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**Case Report**

# When Guillain-Barre syndrome took a wrong path

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## ABSTRACT

The pharyngeal cervical brachial (PCB) variant of Guillain-Barre-syndrome (GBS) is defined by rapidly progressive oropharyngeal and cervicobrachial weakness associated with areflexia in upper limbs. On nerve conduction study PCB represents a localized subtype of GBS characterized by axonal rather than demyelinating neuropathy. PCB is often misdiagnosed as brainstem stroke, myasthenia gravis or botulism. Herein we describe a case of symmetric descending type of acute flaccid paralysis with predominant upper limb involvement and also lower limb involvement diagnosed as a PCB- GBS overlap mainly based on nerve conduction study.

**Keywords:** Pharyngeal cervical brachial variant, Axonal neuropathy, Symmetric descending paralysis

## INTRODUCTION

Guillain barre syndrome (GBS) is an acute onset monophasic inflammatory polyneuropathy characterized by rapidly progressive, symmetric, ascending weakness with areflexia.[1] Annual incidence of GBS is 0.3-1.3 per 1,00,000 persons less than 18 years of age.[1] GBS is often triggered by an antecedent infection with most common pathogen being campylobacter jejuni. Weakness is often symmetrical starting distally in lower limbs and progressively involves upper limbs, respiratory muscles and cranial nerves.[2] There are several variants of GBS diagnosed based on the clinical features and the nerve conduction studies.[3] PCB is one of rare variant of GBS typically presents with rapidly progressive oropharyngeal, neck and upper extremity muscles weakness and the first case was reported by Ropper in 1986.[4-5] Power in the lower limbs is usually preserved or only mildly affected indicating that PCB represents a localized subtype of GBS .Nerve conduction study in PCB variant shows a axonal neuropathy rather than a demyelinating type. Patients with PCB variant carry immunoglobulin G anti-GT1a antibodies which often cross-reacts with GQ1b antibodies. Patients with PCB are initially misdiagnosed as having brainstem stroke, myasthenia gravis/ botulism (Table 1).

**Table 1: Features differentiating between PCB variant and botulism.**

| Variables | PCB variant of GBS | Botulism |
|---|---|---|
| **Onset** | Hours | Hours |
| **Progression** | Gradual | Gradual /descending |
| **Pattern of weakness** | | |
| Ptosis | +/- | ++ |
| Internal ophthalmoplegia | +/- | ++ |
| External ophthalmoplegia | +/- | ++ |
| Facial | +/- | ++ |
| Bulbar | ++ | ++ |
| Arm | ++ | ++ |
| Leg | - | ++ |
| Symmetrical | Yes | Yes |
| Hyporeflexia/ areflexia | + | - |
| Sensory deficit | +/- | - |
| Other clinical features | Antecedent infectious symptoms | Dry mouth, dizziness and GI symptoms |

## CASE REPORT

A 14-year-old child, 1st born to a non-consanguineously married couple was brought with complaints of 4-5 episodes of vomiting 5 days prior to admission. Associated with complaints of pain in both the upper limbs the succeeding day which was insidious in onset progressive dull aching type, followed by weakness of both upper limbs which was more distal type of weakness when compared to proximal. The following day child noticed pain in both lower limbs and weakness of both the lower limbs. Weakness was more in upper limbs when compared to lower limbs. There was no history of fever, convulsions, altered sensorium, bowel and bladder involvement, dog bite or snake bite. There was no significant past history, family history and birth history. Child was immunized appropriate for age.

On examination child was vitally stable, head to toe examination revealed old healed scars of varicella over chest and abdomen. Single breath count was 13, there was poor cough and his voice quality was also poor. Other signs of impending respiratory failure like cyanosis, and pooling of secretions was absent. On systemic examination of central nervous system, higher mental functions was within normal limits, there was no cranial nerve involvement. Motor system examination of neck muscles revealed power of 2/5. Upper limb showed a power of 2/5 in proximal muscles and power of 1/5 in distal muscles. In lower limbs power was 2/5 except in plantar flexors which was 3/5. Deep tendon reflexes like biceps, triceps, ankle and knee jerk were absent. Superficial reflexes were within normal limits. Sensory system examination was within normal limits and there were no signs of autonomic nervous system involvement. Child's power belonged to Hughes grade 4 and according to egress scoring child belonged to high-risk category.

On investigating CBC and electrolytes were within normal limits. Nerve conduction study was suggestive of demyelinating motor polyradiculoneuropathy in all 4 limbs. Motor nerve conduction velocity of both median nerves showed prolonged distal latencies, reduced compound muscle action potential. Ulnar nerve study showed reduced compound muscle action potential and conduction block. Peroneal and tibial showed normal distal latencies, normal compound muscle action potential and normal conduction velocities. Cerebrospinal fluid analysis was planned but as there was no consent from attenders CSF analysis was not done. Hence diagnosis of PCB-GBS overlap was made based on the nerve conduction studies and child received IVIG at 2 gm/kg over 2 days. Daily child was monitored and was discharged after 10 days with physiotherapy. Power had improved from 2/5 to 4/5 over a 1 month follow up.

## DISCUSSION

PCB variant is one of the rare variants of guillian barre syndrome. Involves mainly bulbar, neck and upper limb muscles (Figure 1). These patients typically presents with rapidly progressive oropharyngeal and cervicobrachial weakness. Lower limb involvement in this variant is less. These patients must present in the absence of ataxia and disturbed consciousness[6].Most common initial symptom is arm weakness and second most common is dysphagia.[7] Also these patients are more prone for diaphragmatic paralysis and requirement of mechanical ventilator when compared to other variants. Antibodies seen in this variant is mainly GT1a, GQ1b, and GD1a. Nerve conduction studies shows axonal polyneuropathy.[7] Although nerve conduction studies show axonal variant, in our case nerve conduction study was suggestive of demyelinating variant. But other diagnostic features required for diagnosis of PCB variant was fulfilled. Also, there are other studies of PCB variant where nerve conduction showed demyelinating features.[6,8,9]

**Table 2: Diagnostic criteria for PCB variant of GBS.**

| Features required for diagnosis | Features strongly supportive of the diagnosis |
|---|---|
| **Relative symmetric oropharyngeal weakness and neck and arm weakness** | Antecedent infectious symptoms |
| **Absence of ataxia and disturbed consciousness and prominent leg weakness** | Cerebrospinal fluid-albumino-cytological dissociation |
| **Monophasic illness pattern and interval between onset and nadir of oropharyngeal or arm weakness between 12 hours and 28 days and subsequent clinical plateau** | Neurophysiological evidence of neuropathy |
| **Absence of alternative diagnosis** | Presence of IgG anti-GT1a or anti-GQ1b antibodies |

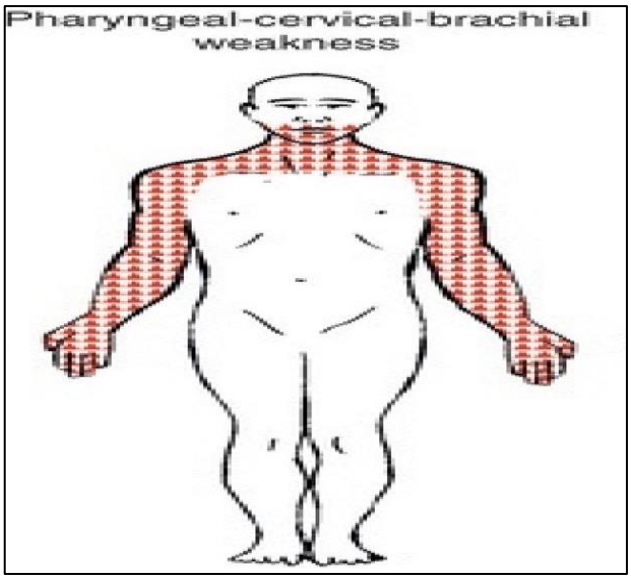


**Figure 1: PCB weakness.**

## CONCLUSION

PCB is a rare unfamiliar variant of GBS that is often misdiagnosed. We have reported a unique case of PCB-GBS overlap. Early clinical recognition of PCB variant is essential for preventing respiratory associated morbidity and mortality.

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