CASE 1997 #1

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CASE REFERENCE NUMBER: B3184 96-S-2111

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 (1983). 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 (Feb 1994) 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. In October 1994, electrophysiological studies showed diffuse moderate to severe mixed
axonal demyelinating polyneuropathy. The patient was lost to followup and re-examined in November
1995 because of worsening of neuropathy. In March 1996, 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