Case Report: An Unusual Case Of Guillain-Barré Syndrome

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Introduction

Guillain-Barré Syndrome (GBS) is an acute immune mediated disease of the peripheral nervous system with a wide range of clinical symptoms and great variability in outcome. The reported incidence ranges from 1-2 cases per 100,000 persons per year[1]. Poor prognostic features include preceeding gastrointestinal illness, age more than 50 years, rapid onset, and the increasing severity of presentation, including bulbar features[2].

GBS is generally viewed as an autoimmune disorder, triggered in most cases by an antecedent bacterial or viral infection. Recent evidence suggests that *Campylobacter jejuni* enteritis may be the most common antecedent infection, preceeding GBS in approximately 25-38% of patients[3]. The target of the aberrant immune response seems to be within the Schwann cell surface membrane or the myelin, resulting in primary inflammatory demyelination as the major pathological finding[4]. Various antibodies to nerve cell components, notably antiglycolipids such as anti-GM1 have been detected in the serum of these patients and are often used in confirming a diagnosis of Guillain-Barré syndrome[5].

Plasma exchange and intravenous immunoglobulin (IVIG) are the mainstay of active treatment[6]. The former directly removes humoral factors such as antibodies, immune complexes, complement and other non-specific inflammatory mediators from the serum of patients[7]. The recovery of a patient with poor prognostic indices is often prolonged even with plasma exchange[2].

We report a patient with *Campylobacter jejuni* associated atypical GBS who responded immediately following the first treatment with plasma exchange.

Case Report

A 55 year old man presented to hospital complaining of generalised weakness. Three days prior to admission he noticed a weak voice and nasal regurgitation. The next day he described lower limb weakness. On the day prior to admission he developed weak arm and neck muscles. He also had abnormal sensation in his hands and left leg. He had no diplopia or blurred vision. Three weeks prior to admission he had a diarrhoeal illness which was confirmed later in hospital to be caused by *Campylobacter jejuni* infection. The patient's clinical symptoms and progression are summarized in Tables 1 and 2. A diagnosis of Guillain-Barré Syndrome with bulbar symptoms was made based on the clinical symptoms.

On day 2 he was commenced on plasma exchange. The patient showed a remarkable recovery by day 3 (Table 1), after one session of plasma exchange. On day 8 the patient had recovered completely and he was discharged on day 12. He was well at follow up 6 months later.

Table 1: Clinical symptoms and progression of illness following admission

Number of Days Admitted	1(admission)	3	8
Total Plasma Exchange Sessions	0	1	4
Gag Reflex (indicating bulbar palsy)	Absent	Present but diminished	Present
Power Upper Limbs			
Proximal	2/5	4/5	5/5
Distal	3/5	4/5	5/5
Power Lower Limbs			
Proximal	2/5	4/5	5/5
Distal	3/5	4/5	5/5
Reflexes Upper Limbs	0	2+	3+
Reflexes Lower Limbs	1	2+	3+
Altered Sensation (dermatomes)	C7,C8 right and left side. L2-L5 left side	C7,C8 right and left side. L2-L5 left side	Normal Sensation
CSF Protein	Raised		
Nerve Conduction	Normal		
Antibodies			
Anti-GM1	Negative		
Anti-GQ1b IgG	Strongly positive	1 in 64,000	

Table 2: Hughes clinical scoring system[8]

	DAY 1	DAY 3	DAY 8
Grade 5 – ventilated			
Grade 4 – bedridden	G4		
Grade 3 – walk 5m aided		G3	
Grade2 – ambulating			
Grade 1 - normal			G1

Discussion

This patient presented in an atypical fashion given that he had predominantly bulbar features and asymmetrical weakness, with his proximal strength greater than his distal. He also had unusual serology. He was strongly positive for Anti-GQ1b IgG antibody. This antibody is highly specific for the Miller Fisher variant of GBS characterised by ophthalmoplegia, areflexia and ataxia, or the newly termed "Anti-GQ1b IgG antibody syndrome"[9,10]. These syndromes are associated with ataxia and ophthalmoplegia neither of which were seen in this patient[10] .

The patient also had poor prognostic indices for GBS including age greater than 50, rapid onset of symptoms, with a predominantly bulbar pattern, and a preceeding *Campylobacter jejuni* diarrhoeal infection[2]. These would all indicate that the patient would have been likely to have had a long and difficult recovery. Surprisingly, the patient made a very rapid recovery. After one session of plasma exchange his gag reflex recovered and his limb weakness had improved significantly. By day 8 he had completely recovered and has remained well on subsequent follow up.

The North American Plasmapheresis Study reported the median days to improvement by one grade in the Hughes Clinical Scoring System (Table 2) was 19 days, and time to walk unaided was 53 days in patients treated with plasma exchange[7,8,9]. The patient returned to normal functioning within 8 days, an improvement of three grades. Shah *et al.*[12] reported a rapid recovery in a child treated with IVIG ambulating aided in 6 days, but this speed of recovery has not been reported in adults or in patients treated with plasma exchange. The rapid recovery of the patient presented here was especially remarkable in view of his very poor initial prognostic indices.

Conclusion

We report a patient with a very atypical presentation of Guillain-Barré Syndrome who responded extremely rapidly to plasma exchange treatment. This underlines that despite poor initial prognosis some patients will respond very well to early treatment. The patient was also Anti-GQ1b IgG antibody positive therefore indicating that the supposed "Anti-GQ1b IgG syndrome" could in fact be broadened to include Guillain-Barré Syndrome[10].

References

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