50th ANNUAL DIAGNOSTIC SLIDE SESSION 2009

CASE 2009-1

Submitted by: Daniel Keith Harrison, M.D. (Fellow in Neuropathology) [1], Cheryl Ann Palmer, M.D. (Professor of Neuropathology and Neurology) [1], Yuki A. Hammers, M.D. (Staff Pathologist) [2]

[1]University of Alabama at Birmingham, Department of Pathology, Division of Neuropathology, PD6A 175, 619 19th Street South, Birmingham, AL 35249-4550
[2] The Children's Hospital of Alabama, Children's Hospital, Department of Pathology and Laboratory Medicine, 1600 7th Avenue South, Birmingham, AL 35233

Clinical History: This 9-year-old boy presented with a three-week history of headaches, nausea, vomiting and subsequent neck and back pain. The headaches were described as diffuse, worse when standing and unimproved with over-the-counter medications. He had no major medical problems during his early childhood and had been performing well in school until this episode. He had been followed in Dermatology clinic since infancy for multiple hairy nevi on his extremities, neck and back.

Cerebrospinal fluid laboratory findings at presentation showed 8 white blood cells (with a differential of 93% lymphocytes), 1 red blood cell, a protein of 23 mg per dL and a glucose of 19 mg per dL. The opening pressure of the lumbar puncture was 44 cm of H_2O. CSF cultures were negative. Computed tomography of the brain showed no acute intracranial abnormality. Two months later, he developed complex partial seizures and a series of magnetic resonance images showed progressive leptomeningeal enhancement with superficial parenchymal involvement. He underwent a left frontal brain biopsy and insertion of a ventriculoperitoneal shunt for elevated intracranial pressure.

Material submitted: 1 H&E-stained section of the left gyrus rectus.

Points for discussion: What is the diagnosis?