

Case 1993-6

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Clinical summary: This 49 year-old female, with a history of systemic lupus erythematosus, developed generalized edema and stomatitis, and was admitted to the hospital. A diagnosis of nephrotic syndrome was made, and therapy with high-dose Solumedrol was instituted. The stomatitis was severe, with oral ulcers and erythema involving the oral cavity and lips, fever with spikes to 104⁰F, and malaise; but this largely resolved and she was discharged on oral steroid therapy. An open renal biopsy, performed one month after the original admission, showed a membranous glomerulonephropathy with focal proliferative lesions. She returned one week later with continued drainage of serous fluid from her biopsy site, peripheral edema, fever and decreasing mental status. Muscle fasciculations progressed to rigidity with flexed arm posturing, and intravenous steroids were reinstated for suspected lupus cerebritis. Blood, urine and cerebrospinal fluid cultures were negative, and computed tomographic scans of her head showed only old lacunar infarcts bilaterally. A four-vessel arteriogram found no evidence of cerebritis. A brain biopsy was performed, which showed lesions similar to those later found at autopsy (see below). Cultures of biopsied brain tissue, including viral cultures, showed no growth.

Acyclovir therapy was added to broad-spectrum antibiotic therapy, but attempts to taper her steroid dosage were complicated by acute thrombocytopenia. Two years previously the patient had had a well-documented episode of thrombotic thrombocytopenic purpura which had been treated with steroids, but the present episode was accompanied by hypocellularity and decreased megakaryocytes in her bone marrow and absence of microangiopathic changes on peripheral smear. This resolved when high-dose steroids were restored. The patient continued to decline, developing a terminal Candidal sepsis, and expired 41 days following admission.

Postmortem findings: Postmortem examination showed no significant gross changes. Histological examination showed numerous small necrotizing lesions throughout the grey matter of the cerebral cortex, basal ganglia, midbrain, medulla, spinal cord and neurohypophysis. Somewhat similar lesions were found in the spleen, pancreas, liver and adrenal. The bone marrow showed mild hypocellularity. Cultures of brain tissue obtained at autopsy, including viral cultures, showed no growth.

Material submitted: Section of cerebral cortex, hematoxylin & eosin.

Points for discussion:

1. Diagnosis
2. Etiology