Intravascular lymphomatosis

Formerly: (malignant) angioendotheliomatosis. A rare lymphoma with no solid mass in which malignant lymphoid cells are found in the lumen of small blood vessels in affected organs. CNS involvement is reported in most cases. Presentation is nonspecific: patients are often febrile and may present with progressive multifocal cerebrovascular events (including stroke or hemorrhage), spinal cord or nerve root symptoms including cauda equina syndrome, encephalopathy, or peripheral or cranial neuropathies [1].

Initial transient cerebral symptoms may mimic TIAs or seizures. The ESR is often elevated prior to the initiation of steroids. Lymphoma cells may be seen in the CSF.

Painful skin nodules or plaques occur in ≈ 10% of cases, generally involving the abdomen or lower extremities, and these cases may be diagnosed with a skin biopsy (differential diagnosis here includes angioendotheliomatosis, a benign capillary, and endothelial cell disorder). Otherwise, diagnosis often requires brain biopsy (open or stereotactic), in which involved areas on imaging studies are targeted. Pathology: malignant lymphoid cells distend and occlude small arteries, veins, and capillaries with little or no parenchymal extension [2].

Treatment with combination chemotherapy can result in long-term remission in some patients, but early diagnosis before permanent damage occurs is critical (diagnosis is rarely made pre-mortem).

1)
Glass J, Hochberg FH, Miller DC. Intravascular Lymphomatosis. A Systemic Disease with Neurologic Manifestations. Cancer. 1993; 71:3156– 3164
2)
Burger PC, Scheithauer BW, Vogel FS. Surgical Pathology of the Nervous System and Its Coverings. 4th ed. New York: Churchill Livingstone; 2002