CASE REPORT

An unusual presentation of Guillain-Barré syndrome in a young man with bilateral upper extremity weakness

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SUMMARY

A 30-year-old man attended the emergency department with a 4-day history of progressive, bilateral upper limb weakness. He had mild shortness of breath and occasional swallowing difficulties. One month prior to presentation, he had flu-like symptoms and diarrhoea. Examination revealed upper limb hypotonia, symmetrical distal arm weakness and hyporeflexia. Power and reflexes in the lower limbs were normal. Nerve conduction studies and lumbar puncture demonstrated features consistent with Guillain-Barré syndrome (GBS). The patient was treated with a 5-day course of intravenous immunoglobulins. He improved significantly over the next 2 weeks. Breathing and swallow function did not deteriorate and required no further intervention. He had a sustained improvement, and remained at baseline 1 year later. Work-up for underlying structural, infectious, inflammatory and paraneoplastic aetiologies were negative. Serum antiganglioside antibodies were positive for the anti-GT1a IgG isotype supporting the clinical diagnosis of the pharyngeal-cervical-brachial variant of GBS.

BACKGROUND

The pharyngeal-cervical-brachial (PCB) variant of Guillain-Barré syndrome (GBS) is rare and may be misdiagnosed by clinicians because of its atypical presenting features with predominantly upper limb symptoms. It responds well to intravenous immunoglobulin, once recognised.

CASE PRESENTATION

A 30-year-old man presented to the emergency department, with progressive upper limb weakness, mild dyspnoea, dysphagia and dysarthria, over 5 days. He worked with computers and had noticed difficulty typing on the keyboard and opening bottles. He felt his arms were less affected proximally and that his legs were minimally affected. He had mild shortness of breath on exercise. He felt he needed to cut food into smaller pieces over the preceding week, in order to swallow safely. He denied any choking episodes.

On examination, his pupils were normal. There was no evidence of ptosis or ophthalmoplegia. His speech was mildly dysarthric. There was no tongue atrophy or fasciculations. Tone was normal. There was distal (Medical research council (MRC) grades 1–2/5) more than proximal weakness (MRC grades 2–3/5) in his upper limbs, with associated neck flexion weakness (MRC grade 4/5). His upper

extremities were hyporeflexic; lower limb examination revealed very mild proximal weakness (MRC grade 4+/5) only, with intact reflexes.

The patient's medical history was significant for migraine. He drank about 20–25 units of alcohol on weekends. He was a non-smoker and denied illicit drug use.

INVESTIGATIONS

Nerve conduction studies (NCS) were performed on the day of presentation. In the upper limbs, there were prolonged distal motor latencies of the right median and ulnar nerves. Sensory responses were relatively preserved, and F wave responses were absent. In the lower limbs, conduction block was demonstrated in the left common peroneal nerve, lower limb NCS were otherwise entirely normal. These findings would be supportive of a diagnosis of GBS predominantly affecting the upper limbs (table 1).

A lumbar puncture was performed on the day of presentation. This supported a diagnosis of GBS with normal cell count but an elevated protein level of 1.05 g/L (range 0.1–0.4 g/L). Cerebrospinal fluid glucose, white cell count, Gram stain, culture and sensitivity were normal. Serum autoimmune panel and vasculitic screen were negative.

Serum infectious screens for Lyme disease, HIV, hepatitis B and C, cytomegalovirus, Epstein-Barr virus and herpes simplex virus were negative. Paraneoplastic antibodies panel was negative. Antivoltage-gated calcium channel antibody and antiacetylcholine receptor antibody were negative. Antiganglioside antibodies were positive for the IgG isotype of anti-GT1a antibody, strongly in support of the clinical diagnosis of the PCB variant of GBS.

MRI of the cervical spine was normal. There was no evidence of cervical disc disease or radiculopathy. There was no abnormal signal to suggest an anterior horn cell syndrome, and no intramedullary T2 signal abnormality ('snake eye' sign) suggestive of a compressive myelopathy or ossification of the posterior longitudinal ligament. The patient also had a normal MRI of the left and right brachial plexus.

Speech and language therapy review revealed mild dysarthria and dysphagia without any aspiration risk, and no need for altered diet. Pulmonary function tests were normal.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis in this case included anterior cervical cord syndromes such as Hirayama

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Table 1 Nerve conduction studies, upper and lower limbs

Nerve	Amplitude (reference normal)	Latency (reference normal)	Conduction velocity (reference normal)	F wave (reference normal)
Left median sensory	19.3 μ V (>20 μ V)	2.4 ms (<3.5 ms)	59 m/s (>50 m/s)	–
Left ulnar sensory	7.0 μ V (>10 μ V)	2.1 ms (<3.7 ms)	50 m/s (>50 m/s)	–
Right median motor				
Wrist	9.2 mV	5.2 ms (<4.4 ms)	48 m/s (>49 m/s)	Absent (<29 ms)
Antecubital fossa	9.2 mV (>4 mV)	8.3 ms		
Left median motor				
Wrist	13.6 mV	4.1 ms (<4.4 ms)	42 m/s (>49 m/s)	33 ms, small and impersistent (<29 ms)
Antecubital fossa	2.0 mV (>4 mV)	9.8 ms		
Right ulnar motor				
Wrist	0.9 mV	5.6 ms (<3.5 ms)	53 m/s (>49 m/s)	Absent (<29 ms)
Elbow	0.9 mV (>6 mV)	9.3 ms		
Left ulnar motor				
Wrist	12 mV	3.7 ms (<4.4 ms)	58 m/s (>49 m/s)	Absent (<29 ms)
Elbow	2 mV (>6 mV)	7.5 ms		
Left peroneal motor				
Ankle	3.0 mV	4.5 ms (<6 ms)	47 m/s (>46 m/s)	–
Knee	0.6 mV (>2 mV)	10.7 ms		
Left tibial motor	7.5 mV (>3 mV)	6.6 ms (<6 ms)	Not calculated	48.5 ms (<50 ms)
Left sural (sensory)	18.1 μ V (>2 μ V)	2.4 ms (<4.2 ms)	48 m/s	–

disease (non-progressive juvenile spinal muscular atrophy), bilateral cervical radiculopathy due to trauma, herniated disc or osteophyte impingement, bilateral brachial plexopathy and neuromuscular junction disorders such as Lambert Eaton myasthenic syndrome or botulism.

Clinically, there was no sensory level or urinary retention to support a myelopathy. The patient's weakness was also relatively symmetrical and would typically be unilateral in Hirayama disease. MRI of the cervical spine was also subsequently normal. Normal sensory NCS in the upper limbs made brachial plexopathy unlikely. Imaging of the brachial plexus bilaterally was also normal. Lambert Eaton syndrome is usually lower limb predominant and voltage-gated calcium channel antibodies were absent in this patient. Myasthenia gravis usually presents with proximal rather than distal limb weakness, and patients may have extraocular and/or bulbar musculature involvement that worsens as the day progresses, which was not the case in this patient. No high-risk behaviours associated with botulism were identified in this patient and his pupils were of normal size with an intact pupillary light response throughout his illness.

TREATMENT

The patient received a 5-day course of intravenous immunoglobulin, to which he responded very well. He underwent inpatient rehabilitation with physiotherapy, occupational therapy, and speech and language therapy.

OUTCOME AND FOLLOW-UP

There was a sustained improvement in power, breathing and swallow within 2 weeks of intravenous immunoglobulin treatment and rehabilitation. The patient was referred to community rehabilitation services and has maintained a full recovery since discharge.

DISCUSSION

GBS is the most common cause of acute muscular paralysis, affecting 0.4–2.4/100 000 people annually.¹ The PCB variant of GBS is rare (0.07–0.25/100 000 annually²) and often

misdiagnosed. This variant is characterised by predominantly oropharyngeal, neck and upper limb weakness. Other features supportive of a diagnosis may include diaphragmatic involvement, sparing of lower limb strength and reflexes, minimal or no sensory deficit and an axonal pattern on NCS. It is an important entity to recognise as, despite its atypical presentation, similar to the majority of GBS cases, it responds well to early treatment with intravenous immunoglobulin. Once treated, most patients experience a complete recovery.³

Learning points

- ▶ The pharyngeal-cervical-brachial variant of Guillain-Barré syndrome (GBS) is rare.
- ▶ It presents with any of upper limb weakness, respiratory failure and/or dysphagia. Lower limbs may be spared.
- ▶ Patients do have the typical cerebrospinal fluid albuminocytological dissociation of GBS.
- ▶ NCS abnormalities are upper limb predominant.
- ▶ It responds to intravenous immunoglobulin, once recognised.

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