Case 2000-1

Submitted by: Richard L. Davis, M.D., and Jane Uyehara-Lock, M.D., University of California, San Francisco, CA

Diagnosis: Ependymomatosis, involving the spinal subarachnoid space, with multifocal intracranial involvement

Comment: The cells were positive on immunocytochemistry (ICC) for glial fibrillary acidic protein (GFAP), and they were also positive for transthyretin (pre-albumin). Rare nuclei were positive for the MIB-1 clone of the proliferative marker Ki-67. It was felt by everyone that there was no relationship of this lesion to cystic fibrosis, which the patient also had.

References:


Case 2000-2

Submitted by: Michael A Stier, M.D., Humaira Khanam, M.D., Scott R. VandenBerg, M.D., M. Beatriz Lopes, M.D., University of Virginia, Charlottesville, VA; and Sérgio Rosemberg, M.D., and Paulo H. Aguiar, M.D., University of São Paulo School of Medicine, São Paulo, BRAZIL

Diagnosis: Solitary fibrous tumor
Comment: Several alternative diagnoses were suggested, including mesenchymal perineurioma of the choroid plexus and hemangiopericytoma. The lesion was positive on ICC for CD34, CD99, vimentin, and Bcl-2.

References:


Case 2000-3

Submitted by: Philip J. Boyer, M.D., Ph.D., David W. Allen, M.D., and Javed Towfighi, M.D., Penn State University-Hershey Medical Center, Hershey, PA; and Peter J. Christ, M.D., and Robert R. Eckert, D.O., St. Joseph Medical Center, Reading, PA

Diagnosis: Myxoid spindle cell neoplasm of low to intermediate malignant potential

Comment: Again, there was a great range of diagnoses, including metaplastic meningioma (myxoid variant), neurofibroma, myxoid malignant fibrous histiocytoma (MFH), and low grade sarcoma. The tumor cells were positive for PAS and Alcian blue, and on ICC for vimentin, but they were negative for everything else, including epithelial membrane antigen (EMA).

References:


Nagatani M et al. Primary myxoma in the posterior fossa: case report. Neurosurgery 1987; 20:329-331. (Letter to editor [Branch CL, Neurosurgery 1987; 21:130] criticizes the clinical evaluation of the heart in this case and suggests that cardiac angiography is needed in addition to echocardiography to rule out cardiac myxoma.)


**Case 2000-4**

Submitted by: Sandra L. Cottingham, M.D., Ph.D., Stephen D. Cohle, M.D., and Susan Millet, Spectrum Health East Campus, Grand Rapids, MI

Diagnosis: Meningioangiomatosis

Comment: The lesion, which involved the cortex and the subarachnoid space, also focally involved the white matter. There were also neurofibrillary degeneration and granulovacuolar degeneration in neurons. This lesion, which can be associated with NF2, can also be an incidental finding at autopsy, and some questioned whether it was related to the patient’s death. It was mentioned that this lesion also occurs in cats.

References:


**Case 2000-5**

Submitted by: Arnulf H. Koeppen, M.D., and Jiang Qian, M.D., Ph.D., Albany Medical College, Albany, NY

Diagnosis: Gliomatosis cerebri

Comment: The lesion was quite widespread, involving the molecular layer and white matter of the cerebellum and also the CA1 region of the hippocampus. Many of the cells have elongated, rod-like nuclei. These cells are positive on ICC for CD45 and also with the lectin RCA-1, but they are negative for HAM-56, raising a question as to whether they
are microglial cells or not. In this case, GFAP and CD45 staining did not coincide, on confocal microscopy, and GFAP staining in general was disappointing.

References:


Case 2000-6

Submitted by: Dennis Dickson, M.D., Mayo Clinic Jacksonville, Jacksonville, FL

Diagnosis: Non-paraneoplastic (idiopathic) limbic encephalitis

Comment: No occult neoplasm was discovered at autopsy. Several observers stated that chronic herpes simplex encephalitis needed to be ruled out, and this might require the use of PCR.

References:


Case 2000-7

Submitted by: Alexandra I. Brower, D. V. M., and Barbara Crain, M.D., Ph.D., Johns Hopkins School of Medicine, Baltimore, MD

Diagnosis: Lymphoplasmacytic meningoencephalitis secondary to trematode (Nasitrema sp.) parasite migration

Comment: Histologic findings (from the presenter): "Within the meninges are numerous refractile brown-yellow triangular parasite eggs, which range from 50 to 70 microns in diameter. Most of the parasite eggs are not viable, but a few are embryonated. Multinucleate giant cells surround many of the ova. Extending into the brain parenchyma are
multiple necrotic tracts with associated edema, mild gliosis, and intralesional parasite eggs. There is a mild to moderate multifocal perivascular lymphoplasmacytosis.

The parasite normally resides in the cranial air sinuses, but it can migrate around the eighth cranial (vestibular) nerve, which is present on the submitted section. This results in vestibular dysfunction, causing “stranding.” Some noted an occlusive vasculitis, with infarcts in the brainstem. A curious layer of cells in the subpial zone is apparently a normal finding in the brainstem in this species.

References:


Case 2000-8

Submitted by: Robert E. Mrak, M.D., Ph.D., University of Arkansas, Little Rock, AR

Diagnosis: Spinal cord infarction resulting from fibrocartilaginous embolization

Comment: The emboli are present in vessels in the leptomeninges, and they were present on all slides submitted. This condition was probably related to trauma in this patient, and it has been associated with many predisposing physical conditions. This phenomenon also occurs spontaneously in dogs. The entire cord was necrotic in the most severely involved segments, while the section submitted was from the margin of the infarct.

References:


Case 2000-9

Submitted by: Leroy R. Sharer, M.D., and Ivan Dressner, M.D., New Jersey Medical School, Newark, NJ

Diagnosis: Adult polyglucosan body disease involving peripheral nerve, in association with the Tyr^{326}Ser mutation in the glycogen-branching enzyme gene

Comment: The patient was of Ashkenazi Jewish descent, and this mutation has been described in this ethnic group. The patient was homozygous for the mutation. The polyglucosan bodies were PAS and Alcian blue positive, and they were filamentous on ultrastructural examination. Most of the reported cases have dementia, which was not clinically apparent in this patient.

References:


Case 2000-10

Submitted by: Juan M. Bilbao, M.D., St. Michael’s Hospital, Toronto, Ontario, CANADA

Diagnosis: Myopathy with tubular aggregates

Comment: Several alternative diagnoses were entertained, including oculopharyngeal dystrophy, inclusion body myopathy, and nemaline myopathy. There is predominantly perinuclear, granular, basophilic material in type 1 and type 2 fibers. Tubular aggregates were seen on electron microscopy. The biopsy also had type 1 fiber predominance, a change that was also seen in the biopsy done at age 11 years, as well as fiber necrosis and phagocytosis.

Reference: