**CASE 1998-1**

Submitted by: Rebecca D. Folkerth, M.D., Boston VA Medical Center, Boston, MA

**DIAGNOSIS:** Solitary fibrous tumor (7cm), arising in the cerebral ventricle, with atypical features, including mitoses (up to 3 per 10 high-power fields), and focal hypercellularity and nuclear pleomorphism; CD34 and O13 (CD99) positive, and S100, smooth muscle actin, and epithelial membrane antigen negative.

**Comment:** In discussion, the view was expressed that solitary fibrous tumor (SFT) and hemangiopericytoma (HPC) are different ends of a spectrum of tumors that clearly differ from meningioma. An opposing view was presented that SFT's are largely benign, in contradistinction to HPC.

**REFERENCES:**


**CASE 1998-2**

Submitted by: Drs. Barbara H. Amaker, M. Gary Hadfield, and Nitya R. Ghatak, Medical College of Virginia/Virginia Commonwealth University, Richmond, VA

**DIAGNOSIS:** Leiomyosarcoma in a child with AIDS, associated with EB virus infection.

**Comment:** EBV was detected by demonstrating nucleic acid sequences within the tumor. It was pointed out that routine immunohistochemistry for EBV, using the
standard antibody to latency membrane protein, is negative in these neoplasms, since this protein is not expressed.

REFERENCES:


CASE 1998-3

Submitted by: Drs. Waldemar Radziszewski, Pier-Luigi DiPatre, Alexander Brooks and Harry Vinters, UCLA Medical Center, Los Angeles, CA

DIAGNOSIS: Focal Mycobacterium avium/intracellulare (MAI) infection mimicking a meningioma.

Comment: The patient had been on prednisone, as treatment for SLE. Acid fast stain disclosed numerous organisms within the cells, and culture was also positive for MAI.

REFERENCES:


CASE 1998-4

Submitted by: Sozos Ch. Papasozomenos, M.D., University of Texas-Houston Medical School, Houston, TX

DIAGNOSIS: Langerhans Cell Histiocytosis.

Comment: The submitted electron micrograph demonstrated a Birbeck granule. The patient subsequently developed Langerhans cell histiocytosis involving the lung.

REFERENCES:

CASE 1998-5

Submitted by: Drs. Brent Harris and Dikran Horoupian, Stanford University Hospital, Stanford, and Dr. L.T. Smythe, Kaiser Hospital, Redwood City, CA

DIAGNOSIS: Extranodal sinus histiocytosis (Rosai-Dorfman disease).

REFERENCES:

CASE 1998-6

Submitted by: Drs. Edwin S. Monuki and Umberto de Girolami, Brigham and Women's Hospital and Children's Hospital, Boston, MA.

DIAGNOSIS: Granulomatous amebic encephalitis due to Balamuthia mandrillaris.
Comment: Dr. G.S. Visvesvara of the Parasitology Branch of the CDC identified the causative agent in this case by immunofluorescent staining of the tissue with a specific antibody for Balamuthia.

REFERENCES:


CASE 1998-7

Submitted by: Drs. Jiang Qian and Richard Prayson, Cleveland Clinic, Cleveland, OH.

DIAGNOSIS: Acute necrotizing myopathy of intensive care.

Comment: Electron microscopy revealed loss of myosin thick filaments. Patients with this disorder, particularly those with status asthmaticus, have usually been given a combination of corticosteroids and neuromuscular blocking agents. Other names for this disorder include critical illness myopathy, acute steroid myopathy, and myopathy of asthma.

REFERENCES:


CASE 1998-8

Submitted by: Drs. Juan M. Bilbao and Felix Tyndel and Ms. Sandra M. Cohen, St. Michael's Hospital, Toronto, Ontario, CANADA.

DIAGNOSIS: IgM (anti-MAG) paraprotein-associated polyneuropathy, with features of segmental demyelination, remyelination, onion bulbs, axonal changes and widely spaced myelin.

COMMENT: Approximately one half of patients with peripheral neuropathy and IgM monoclonal gammopathy have antibodies that bind to MAG. Immunogold labeling demonstrates localization of IgM to the separated myelin lamellae. About 17% of patients with monoclonal gammopathy and neuropathy develop a malignant lymphoproliferative disorder within 10 years, and 33% after 20 years.

REFERENCES:


CASE 1998-9

Submitted by: Professor Francesco Scaravilli, Institute of Neurology, Queen Square, London, UNITED KINGDOM.

DIAGNOSIS: Mitochondrial cytopathy (Kearns-Sayre).

Comment: Muscle biopsy during life disclosed mitochondrial myopathy. Mitochondrial DNA in this patient had tandem repeats 8 Kb in length, as well as focal deletions. The status spongiosus in the white matter in this case consisted of ovoid vacuoles that were parallel to the axons.
REFERENCES:


CASE 1998-10

Submitted by: Drs. Martha Quezada, Peter Bryant-Greenwood and Nancy Tresser, NINDS, NIH, Bethesda, MD.

DIAGNOSIS: Carney complex: Melanotic neuroectodermal tumor (psammomatous melanotic schwannoma).

Comment: The Carney complex is an autosomal dominant disorder with variable penetrance. It is not entirely clear whether the current lesion is a schwannoma or a melanoma.

REFERENCE: