Case 2009-1
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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.

Cutaneous hairy nevi

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 H2O. 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
Diagnosis: Neurocutaneous melanosis with leptomeningeal and brain parenchymal melanocytic lesions favoring progression to malignant melanoma

Immunohistochemistry

- **HMB-45**
- **MITF**
- **MART-1**
- **S100**

**Comment:** Imaging in this case demonstrated involvement particularly in the region of the temporal lobe. The MIB-1 labeling index was 30 to 40%. This patient eventually died of disease, despite the use of chemotherapy.

**From the Presenter:** Neurocutaneous melanosis is a rare condition, and differentiation of leptomeningeal melanosis in the syndrome from frankly malignant melanoma can be difficult. A high proliferative rate and an invasive pattern of tumor into the brain parenchyma are useful criteria favoring melanoma. In the present case, the high proliferative rate and the presence of melanocytic cell clusters within neuropil are most consistent with a diagnosis of malignant melanoma in the clinical setting of neurocutaneous melanosis.

**References:**


**Note**: At the suggestion of Dr. Herbert Budka, made at the Diagnostic Slide Session, we have included some classical references on this entity:

