

CASE 9

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This 43 year-old male college graduate noted the insidious onset of difficulty in using his right hand, with deterioration of his handwriting. Six months later he had intermittent dizziness, vertigo, poor balance in walking and paresthesias of his right second and third fingers. Examination one year after onset showed no organic mental changes, normal strength and sensation, impaired alternate motion rate in the right hand, hyperreflexia of the right arm and leg, but no Babinski signs. In subsequent months he developed a short attention span, weakness of both upper extremities, awkwardness and posturing of both hands, and gait difficulty. An adverse seizure with automatisms was observed. *side ?*

An EEG showed severe generalized slowing. Spinal fluid, brain scan, left carotid arteriogram and pneumoencephalogram were normal. Blood pressure varied from 160/110 to 180/130. Urinalysis showed 3-4 + proteinuria, with up to 100 RBC per hpf and occasional granular and hyalin casts. BUN was 39 mg.%, creatinine 2.0 mg.% and uric acid 9.9 mg.%. An IVP and a radioisotope renogram were normal, but hippuran passage was slow bilaterally. An EMG showed denervation changes along the right median nerve. ?

Subsequently the patient was intermittently confused and euphoric, with severe memory impairment. Position sense in fingers and toes was absent, but pin-prick and touch were intact. His course was one of progressive deterioration, first intellectually, then in motor performance, until he was unable to read, sit or care for his personal needs. His speech became unintelligible. He had difficulty in swallowing, developed pneumonia and died at 46, three years after the onset of his first neurological symptoms.

Necropsy: Old and recent bland vegetations involving both aortic and mitral valves. Small old ischemic foci in heart, kidney, spleen, lymph nodes, adrenals and pituitary, often with arterioles partially or completely occluded by thrombi in varying stages of organization. Splenic arterioles demonstrated "onion-skin" appearances. Glomeruli primarily showed membranous thickening. Many urate crystals in one renal pelvis and calyceal system. The cerebral hemispheres showed many meningeal and cortical arteriolar occlusions, with extensive cortical ischemic destruction, and secondary demyelination of the white matter. Brainstem, cerebellum and spinal cord were involved to a lesser degree.

Diagnoses: Collagen disease with valvular vegetations, widespread emboli, thromboses, and arteriolitis, consistent with lupus erythematosus.

Submitted: One H & E-stained and two unstained slides; also a gross Kodachrome.

Points for Discussion:

1. Nature of the cerebral lesions - - - embolic, thrombotic, arteriolitis? How to prove?
2. How frequently is the CNS involved in vasculitis and the collagen diseases? Why is it so often uninvolved?