

Case Report

A Rare Presentation of Guillain-Barre Syndrome: Pharyngeal Cervical-Brachial Variant

Mohamed Nadheem ¹✉¹National Hospital of Sri Lanka**Abstract**

Guillain-Barre syndrome is characterized by acute onset symmetrical, generalized, ascending weakness with areflexia. It has wide variety of subtypes according to the involvement of muscle and nerve groups. We describe a case of 70 year old lady developing a rare form of GBS, the pharyngeal-cervical-brachial variant of the disease.

Key words: Guillain-Barre Syndrome, Pharyngeal-cervical-brachial variant, Sri Lanka**Copyright:** ©. 2017 Nadheem M. This is an open access article distributed under the [Creative Commons Attribution License](#), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.**Funding:** None**Competing interest:** None✉ **Correspondence:** nadheem300@gmail.com**DOI:** <http://doi.org/10.4038/amj.v11i1.7626>**Introduction**

Guillain-Barre Syndrome (GBS) is an acute immune mediated polyneuropathy. It includes a heterogeneous group of disorders with various subtypes and variants. GBS is clinically characterized by symmetrical, generalized ascending weakness with areflexia due to peripheral nerve involvement. Pharyngeal-cervical-brachial (PCB) variant is a rare form of Guillain-Barré syndrome and presents with a rapidly progressive oropharyngeal and cervico brachial weakness with areflexia predominantly of the upper limbs. Lower limb muscle power is usually preserved or only mildly affected. Here we report a classical case of PCB variant of GBS affecting a 70-year-old.

Case report

A 70-year-old known hypertensive woman on treatment presented with weakness of left upper limb, more proximal than distal, progressed into weakness of right upper limb and subsequently left lower limb for 4 days duration. Lower limb weakness was less severe compared to the upper limbs. She had difficulty in swallowing for a similar duration, with mild respiratory discomfort. There was no preceding diarrhoeal illness but she described a respiratory illness one month ago.

On examination she was conscious and oriented. Facial power, eye movements and pupillary response were normal. Palatal movements were impaired. There was

weakness of neck muscles with neck flexion of 3/5. Single breath count was more than 25. Shoulder abduction was 2/5 on the right and 1/5 on the left. Distal upper limb power was 2/5 on the left and 3/5 on the right. Power in the lower limbs was 4/5. There was global are flexia with no sensory involvement. Plantar response was flexor bilaterally and there was no demonstrable fatigability or ataxia.

She was afebrile with a blood pressure of 140/80 mmHg and pulse rate of 80 bpm. Cardiovascular, respiratory and abdominal examinations were unremarkable.

Her baseline blood investigations were as follows. White cell count $12.3 \times 10^9/L$, platelet count $197 \times 10^9/L$, serum potassium 4.2 mmol/L, serum sodium 138 mmol/L, , fasting blood glucose 103 mg/dl and serum creatinine 57 umol/L. Haemoglobin level was 13.9g%, Liver function tests were normal. Chest radiograph was unremarkable. Viral panel (EBV, CMV, herpes virus), cultures for Salmonella, Shigella and Campylobacter jejuni and serological examination for mycoplasma pneumonia were negative. Neurophysiological studies done on day 3 of admission indicated motor conduction abnormalities with conduction blocks, and sensory responses were normal suggesting GBS. Cerebrospinal fluid (CSF) examination on day 08 of illness revealed typical "albumino cytological dissociation" with no white or red blood cells. The protein

level was high at 60 mg/dl, with normal glucose. CSF cultures and gram stain were unremarkable.

With clinical and neurophysiological evidence, a diagnosis of PCB variant of GBS was made and the patient was initiated on intravenous immunoglobulin 2mg/kg over 5 days. Strict monitoring of respiratory functions was done with respirometer observing for potential complications. Over the following 10 days, with physiotherapy and supportive care, she made a remarkable recovery with upper limb power of 4/5, normal lower limb strength, neck flexion and normal swallowing.

Discussion

Guillain-Barre syndrome is characterized by bilaterally symmetrical ascending paralysis, absence of deep tendon reflexes, sensory loss, cytoalbuminologic dissociation in cerebrospinal fluid and typical findings in nerve conduction studies.

In 1986, Ropper (1) described the first patients who developed rapidly progressive oropharyngeal, neck and shoulder weakness, with relative sparing of the lower limbs in the absence of sensory disturbance and, mimicked like descending paralysis seen in botulism although relative sparing of the lower limbs were initially thought as the hallmark of the disease some patients were later described to have minimal limb weakness as in our patient.

According to the proposed new criteria for the PCB variant of GBS (2), our patient fulfilled all the features required for the diagnosis. She had relatively symmetrical oropharyngeal weakness, neck weakness, arm weakness and arm areflexia. There was absence of ataxia, disturbed consciousness and prominent leg weakness. She had a monophasic illness pattern and interval between onset and nadir of oropharyngeal or arm weakness was around 2 weeks and there was absence of identifiable alternative diagnosis.

In addition to these she had other strongly supportive features such as, antecedent infectious symptoms, cerebrospinal fluid analysis showing albumino cytological dissociation, neurophysiological evidence of neuropathy.

GBS is one of the auto immune illness preceded by an infectious illness. Auto antibodies against specific

neuronal gangliosides have been implicated in the pathogenesis of different GBS variants. The strongest association for PCB is the presence of IgG anti-GT1a antibodies and is thought to be a useful marker in supporting the diagnosis (3) In our case serological assays were not available in our country.

The presence of additional ophthalmoplegia and ataxia indicates overlap with Fisher syndrome. This is the commonest association with the variant

Patients with pure PCB were more likely to require intubation than those with overlap syndromes and this correlated with degree of bulbar involvement (3). Fortunately, our patient did not need ventilation.

Conclusion

PCB variant of GBS should be remembered in patients with symptoms of bulbar and upper extremity weakness not only for early diagnosis but also to plan treatment early and follow up potential complications. Due to the unfamiliarity with PCB variant, clinical picture is often misdiagnosed as brainstem stroke, myasthenia gravis or botulism.

References

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