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CASE 2002 - 7

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Clinical History: The 42 year-old female patient underwent resectosigmoidal resection for adenocarcinoma (pT3N0M0, G2); she had neither a recurrence nor metastases. Shortly thereafter, the patient developed neuropsychologic and sensorimotor deficits followed by seizures. Cerebrospinal fluid analyses showed increased mononuclear cell count and elevated protein. Neuroimaging revealed multiple contrast-enhancing supra- and infratentorial lesions (Kodachrome submitted). In the cerebellar biopsy specimen there were dense lymphocytic infiltrates partially involving the leptomeninges. Serological analyses for various infectious agents were all negative. Treatment with steroids, substituted later by alkylating agents, had a transient success. One year later, while kept under low dose chemotherapy, the patient developed a relapse of her neurologic deficits and novel skin lesions; skin biopsy revealed lymphocytic and necrobiotic panniculitis. Administration of immunoglobulins had some beneficial effects, but the patient's condition deteriorated with pancytopenia and loss of B cells, and she died at age 45 of cerebral hemorrhage.

Necropsy findings: There were multiple recent haematomas distributed over the entire central nervous system. Lymphocytic perivascular cuffs and multiple lymphocytic infiltrates were present in the central nervous system including cerebral grey and white matter, cerebellum, spinal cord and cranial nerves, in the peripheral nerve system and tubulointerstitially in the kidneys, while significant glomerulopathy was not observed. There were chronic periportal hepatitis, hyperplasia of bone marrow and splenomegaly with extramedullary haematopoiesis in a setting of red pulp hyperplasia, while splenic white pulp and nodal lymph follicles showed partial depletion consistent with acquired B cell deficiency.

Material submitted: Kodachrome of MRI, slide of cerebellar biopsy specimen (H&E)

Points for discussion: Differential diagnosis