

## Acute Bibrachial Palsy: Another Forme Fruste of Guillain-Barré Syndrome

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### Abstract

An acute onset of restricted brachial diparesis in a healthy individual is a rare clinical scenario, which requires prompt assessment through a wide differential diagnosis. In this paper, we report the interesting case of a young patient who developed acute bibrachial paresis, in the context of an atypical manifestation of Guillain-Barré syndrome (GBS). In this case, bibrachial paresis represented an incomplete form of Pharyngeal-Cervical-Brachial (PCB) variant, a well-defined clinical subtype of GBS, where the disturbance of axonal function is supposed to be the main underlying mechanism.

**Keywords:** *Pharyngeal-Cervical-Brachial (PCB) Variant; Guillain-Barré syndrome (GBS); Bibrachial Paralysis; Acute Motor Axonal Neuropathy (AMAN)*

### Introduction

Pharyngeal-Cervical-Brachial (PCB) variant is a well-defined clinical subtype of Guillain-Barre syndrome (GBS). It is characterized by rapidly progressive weakness affecting oropharyngeal, neck and brachial muscles in conjunction with upper limbs areflexia [1]. In this paper, we report the interesting case of a patient who developed an incomplete form of PCB, namely acute bibrachial paralysis.

### Report of a Case

A 18-year-old man was referred to our department with a 6-days history of painless muscular weakness in his arms. He denied recent infection, vaccination or consumption of illicit drugs. Otherwise his past medical history was unremarkable.

Physical examination was unrevealing. The patient was alert without obvious cranial nerve dysfunction. Neurological examination revealed moderate weakness in the upper limbs with Medical Research Council (MRC) graded strength of 3/5 in proximal and 2/5 in distal muscles. Motor power was normal in the lower limbs apart from a mild (MRC 4/5) paresis of left foot and toes extension. Deep tendon reflexes were absent in the arms, whereas trace reflexes were noted in the legs. Plantar responses were flexor. Sensory examination as well as cerebellar function proved normal.

An MRI of the brain and cervical spine, as well as ultrasound examination of the extracranial and intracranial arteries, were unremarkable.

Routine hematological and biochemical tests (including thyroid function tests and electrolytes) were normal. In addition, extensive laboratory studies, including AChR-Ab, urine porphobilinogen, heavy metals, autoimmune panel, HIV and Lyme antibodies, were either normal or negative. Subsequently, an enzyme-linked immunosorbent assay study disclosed that immunoglobulin G (IgG) anti- GT1a antibodies were positive, whereas GQ1b-Ab were positive in grey zone.

A lumbar puncture revealed the following: 4 leukocytes per mm<sup>3</sup>, protein 80 mg/dl, glucose 64 mg/dl. CSFs venereal disease research laboratory, viral PCRs (herpes simplex virus, varicella zoster virus, West Nile virus, cytomegalovirus, Epstein-Barr virus, and enterovirus) and oligoclonal bands were all negative.

Nerve conduction studies (NCS) of the median, ulnar and sural nerves demonstrated normal sensory nerve action potential (SNAP) amplitudes and conduction velocities. The most prominent abnormality of the motor NCS was the presence of partial conduction blocks in the right median, left ulnar and left common peroneal nerve (Table 1). Additionally, a significantly low persistence of F-waves in the upper limbs was noted.

Based on the results of the electrodiagnostic testing, the CSF findings and the positive GT1a-Ab, we diagnosed our patient as having a localized type of GBS and treated him with intravenous immunoglobulin (IVIg 0.4g/kg/day for 5 days). Over the following 10 weeks, the patient recovered considerably from his deficits (MRC of 5/5 in the lower limbs and MRC of 4+/5 in the upper limbs). Repeat NCS testing showed resolution of the conduction blocks (Table 1).

|                                 | Day 6        | Day 70      |
|---------------------------------|--------------|-------------|
| <b>Motor conduction studies</b> |              |             |
| <b>Right median</b>             |              |             |
| DML                             | 3.62         | 4           |
| CMAP (wrist/elb)                | 8.1/4.1 (CB) | 8/6.9       |
| CV                              | 58.3         | 56.8        |
| <b>Left ulnar</b>               |              |             |
| DML                             | 3.6          | 3.7         |
| CMAP (wrist/eib./ab. elb)       | 6.3/5/1 (CB) | 8.8/6.9/6,5 |
| CV                              | 63/43        | 56.4/46.8   |
| <b>Left peroneal</b>            |              |             |
| DML                             | 4.84         | 4.2         |
| CMAP                            | 4.9/2.4 (CB) | 5/3.2       |
| CV                              | 39.7         | 47.3        |

**Table 1:** Abnormal findings on serial motor nerve conduction studies.

DML: Distal Motor Latency; CMAP: Compound Muscle Action Potential; CV: Conduction Velocity; CB: Conduction Block.

The abnormal values have been highlighted in bold.

### Discussion

According to large studies, the incidence of PCB variant among patients with GBS has been estimated between 3% and 5% [2,3]. Recently, Wakerley and Yuki have outlined novel diagnostic criteria for PCB variant, based on a retrospective study of 100 neuropathy patients who manifested pharynx, neck and upper limbs weakness within 4 weeks of initial onset [1,4]. The authors suggested that patients with PCB and a minor component of legs weakness, which was also observed in our case, should be still classified as having PCB.

Our patient showed paresis largely confined to his upper limbs. His presentation could be in keeping with the above type of PCB, if one assumes that a minor degree of neck or pharyngeal weakness might have been escaped from clinical detection. Alternatively, the pathophysiological process implicated might have targeted the peripheral nerves of the upper limbs almost exclusively. In this regard, it is of interest that in the large cohort of patients diagnosed with GBS according to the criteria of the Brighton Collaboration, there were 3 (< 1%) cases similar to ours having isolated upper limbs paresis [5]. Among them, 2 demonstrated pure arm weakness persisting at the nadir of their disease. There is also a similar report in the Japanese literature, which describes an individual with acute upper limb dominant GBS sparing oropharyngeal and neck musculature [6].

Despite this highly localized presentation of our patient, there was strong evidence that GBS was the correct diagnosis. Indeed, the CSF, neurophysiological and serological profile was consistent to a GBS variant. Furthermore, other competing disorders with a potential to lead to isolated brachial weakness were excluded. Typical examples comprise watershed brain infarcts [7], bilateral prerolandic cortical and subcortical lesions [8], cervicomedullary junction insults [9] and cervical cord gray matter involvement due to infectious, traumatic or vascular causes [10].

Additionally, bilateral brachial plexopathies [11], paraneoplastic [12], as well as porphyric [13] and toxic neuropathies [14] may rarely produce this striking phenotype.

Although the pathophysiological background of PCB has not been formally tested, there are a few case reports and small series which strongly indicate disturbance of axonal function as the main underlying mechanism. Indeed, in the original series of PCB patients reported by A. Ropper, there were no apparent demyelinating features [15], whereas axonal conduction failure was detected in a subsequent report of two PCB cases [16]. More recently, Chan, *et al.* [17] as well as Capasso, *et al.* [18] described three PCB patients who displayed a characteristic non-demyelinating, reversible conduction failure pattern on NCS.

This is ascribed to antibody-mediated microstructural changes at the nodes and paranodes of motor axolemma leading to nerve impulse conduction failure. The rapid recovery of NCS abnormalities has been attributed to reorganization of sodium channels at the nodal region based on the putative arrest of the immune cascade at the stage of functional impairment [19].

Interestingly, the neurophysiological findings of our patient, as previously mentioned, consisting of multifocal conduction blocks resolving within 10 weeks, without development of remyelinating features (such as prominent temporal dispersion), were compatible with early reversible axonal failure pattern [19]. Not surprisingly, the initial NCS abnormalities of our case fulfilled the recently modified electrophysiological criteria for axonal GBS, proposed by Rajabally, *et al* [20].

### Conclusion

In conclusion, PCB variant of GBS may exceptionally manifest with incomplete phenotype, such as bibrachial paralysis. In these atypical cases, meticulous diagnostic work-up should be introduced, in order to exclude other serious aetiologies. Overall, our case further supports the proposed concept, that PCB represents a localized form of acute motor axonal neuropathy.

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### Declarations of Interest

None.

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