The clinical picture of a slowly progressive sensory and motor peripheral neuropathy with loss of tendon reflexes certainly fits the diagnosis (Burns 2004). The early sensory symptoms in the territory of individual peripheral nerves is compatible although they would also be consistent with a vasculitic neuropathy. The severe pain is unusual and, although not unknown in CIDP, is again more suggestive of vasculitic neuropathy. The occurrence of intermittent fever is not a recognized part of CIDP, although Dr Bodley-Scott’s own enquiries suggest that this may require further consideration.

The diagnosis depends critically on the nerve conduction studies and these, we are told, showed clear-cut evidence of demyelination, presumably marked slowing of nerve conduction, partial conduction block or dispersion of elicited compound muscle action potentials, or a combination of all three.

The negative sural nerve biopsy certainly does not rule out the diagnosis of CIDP as it is often unhelpful, showing either no change or non-specific axonal degeneration and loss of axons. Only about 25% of biopsies show the diagnostic hallmarks of endoneurial lymphocytic infiltration, macrophage-associated demyelination and ‘onion bulb’ formation.

His renal failure is unlikely to have contributed because it does not cause a demyelinating sensory and motor neuropathy, and has worsened following the institution of haemodialysis.

THE HETEROGENEITY OF CIDP

The cause of CIDP remains unknown but the inflammatory features in active lesions and usual response to immunotherapy of different types suggest an autoimmune pathogenesis. It affects about 2–7 per 100 000 population, is more common in men than women, and in the elderly. The antigens that are the targets of the supposed immune response are obscure.

CIDP has now been subdivided into different subtypes, the usual symmetrical type which is Dr Bodley-Scott’s likely diagnosis, pure sensory and pure motor CIDP, and a multifocal sensory and motor type first described by Lewis and Sumner and often called after them (being an inveterate enemy of eponyms I prefer the name multifocal acquired demyelinating sensory and motor neuropathy – MADSAM). Different again is multifocal motor neuropathy in which nerve conduction remains normal to conventional examination between sites of conduction block.

Some patients with CIDP have paraproteins: in some these may be coincidental while in others...
they mark specific syndromes of which the commonest is the progressive sensory predominant demyelinating neuropathy with IgMκ paraprotein and antibodies to myelin-associated glycoprotein. CIDP sometimes occurs in association with other diseases, including vasculitic disorders.

THE RESPONSE TO RITUXIMAB

It is noteworthy that Dr Bodley-Scott had not responded adequately to corticosteroids, intravenous immunoglobulin, plasma exchange, ciclosporin and mycophenolate but he did respond to rituximab, an anti-CD20 monoclonal antibody which is useful in the treatment of B-cell lymphoma. There are reports in the literature of the treatment of over 30 patients with inflammatory neuropathy of different types with rituximab, but none with CIDP. Pestronk described 21 patients, 14 with multifocal motor neuropathy and IgM antibodies to ganglioside GM1 and seven with paraproteinemic demyelinating neuropathy and antibodies to myelin-associated glycoprotein (Pestronk et al. 2003). The group as a whole showed significantly more improvement in muscle strength than untreated historical controls over one and two year epochs. The two different groups were not presented separately. Renaud et al. reported nine patients with paraprotein-associated demyelinating neuropathy of whom six improved on a neuropathy impairment scale but the improvements were usually only modest (Renaud et al. 2003). There are several other smaller series, including one that described a patient who got much worse on rituximab (Rojas-Garcia et al. 2003). None of these series described treatment of patients with straightforward CIDP.

Dr Bodley-Scott’s vivid description of his own illness is a compulsive read in its own right and one can but admire his resourcefulness in the face of adversity. I think he must have his own variety of CIDP, with neuropathic pain and fever, which responded to rituximab.

REFERENCES


