Severe vasculitic neuropathy in systemic lupus erythematosus and response to cyclophosphamide

Neuropathy may be caused by illnesses producing systemic vasculitis1 including polyanerteritis nodosa, rheumatoid arthritis, Wegener’s granulomatosis, various forms of granulomatosis and also a more benign non-systemic vasculitic disorder: severe vasculitic neuropathy due to systemic lupus erythematosus (SLE) appears to be very rare.2 We report a case in which the neuropathy occurred in an identical clinical feature of a relapse of SLE and which responded to pulses of cyclophosphamide.

Over a period of four weeks, a 24 year old woman, of mixed Philippino and West Indian race, developed night sweats, fever, numbness ascending to the face and substantial weight loss, which was followed by proximal rala and distal retention of sensation. She could walk with the aid of one assistant after six weeks, and independently after six months with a walking stick. She was unable to continue on azathioprine and a reducing dose of prednisolone. On review one year later, she had marked wasting and weakness in the muscles of her hands, foot and anterior tibial compartment. Her maximum power was normal. There was residual loss of proprioception and cutaneous sensation in her fingers and left foot. On repeat electro-physiology, compound motor action potentials and sensory action potentials were again absent or greatly diminished. Conduction velocities were normal or mildly reduced. Electromyography showed signs of denervation in all muscles tested, including the ulnar and radial nerves and evidence of reinervation in all except distal muscles.

Neuropathies in SLE may present as a multiple mononeuropathy, sometimes with a vasculitic pathophysiological pattern, and as polyneuropathy, as in this case. The pathogenesis of this latter distribution may or may not be vasculitic.3 In our case the vasculitic nature was suggested by the polyphasic distribution and confirmed by sural nerve biopsy.

Nerve conduction studies revealed that compound motor action potentials and sensory action potentials were absent or greatly diminished in the legs and arms, but with normal conduction velocities. A sural nerve biopsy showed acute Wallerian degeneration of axons. One small epineurial vessel had fibrinoid material within its wall, and was also infiltrated by chronic inflammatory cells, as were almost all epineurial vessels. After 13 days of 40 mg/1 d prednisolone without any effect, five daily injections of 50 mg methylprednisolone were given but this had no effect on her clinical condition, fever, CRP or platelet count, though her white cell count and alamine transaminase did return to normal. Prednisolone was continued at 60 mg per day. Eight days after the methylprednisolone, cyclophosphamide (9 mg/kg) was given intravenously at weekly intervals for four weeks (each with Mesna cover), without any serious complication, being increased to 15 mg/kg orally. Her fever settled within 36 hours of the first cyclophosphamide dose, as did her CRP and platelet counts over the next few days. Her power began to improve within 12 hours of the first dose, closely followed by a proximal to distal return of sensation. She could walk with the aid of one assistant after six weeks, and independently after six months with a walking stick. She was unable to continue on azathioprine and a reducing dose of prednisolone. On review one year later, she had marked wasting and weakness in the muscles of her hands, foot and anterior tibial compartment. Her maximum power was normal. There was residual loss of proprioception and cutaneous sensation in her fingers and left foot. On repeat electro-physiology, compound motor action potentials and sensory action potentials were again absent or greatly diminished. Conduction velocities were normal or mildly reduced. Electromyography showed signs of denervation in all muscles tested, including the ulnar and radial nerves and evidence of reinervation in all except distal muscles.

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