Individual patients who experienced both Guillain-Barré syndrome and CIDP

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INTRODUCTION

Although Guillain-Barré syndrome (GBS) and chronic inflammatory demyelinating polyneuropathy (CIDP) have been considered as separate entities, some authors have argued that they may be part of a continuum of inflammatory demyelinating neuropathies.[1 2, 3] GBS is defined as having a progressive onset phase of less than 4 weeks; CIDP of at least 2 months. [4-6] However, some CIDP patients have an acute onset, resembling GBS, and some patients with GBS may relapse or experience treatment-related fluctuations (TRFs).[1, 7] We describe four patients all having had separate episodes of both GBS and CIDP that fulfilled the clinical and diagnostic criteria.[4-6] These four patients came to our attention during a period of twenty years in which over 700 GBS and 150 CIDP patients were seen or enrolled in clinical studies. Most GBS patients were followed up for a period of 1 year, the majority of CIDP patients for many years.

RESULTS

Patient 1

A 46-year-old man developed weakness and numbness in his limbs, worsening over 1 year. He fulfilled the clinical and electrodiagnostic criteria for CIDP and responded well to intravenous immunoglobulin (IVIg) treatment, every two weeks. The IVIg dosage was reduced several times with immediate deterioration as a result. Treatment was continued for 6 years and then stopped; thereafter he remained in remission. Twelve years later, he developed tetraparalysis and facial palsy within 48 hours, 2 weeks after a bout of diarrhoea. He fulfilled criteria for GBS. He was treated with IVIg, and he required artificial ventilation for 2 months. He reached a near complete recovery and no new episodes occurred (Table 1).

Patient 2

A 33-year-old man developed progressive weakness and paraesthesia in all limbs over the course of 1 week. Three days later he was unable to walk. He fulfilled diagnostic criteria for GBS. After IVIg, he initially improved, but 9 days later, he developed facial palsy and opthalmoplegia and he was re-treated with IVIg. He subsequently became tetraplegic needing artificial ventilation. Initially it was thought that he had TRFs, although after five exacerbations all rapidly responding to IVIg, a diagnosis of CIDP was more appropriate. His symptoms required maintenance IVIg treatment for two and a half years. Electrodiagnostic studies were compatible with CIDP (Table 1).

Table 1. Case characteristics

Case	Episode	Sex	Age	Diagnosis	Preceding infection/ incident	Cranial nerve dysfunction	Need for artificial respiration
1	1	M	46	CIDP	-	-	-
1	2	M	64	GBS	Gastrointestinal	VII	+
2	1	M	33	GBS	Cytomegalovirus	VII,ophtalmoplegia	+
2	2	M	34	CIDP	-	-	-
3	1	M	34	GBS	Gastrointestinal	-	-
3	2	M	39	Recurrent GBS with TRF	-	-	-
3	3	M	40	CIDP	-	-	-
4	1	M	39	GBS	Flu-vaccination	VII	+
4	2	M	68	CIDP	-	-	-

GBS, Guillain-Barré syndrome; CIDP, chronic inflammatory demyelinating polyneuropathy; TRF, treatment-related fluctuation.

Patient 3

A 34-year-old man developed a rapidly progressive tetraparesis after an episode of diarrhoea. He was diagnosed with GBS and recovered fully after IVIg treatment. Five years later, he was readmitted with similar symptoms that had developed over 1 week. A lumbar puncture showed a normal protein level of 0.43 g/l. Two to three months later his symptoms returned and were successfully treated with IVIg. One and a half years later he experienced another relapse over a 2-month period. His disease course suggested CIDP, and during another IVIg treatment his symptoms stopped and did not recur (Table 1). Electrodiagnostic studies were compatible with the diagnosis of CIDP.

Patient 4

A 39-year-old man developed tingling sensations and weakness in his limbs after a flu vaccination earlier that month. He then developed bilateral facial palsy and fulfilled diagnostic criteria for GBS. Within 3 days he needed artificial ventilation, which continued for 6 weeks. He recovered slowly. Twenty-nine years later, he developed progressive weakness and numbness over a period of 2 months, compatible with CIDP. No preceding illness or vaccination was reported. Electrodiagnostic studies were compatible with the diagnosis of CIDP. He has been successfully treated with maintenance IVIg treatment, every 4 weeks, for a period of 1 year so far (Table 1).

DISCUSSION

These case histories show that GBS and CIDP can occur in the same patient, and underline the difficult differential diagnosis of GBS or acute CIDP. Patient 1 initially had a course fully compatible with CIDP. Many years later he developed an acute polyneuropathy with severe weakness and respiratory failure after a gastrointestinal infection, a classic example of GBS. Patient 2 had a GBS-like acute onset with respiratory insufficiency, but finally developed CIDP. He experienced five exacerbations and needed maintenance IVIg for more than 2 years thereafter, which suggests that the diagnosis of CIDP with acute and severe onset was more appropriate.[7] Patient 3 experienced two episodes of weakness with acute onset after an infection, suggesting recurrent GBS. The minor deteriorations after each IVIg treatment were considered to be TRFs. The progressive symptoms which developed 1 year later over a longer period of 2 months were compatible with CIDP. [5] Patient 4 had rapidly progressing symptoms of GBS after a flu vaccination. Twenty-nine years later weakness gradually returned: this episode was compatible with CIDP. The patient improved but required intermittent IVIg treatment.

Case histories of patients with multiple episodes of weakness and characteristics of both GBS and CIDP are rare.[3, 8] One patient with GBS-like deterioration after administration of IVIg for CIDP has been reported.[9] These four case histories in which individual patients were affected with both GBS and CIDP suggest that in a proportion of patients GBS and CIDP may constitute a clinical continuum, or that there are common host factors which influence susceptibility to these disorders. Apart from striking similarities, GBS and CIDP also show clear differences. Anti-ganglioside antibodies are frequently detected in GBS, but generally absent in CIDP.[10] Preceding infections are less frequently reported in CIDP, but infections during the course of CIDP may clearly worsen symptoms.[10] It should be noted that most CIDP patients improve after steroids, whereas GBS patients do not.[11] On the other hand, most GBS and CIDP patients improve after IVIg or plasma-exchange. Patients with subacute idiopathic demyelinating polyradiculoneuropathy with progressive weakness of 4-8 weeks have been described, and this entity bridges the gap between GBS and CIDP.[2, 12]

Although we are aware that most patients clearly fit the diagnostic criteria of GBS or CIDP alone and that patients having separate episodes of GBS and CIDP are extremely uncommon, it is important to be aware of the possibility that both GBS and CIDP can co-occur in a single patient and should be diagnosed and treated accordingly. Although it can not be excluded that individuals get both GBS and CIDP by chance, these case descriptions may indicate that in some patients CIDP and GBS are part of a clinical and pathophysiological continuum instead of fully separate entities.

REFERENCES

1. Grand'Maison F, Feasby TE, Hahn AF, Koopman WJ. Recurrent Guillain-Barré syndrome. Clinical and laboratory features. *Brain* 1992;115:1093-106.
2. Hughes R, Sanders E, Hall S, Atkinson P, Colchester A, Payan P. Subacute idiopathic demyelinating polyradiculoneuropathy. *Arch Neurol* 1992;49(6):612-6.
3. Mori K, Hattori N, Sugiura M, et al. Chronic inflammatory demyelinating polyneuropathy presenting with features of GBS. *Neurology* 2002;58(6):979-82.
4. Asbury AK, Cornblath DR. Assessment of current diagnostic criteria for Guillain-Barré syndrome. *Ann Neurol* 1990;27 Suppl:S21-4.
5. Research criteria for diagnosis of chronic inflammatory demyelinating polyneuropathy (CIDP). Report from an Ad Hoc Subcommittee of the American Academy of Neurology AIDS Task Force. *Neurology* 1991;41(5):617-8.
6. Joint Task force of the EFNS and the PNS. European Federation of Neurological Societies/ Peripheral Nerve Society Guideline on management of chronic inflammatory demyelinating polyradiculoneuropathy. *J Peripher Nerv Syst* 2005(10):220-8.
7. Ruts L, van Koningsveld R, van Doorn PA. Distinguishing acute-onset CIDP from Guillain-Barré syndrome with treatment related fluctuations. *Neurology* 2005;65(1):138-40.
8. Odaka M, Yuki N, Hirata K. Patients with chronic inflammatory demyelinating polyneuropathy initially diagnosed as Guillain-Barré syndrome. *J Neurol* 2003;250(8):913-6.
9. Krasenbrink I, Kaps M, Blaes F. IVIg-induced acute polyneuroradiculitis in a patient with CIDP? *Eur J Neurology* 2007;14(5):e9.
10. van Doorn PA. Treatment of Guillain-Barré syndrome and CIDP. *J Peripher Nerv Syst* 2005;10(2):113-27.
11. Hughes RA, Swan AV, Raphaël JC, Annane D, van Koningsveld R, van Doorn PA. Immunotherapy for Guillain-Barré syndrome: a systematic review. *Brain* 2007;130(Pt 9):2245-57.
12. Oh SJ, Kurokawa K, de Almeida DF, Ryan HF, Jr., Claussen GC. Subacute inflammatory demyelinating polyneuropathy. *Neurology* 2003;61(11):1507-12.