Case 1996-1

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Clinical History: The patient was a 32 y.o. woman with a long standing disease beginning at age 15, admitted with a one-day history of dysarthria, gait ataxia, increased right-sided weakness, and a six week history of a 20 pound weight loss with post-prandial nausea.

At age 15, the patient had fatigue, arthralgia, and hypertension. At that time she had a biopsy of a subcutaneous nodule and a diagnosis was made, but the records and slides were not available. She was treated with steroids and did well until age 30, when she developed fatigue, arthralgia, intermittent diplopia, and Raynaud's phenomenon. She had an extensive workup that showed: high titer ANA with a speckled pattern, negative SM, SSA and SSB antigens, rheumatoid factor, normal C3, low C4, normal ESR, C-reactive protein, negative anti-JO-1, and no antiphospholipid antibodies. She had mild sclerodactyly, Raynaud's phenomenon, and livido reticularis. During the next two years she developed intermittent bilateral amaurosis fugax, intermittent diplopia, right-sided tinnitus, progressive weakness, arthralgia, and arthritis. She was on various steroids and aspirin. She was relatively stable with persistent right-sided weakness and hypertension.

At admission, imaging studies showed changes consistent with intermediate and remote infarcts. She was treated with high dose steroids; during the workup, she was found unarousable one morning, and imaging showed extensive intracerebral hemorrhage. She went rapidly downhill and died.

Necropsy Findings: Left thalamic and intraventricular hemorrhage, recent, with multiple recent and intermediate evolving infarcts.
Multiple organ involvement by same process as in brain; (kidneys, heart, stomach, spleen, small and large intestine, ovaries, skeletal muscle, lymph nodes, adrenal glands, and peripheral nerves.)

Material submitted: One (1) H&E stained slide of either cerebrum or midbrain;
One 2X2 Ektachrome slide of gross basilar vessels