglioside Ab Panel (Anti-T1a not included) were negative. Cerebrospinal fluid (CSF) examination done within 1st week revealed TLC of 2 cells, protein of 34.4mg/dl and glucose of 71.1mg/dl. Nerve conduction study shows findings consistent with motor axonal polyneuropathy. A presumptive diagnosis of PCB variant of GBS was made and the patient received 5 day course of IVIG. She showed significant improvement in her weakness over the next 5 days. Her single breath count improved to 40, power in nec flexor and extensor improved to 4/5, power in upper limb also improved to 4/5. She was able to support her balance as truncal weakness also improved. After rehabilitation she was discharged (12 days later) on supportive medications.

Discussion

Guillain Barre Syndrome is characterized by acute onset flaccid are flexic ascending paralysis with or without cranial nerves involvement (mainly seventh nerve).[1] There are several less common variants of GBS that have unusual patterns of muscle and cranial nerve involvement. The Pharyngio-Cervical- Brachial variant of GBS is rare (0.07-0.25/ 100,000)[2] and is characterized by oropharynx, neck and proximal upper limb weakness.[3,4] Preceding infection with cytomegalovirus or Campylobacter jejuni is often found[1] with anti-GT1a antibodies being the hallmark of this variant.[1,5] Other diagnostic criteria for PCB variant include diaphragmatic weakness, relative sparing of lower limbs power and deep tendon reflexes, minimal or no sensory deficit, raised CSF protein and axonal neuropathy.[4,7]

Our case illustrates many of the classic characteristics of PCB variant including bulbar weakness, areflexia, neck and upper extremity weakness.an unusually significant truncal weakness was seen at the time of presentation. We wish to raise awareness of this rare variant of GBS so that neurologists and physicians consider this in patients with bulbar, neck and upper limb weakness (resembling descending paralysis) as diagnosis and early intervention greatly benefit the patient's eventual outcome. Although the patient did nit had any prior or antecedent infection or vaccination, this has not been described in many cases of GBS abd is not a major criterion in diagnosing GBS. In addition as patient presented with rapid progression of symptoms, hence CSF studies did not revealed the classical albumin-cytological dissociation, otherwise seen after 2 weeks.

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