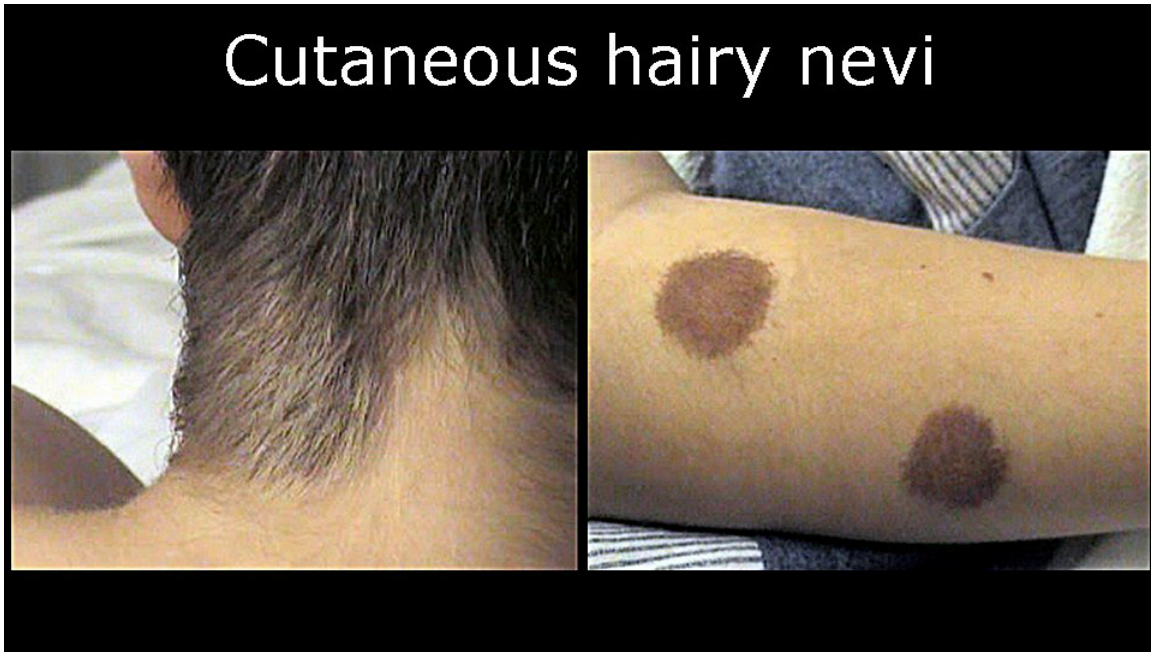
50th ANNUAL DIAGNOSTIC SLIDE SESSION, 2009
DIAGNOSES AND REFERENCES
MODERATOR: Anthony T. Yachnis, M.D.
EDITOR: Leroy R. Sharer, M.D.

Case 2009-1

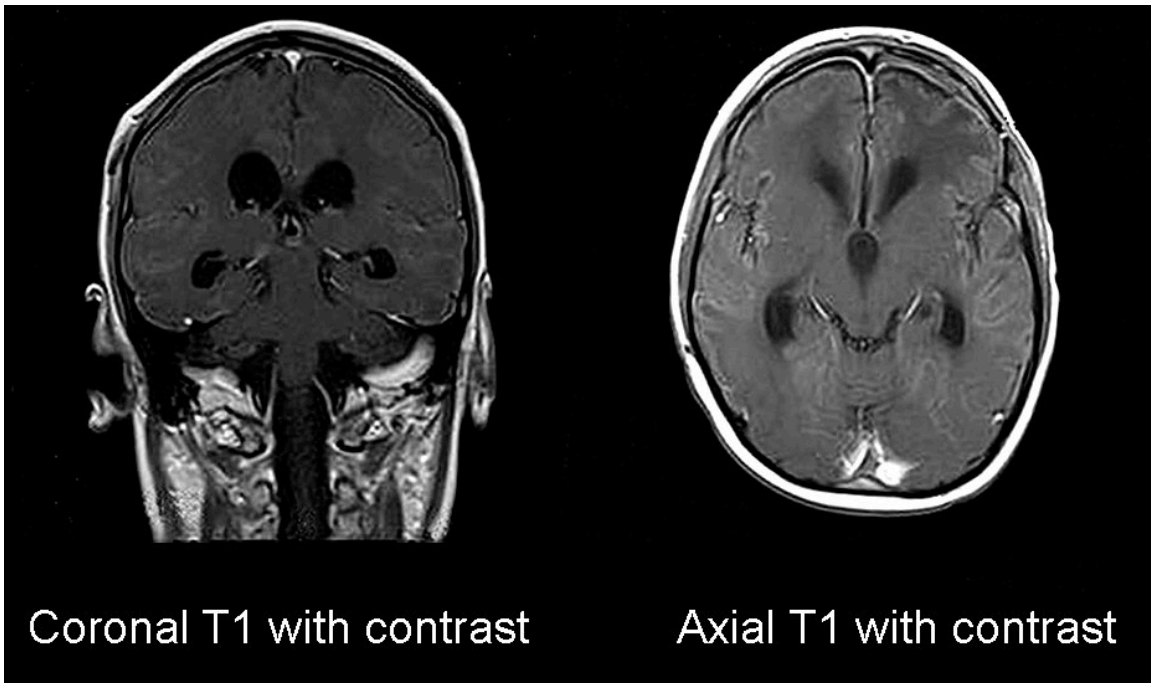
Submitted by: Daniel Keith Harrison, M.D., and Cheryl Ann Palmer, M.D., University of Alabama at Birmingham, and Yuki A. Hammers, M.D., The Children's Hospital of Alabama, Birmingham, AL.

Clinical History

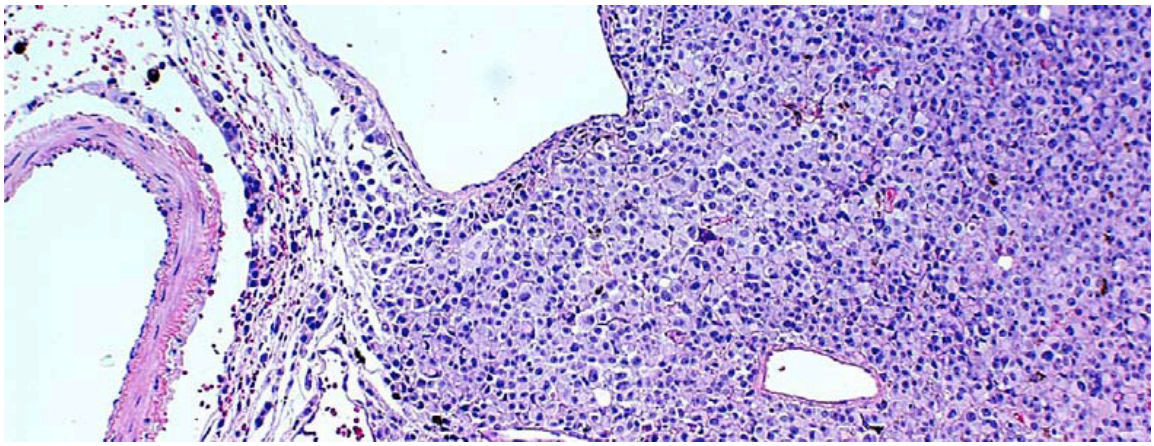
This 9-year-old boy presented with a three-week history of headaches, nausea, vomiting and subsequent neck and back pain. The headaches were described as diffuse, worse when standing and unimproved with over-the-counter medications. He had no major medical problems during his early childhood and had 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.

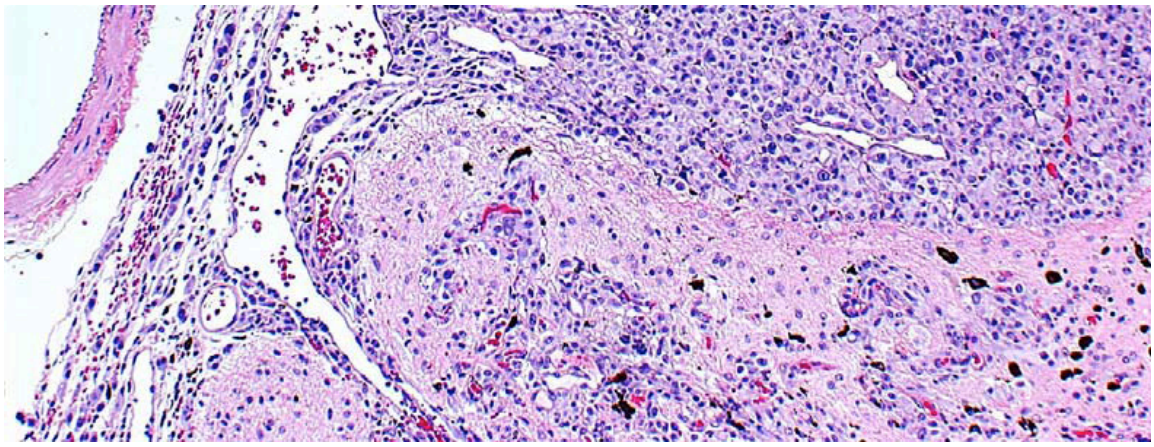


Cerebrospinal fluid laboratory findings at presentation showed 8 white blood cells (with a differential of 93% lymphocytes), 1 red blood cell, a protein of 23 mg per dL and a glucose of 19 mg per dL. The opening pressure of the lumbar puncture was 44 cm of H2O. CSF cultures were negative. Computed tomography 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.

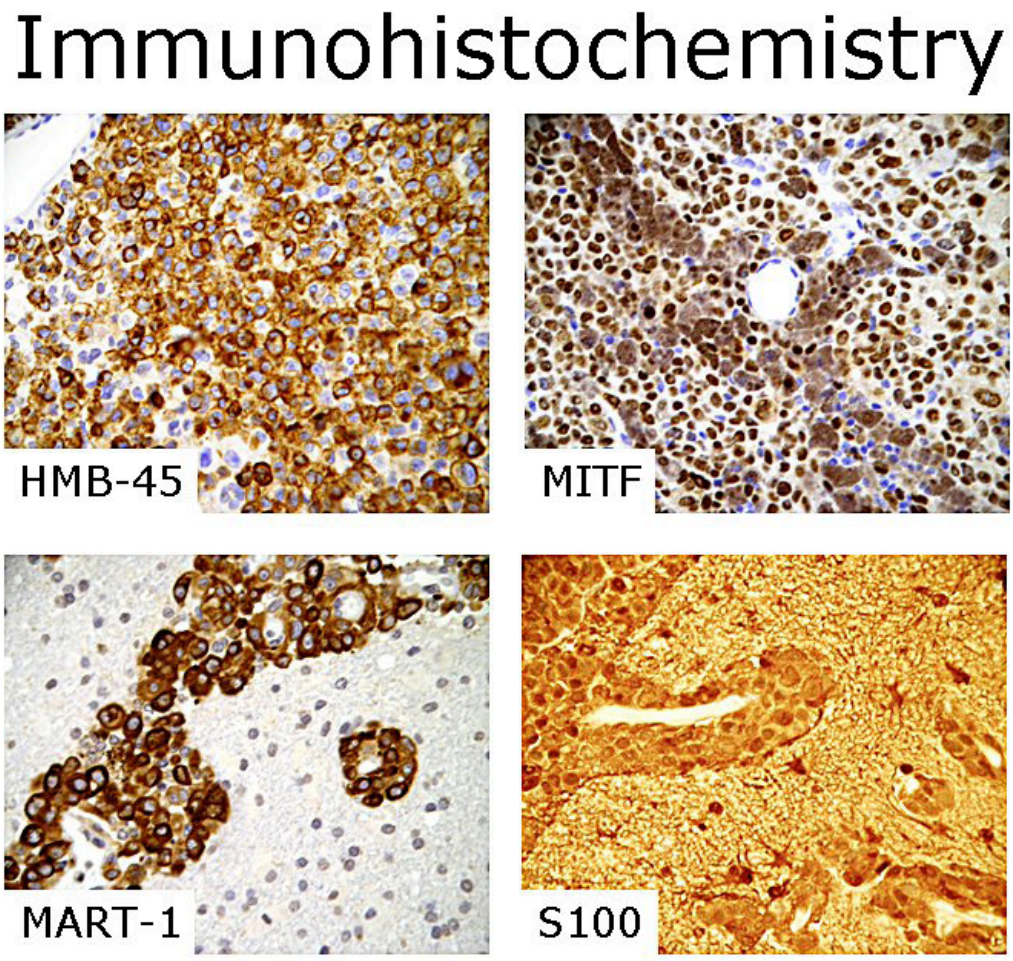


Material submitted: 1 H&E-stained section of the left gyrus rectus





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



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:

Burstein F, Seier H, Hudgins PA, Zapiach L: Neurocutaneous melanosis. J Craniofacial Surg 2005; 16:874-876.

Cajaiba MM, Benjamin D, Halaban R, Reyes-Múgica M. Metastatic peritoneal neurocutaneous melanocytosis. Am J Surg Pathol. 2008 Jan;32(1):156-61.

DeDavid M, Orlow SJ, Provost N, et al: A study of large congenital melanocytic nevi and associated malignant melanomas: review of cases in the New York University Registry and the world literature. J Am Acad Dermatol 1997; 36:409-416.

Di Rocco F, Sabatino G, Koutzoglou M, Battaglia D, Caldarelli M, Tamburrini G: Neurocutaneous melanosis. Childs Nerv Syst 2004; 20:23-28.

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

Rokitansky J: Ein ausgezeichneter Fall von Pigment-Mal mit ausgebreiteter Pigmentierung der inneren Hirn- und Rückenmarkshäute. Allgem Wien Med Z 1861; 6:113–116.

Virchow R: Pigment und diffuse Melanose der Arachnoides. Virchows Arch Pathol Anat 1859; 16:180.

Touraine A: La mélanoblastose neurocutanée. Presse médicale, 1941: 49:1087-1088.