CASE 1998 - 08

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Clinical History:

A 57-year-old man developed recurring and remitting symptoms of polyneuropathy beginning in the legs and then the arms. Family history was negative for neurological disease. Nerve conduction studies showed generalized demyelinating sensory motor neuropathy with motor conduction velocities of 23 and 18 m/s in the median and ulnar nerves respectively, and unobtainable sensory responses in the arms and legs. Investigations showed hypothyroidism, and replacement therapy did not influence the polyneuropathy. Biopsy of left sural nerve showed no inflammation, decreased number of fibers and onion bulbs. Records indicate presence of an IgM paraproteinemia which was not investigated further. Symptoms progressed slowly over the years with worsening numbness, increasing weakness of lower extremity and wide-based gait. A short trial of Prednisone treatment did not alter disease severity, however, the patient improved slightly over subsequent years spontaneously.

At the age of 69, examination revealed minimal weakness, postural tremor, generalized areflexia, loss of vibration sense and ataxia of lower extremities, and unsteady wide-based gait. Sensory responses were not obtainable. Motor conduction velocity was 15 m/s in the median nerve.

At the age of 72 years, the patient presented with fatigue, anemia, lymphadenopathy and inability to walk unassisted. Laboratory investigation revealed IgM/kappa paraprotein with a total IgM concentration of 54.5 gms per liter and relative plasma viscosity of 12.2. Lymph node and bone marrow biopsies revealed a diffuse small lymphocytic malignant lymphoma with a monoclonal plasmacytoid B cell population expressing immunoglobulin light chain kappa and immunoglobulin heavy chain IgM. Treatment with Chlorambucil and Prednisone was instituted with apparent remission of the lymphoma. Plasma exchanged was performed to treat hyperviscosity syndrome. The patient noted marked subjective improvement. A second nerve biopsy was performed 16 years after the onset of his original symptoms. The patient died three years later of disseminated lymphoma.

Material Submitted: Semithin sections stained with toluidine blue of left sural nerve at age 57 (lower left corner close to label) and right sural nerve at age 72 (upper right corner); and a lantern slide of an electron photo-micrograph.

Point of Discussion:

Diagnosis.