Nevertheless, in the largest case series SLE developed in only 2 of 108 patients, so the association may be a chance finding.⁶

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Chronic inflammatory demyelinating polyradiculoneuropathy with ataxia

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Occasionally, chronic inflammatory demyelinating polyradiculopathy presents with ataxic features. This should be borne in mind to prevent misdiagnosis and delay in treatment.

CASE HISTORY

A woman aged 80 had for six months been experiencing unsteadiness, right shoulder pain, paraesthesia in her

forearms and fatigue. Previously she had been fit and well, and her only medication was quinine sulphate for muscle cramps. She was a non-smoker, drinking alcohol occasionally. On examination, muscle power was sub-normal (4/5) in the distal arms and legs but 5/5 elsewhere; tone was normal. Reflexes were reduced, with flexor plantar responses. There was mild ataxia in the upper limbs but more severe ataxia of the lower limbs prevented her from standing. The cranial nerves were normal. Initially there were no sensory changes but after three weeks sensory impairment was noted in the distal upper limbs, with pseudoathetosis. Laboratory indices, including full blood count, blood sugar, vitamin B12, red cell folate, protein electrophoresis, autoantibody screen and tests of liver and thyroid function were unrevealing. MRI of the head showed no focal lesions and normal ventricular size. On neurophysiological testing all sensory action potentials were absent; in motor conduction there was no definite block but distant latencies were delayed, proximal conduction was slow (Table 1) and there was temporal dispersion of compound action potentials. The suggested diagnosis was chronic inflammatory demyelinating polyradiculoneuropathy (CIDP). Cerebrospinal fluid had a raised protein at 1.87 g/L with no organisms or cells seen.

The patient was then further investigated to exclude paraneoplastic inflammatory neuropathy, all tests being negative. Because paraprotein was absent, antibody to myelin-associated glycoprotein was not sought. CIDP was diagnosed and she received a five-day course of intravenous immunoglobulins followed by prednisolone, 60 mg daily with reducing dose, and additionally bisphosphonate for bone protection and gabapentin for neuropathic pain. She recovered well and six months later she was mobile and independent.

COMMENT

CIDP can be difficult to diagnose because of its wide variation in symptoms and signs and its slowly progressive nature. Without treatment, mortality is about 10%. It occurs at all ages (prevalence 1–2 per 100 000) and affects men more frequently than women. The disease is seen as the chronic form of Guillain-Barré syndrome. Because of the inflammatory cells and macrophage-mediated demyelination seen on electron microscopy of sural nerve biopsy specimens and its responsiveness to immune-modulating treatments, an autoimmune basis is suspected. No target has definitely been identified, although myelin protein P0 has been suggested.¹ Demyelination is multifocal and can involve the distal nerve terminals, intermediate nerve segments, and nerve roots. Cranial nerves are mildly affected in some patients. On occasion, as here, CIDP presents with a mainly ataxic clinical picture,² caused by demyelination of proprioceptive

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Table 1 Results of motor nerve conduction studies

Nerve	Distal ms (mV)	Proximal ms (mV)	Velocity m/s
Right median	18.9 (1.0)	29.0	21
Left ulnar	10.2 (0.6)	25.0	16
Right posterior tibial	34.3 (1.0)		
Left posterior tibial	24.3 (0.3)		

neurons. Romberg's sign will be positive and pseudoathetosis may be seen. The condition is then easily confused with other causes of sensory ataxic neuropathy such as chronic idiopathic ataxic neuropathy, paraneoplastic neuropathy, anti-myelin-associated-glycoprotein neuropathy,³ and the neuropathy associated with Sjögren's syndrome. CIDP may be superimposed on diabetic neuropathy as a secondary immunological event.⁴ The condition is diagnosed clinically by the finding of peripheral nerve dysfunction affecting more than one limb for greater than two months together with hyporeflexia or areflexia. Cerebrospinal fluid protein is raised, and nerve conduction studies show loss of sensory conduction, multifocal slowing of motor conduction, motor conduction blocks and temporal dispersion of waveforms. The usual treatment of CIDP is high-dose steroids for at least

two weeks. A long-term maintenance dose is usually required to prevent relapse. Intravenous immunoglobulins (IVIg) for five days or plasma exchange are first-line treatment for certain groups—patients with severe disability on presentation; elderly patients (who tend to respond only slowly to steroids); and patients in whom high-dose steroids are contraindicated. At least half of the responders to IVIg require regular treatment, which may have to be as frequent as every four weeks. This treatment is expensive and needs continuing review.⁵

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