DSS 1990-2

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Clinical History:

This 62 year old man, previously well, was seen for a complaint of shortness of breath for which he received a diuretic. Shortly thereafter, he was referred to a Neurology Clinic with a complaint of radiating right arm and left leg pain. Spine films revealed degenerative disc disease and he was begun on corticosteroids with the presumed diagnosis of radiculopathies of right C8 and left L4. The following month the steroids were tapered. However, the pain worsened so the steroid were then increased for a period and then decreased and discontinued. A few weeks later he came to the Emergency Room with an apparent myelopathy. There was a positive Lhermitte's sign, a sensory level at T4-T6, incontinence, neck pain, marked leg weakness with bilateral radiating pain, and weakness of the intrinsic muscles of the hands. Evaluation for spinal cord compression included a negative MRI and a negative myelogram. Lumbar puncture showed a CSF protein of 64 with a normal glucose, 2 white cells and normal cytology. Electro-diagnostic testing revealed changes consistent with axonal neuropathy in a mononeuritis multiplex distribution.

Additional laboratory results included elevated LDH, IgM Kappa monoclonal gammopathy, light chains in the urine on protein electrophoresis and decreased platelets. The ANA titer was 1:160 with nucleolar pattern and decreased C3 and C4. Raji cell assay was positive but the sedimentation rate was normal. Anti double stranded DNA, and hepatitis serologies were normal. The patient underwent sural nerve biopsy.

Material Submitted: H&E section of sural nerve biopsy and unstained section

Points for Discussion: Diagnosis.