Chronic Inflammatory Demyelinating Polyneuropathy: Treatable Hypertrophic Neuropathy

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Chronic inflammatory demyelinating polyneuropathy (CIDP) is a chronic demyelinating disorder presenting with progressive, stepwise, or relapsing muscle weakness (1). It is characterized by the presence of multifocal areas of demyelination in the peripheral nervous system including the spinal nerve roots. Cranial nerves are also involved occasionally (2). Since the causal mechanisms are not yet understood, some authors (3) approve of using the word 'idiopathic' in place of 'inflammatory' for the 'i' of 'CIDP'. To recognize CIDP is important in any neurological units, because this disorder is basically treatable. It is, however, often difficult or impossible to differentiate from other neuropathies on the basis of the phenotypic expression in an individual patient. Clinical diagnosis totally depends on skilled peripheral nerve conduction study; electrophysiological evidence of active or ongoing multifocal demyelination, or motor nerve conduction block associated with considerable conduction delay, is essential (1).

Since the early recognition of this disorder, peripheral nerve thickening has been noted by some careful observers. In the late 50's, Austin first described palpable thickened nerves in his classic steroid-dependent case (4). He mentioned interstitial edema to be an important cause of enlarged nerves, although true interstitial hypertrophy associated with onion bulb formation could develop after recurrent demyelination and remyelination. More recently, Thomas et al presented 4 cases with thickened nerves from among 6 CIDP cases associated with multiple sclerosis (MS)-like central nervous system (CNS) demyelination (5). In 1995, we reported a 9-year-old boy presenting visible posterior auricular nerves under the skin of the lateral sides of the neck (6). Because his weakness had been noticed early in his infancy and he had remarkable pes cavus deformity, he was under observation as an unusual case with genetically-determined neuropathy, or hereditary motor sensory neuropathy (HMSN). Our electrophysiological investigation, however, revealed multifocal conduction abnormalities suggesting acquired demyelination rather than homogeneous abnormalities due to genetic setting. Luckily he showed considerable beneficial response to steroid therapy, in spite of the long history of his disease of more than 8 years and the severe axonal damage revealed in the biopsied sural nerve. It was particularly interesting that the peripheral nerve thickening quickly subsided during the initial two-month steroid therapy. In this volume, Niino et al (7) reported, another CIDP patient associated with hypertrophic nerves; they found massive thickening of the nerve roots and the peripheral nerves, which were beautifully demonstrated by magnetic resonance imaging (MRI) technique. It was regretful that their patient showed a poor response to steroid administration, which is, however, not uncommon in cases with a long disease history or after many recurrences.

See also p 445.

CIDP was previously believed to be an uncommon disorder; in one series of patients with inflammatory demyelinating neuropathies (8), there were 102 patients with Guillain-Barré syndrome (GBS), and 28 with CIDP. From this data, the annual incidence of CIDP must be only about 0.25 to 0.5 per 100,000 of population, or 2.5 to 5 per million, since it is known that the annual incidence of GBS is 1 to 2 per 100,000. This estimate is, however, based on very uncertain assumptions. A recent report stated that CIDP represents a significant number of initially-undiagnosed acquired neuropathies, with estimates varying from 10 to 20% (9). Moreover, our recent survey (10) revealed that the prevalence rate is roughly estimated to be around 2 to 3 per 100,000 of population in the northern Honshu area. Therefore, it is now becoming clear that CIDP is not merely an uncommon disorder, but it could be another neuropathy in a neurological service. Actually, in our series of CIDP patients, the initial diagnosis included hereditary motor sensory neuropathy, diabetic polyneuropathy, motor neuron disease, recurrence of GBS, and so on (11, 12).

Some patients show considerable resistance to steroid therapy, however, recent randomized controlled trials have confirmed that plasmapheresis and intravenous human immunoglobulin are also effective (13, 14). Both therapies are often advantageous for steroid-resistant patients as well (10). It should be mentioned, however, that the beneficial response is most likely in patients with acute relapse or with disease of one year or less (10, 13, 14). Therefore, to diagnose CIDP during the early stage of the disease, and to start appropriate therapy as early as possible are two of the basic principles of strategy for the treatment of CIDP; complete remission is infrequent but possible (10).
References