‘Hyperacute’ Guillain-Barré Syndrome

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Dear Sir,

The criteria for the diagnosis of acute inflammatory demyelinating polyneuropathy (AIDP) or Guillain-Barré syndrome (GBS) require the development of progressive weakness up to maximal motor disability within 4 weeks. Several studies have evaluated the time to maximal motor incapacity in the disorder. In one survey, 75% of patients reached their nadir within 2 weeks, 92% within 3 weeks and 94%

within 4 weeks [1]; in another, the mean time from onset of symptoms to nadir was 9.7 (± 6.9) days (median time, 7 days) [2]. In an additional study from Japan [3], the median time to nadir was 18.0 $\pm$ 10.4 days for AIDP and 11.5 days $\pm$ 7.8 days for the acute motor axonal neuropathy (AMAN) variant.

The occurrence of GBS, in which maximal incapacity is reached within a day or

two, ‘hyperacute’ GBS, is a rather unusual event. We collected 5 patients who were admitted to 4 academic institutes in Israel (Hadassah and Shaare-Zedek, Jerusalem, Hillel-Yaffe, Hadera and Soroka, Beer-Sheva) over a 10-year period (1997–2006) with a fulminant course of disease. These medical centers serve a combined population of approximately 1.25 million and the cases were retrospectively collect-

Table 1. Clinical and laboratory features in 5 patients with ‘hyperacute’ GBS

Patient No.	Sex/age	Antecedent infection	Time to nadir	Maximal deficit	CSF	Therapy	Outcome
1	F/30	URI within the previous week	20 h	quadriplegia, respiratory failure	normal protein, 3 cells/ μ l	PE	full functional recovery, residual areflexia
2	M/40	gastroenteritis 5 days before admission	24 h	quadriplegia	normal protein, 2 cells/ μ l	IVIg	full functional recovery
3	M/42	gastroenteritis 10 days prior to admission	36 h	quadriplegia, respiratory failure	protein, 55 mg/dl (n < 45) acellular	IVIg	fully recovered
4	M/57	no noticeable trigger	36 h	quadriplegia, respiratory failure, ophthalmoplegia, dysphagia, facial diplegia	normal protein acellular	IVIg	6 months after disease onset, independent gait and driving ability, intelligible speech and intact glutition
5	F/52	influenza vaccine	24 h	severe quadriparesis and moderate dyspnea	normal protein acellular	IVIg	5 months after disease onset, still in rehabilitation, regaining independent gait capability

IVIg = Intravenous immunoglobulins; URI = upper respiratory infection; PE = plasmapheresis.

Table 2. Electrodiagnostic studies in 5 patients with 'hyperacute' GBS

Patient No.	Median nerve				Ulnar nerve				Peroneal nerve				Needle EMG	Interpretation
	DL	CMAP	CV	F lat	DL	CMAP	CV	F lat	DL	CMAP	CV	F lat		
1	5.7	1.2	26	NE	4.7	0.9	27	NE	NE				active denervation in several muscles	demyelination with secondary axonopathy
2	4.4	2.2	46	32	3.0	2.3	45	34	5.7	0.8	35	57	no signs of denervation	predominantly axonal motor polyneuropathy
3	4.2	2.4	53	29	3.3	1.7	48	30	5.1	0.65	38	58	scattered fibrillation potentials	early axonal damage
4	5.2	9.0	52	NE	3.3	6.9	52	NE	4.5	4.5*	44	54	no signs of denervation	scattered demyelination
5	4.9	3.1*	28	NE	4.6	5.2	50	NE	7.2	1.1*	39	NE	no signs of denervation	asymmetrical demyelination and conduction blocks

CMAP = Compound muscle action potential (mV); DL = distal latency (ms); CV = conduction velocity (m/s); F lat = F wave latency (m).

* Abnormal proximal/distal amplitude reduction (conduction block), NE = not elicited; ND = not done.

ed from about 160 GBS patients treated during this period in the 4 hospitals. We hereby report their clinical features and outcome.

Case Report (Patient 1) (tables 1, 2)

A 30-year-old previously healthy woman came to the emergency room several hours after the onset of paresthesias in all 4 limbs. Her neurological examination was normal, but since she complained of shortness of breath, she was admitted for observation. Within several hours she developed progressive weakness in all four limbs and respiratory failure. An urgent endotracheal intubation was performed and assisted ventilation was initiated. At that point she had global areflexia, with weakness in all four limbs of 1–2/5 on the MRC scale and superficial sensory deficit in a glove and stocking distribution. No cognitive deficit was noted, her cranial nerves (including the facial) were intact and no pyramidal reflexes were elicited. Lumbar puncture revealed clear fluid under normal pressure with normal protein level of 350 mg/l, normal glucose and 3 WBC/mm³. She was transferred to the intensive care unit where treatment with plasma exchange was initiated. Cervical and brainstem MRI scans were normal, but at that stage nerve conduction studies were not obtained. Twelve-day posthospitalization elective tracheotomy was carried out. Her condition started to improve 3 weeks from disease onset, and she was transferred to the neurological depart-

ment and later on to rehabilitation. Two electrodiagnostic studies were then performed and demonstrated reduced compound muscle action potentials, prolonged distal latencies, prolonged F waves and active denervation in several muscles interpreted as demyelinating polyneuropathy with secondary axonal changes. Within 9 months of rehabilitation she sustained a complete functional and neurological recovery, except for global areflexia.

Histories of the 5 patients with 'hyperacute' GBS are summarized in table 1. The age range was 30–57, 4 of the 5 had a recent history of infection or vaccination, and all developed complete or nearly complete quadriplegia with at least partial respiratory compromise within a day or two. Two patients, numbers 3 and 5, also developed severe autonomic instability with marked fluctuations of blood pressure and heart rate requiring frequent pharmacological modification. Four of the 5 patients underwent electrodiagnostic testing during the initial 3 days of their illness (sample data seen in table 2), with 2 showing early signs of demyelinating and 2 having evidence for axonal involvement. The fifth patient (No. 1) underwent her first study on day 12 after symptom onset. The CSF was normal in 4 of the 5 cases and mildly elevated protein content was found in the fifth. All patients either completed a satisfactory functional and neurological recovery or are still in active stage of rehabilitation (table 1).

Discussion

'Hyperacute' GBS appears to be an unusually rare condition. While drawing generalizations from our very small series might be impossible, this group seems to be indistinguishable from most Israeli GBS patients in their demographic features, antecedent history and outcome. Clinical, laboratory and electrodiagnostic evidence suggest distal involvement with relative sparing of the nerve roots: none of the patients had low back pain, the CSF protein levels were normal or only mildly elevated, and the nerve conduction studies were suggestive of distal rather than proximal peripheral nerve involvement. Only 2 of the 4 patients with electrodiagnostic studies showed evidence for axonal involvement in accordance with findings suggesting shorter time to nadir in axonal GBS subtypes [3].

The outcome of the disorder was favorable in all 5 patients. While a shorter latency to nadir was found to be an adverse prognostic factor in several studies [4], this was not the case in others [1]. Similarly, although axonal involvement was associated with poorer prognosis in some studies [1, 3] this is not universally so [5]. The younger age of some of our patients is considered a favorable prognostic feature [1]. Likewise, presence of a history of infection is associated with better prognosis [2]. Despite the severe autonomic involvement seen in 2 of the patients, their outcome did not differ from that of the others.

This report should draw attention to an unusual and apparently uncommon variant of GBS. The small number of cases does not enable to fully analyze its anatomical, pathogenetic and clinical features but it seems that there is no clear clinical or laboratory marker to distinguish them from 'usual GBS'. This entity should be kept in mind along with diagnoses such as brainstem stroke, botulism and CNS inflammatory disorders when facing baffling cases with acute neurological deterioration.

References

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