CASE 1997 #1

SUBMITTED BY: Dr. Juan M. Bilbao
Department of Pathology
St. Michael's Hospital
30 Bond Street
Toronto, Ontario
M5B 1W8, Canada

CLINICAL HISTORY:

This 70 year old woman developed at the age of 58 polyarticular arthritis, butterfly rash, sicca syndrome and was found to be ANA positive, coombs positive and ENA positive. The diagnosis of SLE was made and other investigations revealed IgG lambda paraprotein in serum and urine. A bone marrow biopsy showed 10% plasma cells. Skeletal survey was negative.

Plaquenil, 250 mg/day, was started for treatment of cutaneous manifestations of SLE. At the age of 63, hand tingling developed. At the age of 65, the patient's overall condition improved and all medication was discontinued. Following a flare of skin rash that year, chloroquine was started but symptoms progressed. With a regimen of Prednisone and Isoniazid, the patient's disease was suppressed.

At the age of 68, the patient complained of symmetrical numbness and tingling in hands and feet. Examination was normal and electrophysiological studies showed slightly reduced motor amplitudes in the legs. Symptoms progressed, weakness of foot dorsiflexion developed and reduced vibration and pin prick sensation in the feet were demonstrated. Deep tendon reflexes were decreased throughout. Ten months later, electrophysiological studies showed diffuse moderate to severe mixed axonal demyelinating polyneuropathy. The patient was lost to followup and re-examined at age 70 because of worsening of neuropathy. Three months later, at the time of sural nerve biopsy, the patient was on Plaquenil 250 mg/day, Prednisone 10 mg/day, Isoniazid 300 mg/day, and Pyridoxine 50 mg/day. A circulating paraprotein was still present, in concentration unchanged from earlier determinations. There was no evidence of active SLE.

Serial paraffin sections (x30) of sural nerve disclosed a focal collection of mature lymphocytes in epineurium, no immunostaining for light chains, no amyloid deposits and no vasculopathy.

MATERIAL SUBMITTED: Plastic resin section of sural nerve stained with Toluidin blue and one kodachrome of two electron micrographs: a myelinated nerve fiber and a smooth muscle cell.

POINTS FOR DISCUSSION:

1) Diagnosis
2) Pathogenesis