39th ANNUAL DIAGNOSTIC SLIDE SESSION
1998
REFERENCES AND DIAGNOSES

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:

1. Van de Rijin M, Lombard CN, Rouse RV. Expression of CD34 by solitary fibrous tumors of the pleura, mediastinum, and lung. Am J Surg Path 1994; 18:814-20.

2. Fukunaga M, Naganuma H, Nikaido T, Harada T, Ushigome S. Extrapleural solitary fibrous tumor: a report of seven cases. Mod Pathol 1997; 10:443-50.

3. Prayson RA, McMahon JT, Barnett GH. Solitary fibrous tumor of the meninges. Case report and review of the literature. J Neurosurg 1997; 86:1049-52.

4. Malek AM, Weller SJ, Price DL Jr, Madsen JR. Solitary fibrous tumor presenting as a symptomatic intraspinal mass: case report. Neurosurgery 1997; 40:844-7.

5. Natarajan S, Morgello S. CNS solitary fibrous tumors lacking dural attachment (abstract). J Neuropathol Exp Neurol 1998; 57:526.

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:

1. McClain KL, Leach CT, Jenson HB, Joshi VV, Pollock BH, Parmley RT, DiCarlo FJ, Chadwick EG, Murphy SB. Association of Epstein-Barr virus with leiomyosarcomas in young people with AIDS. N Engl J Med 1995; 332:12-18.

2. Creager AJ, Maia DM, Funkhouser WK. Epstein-Barr virus-associated renal smooth muscle neoplasm: report of a case with review of the literature. Arch Pathol Lab Med 1998; 122:277-281.

3. Morgello S, Kotsianti A, Gumprecht SP, et al. Epstein-Barr virus-associated dural leiomyosarcoma in a man infected with human immunodeficiency virus: case report. J Neurosurg 1997; 86:883-887.

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:

1. Gyure KA, et al. Symptomatic Mycobacterium avium complex infection of the central nervous system. Arch Pathol Lab Med 1995; 119:836-839.

2. Umlas J, et al. Spindle cell pseudotumor due to Mycobacterium avium-intracellulare in patients with AIDS. Am J Sug Path 1991; 15:1182-1187.

3. Baksh FK, Handler MS. Central nervous system Mycobacterium avium complex infection (letter to editor). Arch Pathol Lab Med 1996; 120:613.

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:

1. Lieberman PH, Jones CR, Steinman RM, et al. Langerhans cell (eosinophilic) granulomatosis. A clinicopathologic study encompassing 50 years. Am J Surg Path 1996; 20:519-552.

2. Wilman CL, Busque L, Griffith BB, et al. Langerhans-cell histiocytosis (histiocytosis X) - A clonal proliferative disease. N Engl J Med 1994; 331:154-60.

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:

1. Deodhare SS, Ang LC, Bilbao JM. Isolated intracranial involvement in Rosai-Dorfman disease. Arch Pathol Lab Med 1998; 122:161-165.

2. Resnick DK, Johnson BL, Lovely TJ. Rosai-Dorfman disease presenting with multiple orbital and intracranial masses. Acta Neuropathol 1996; 91:554-557.

3. Panicker NK, Sabhikhi AK, Rai R. Rosai-Dorfman disease presenting as a meningioma. Indian J Cancer 1996; 33:192-194.

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:

1. Martinez AJ, Visvesvara GS. Free-living, amphizoic and opportunistic amebas. Brain Pathol 1997; 7:583-598.

2. Janitschke K, Martinez AJ, Visvesvara GS, Schuster F. Animal model of *Balamuthia mandrillaris* CNS infection: contrast and comparison in immunodeficient and immunocompetent mice: a murine model of "granulomatous" amebic encephalitis. J Neuropathol Exp Neurol 1996; 55:815-821.

3. Visvesvara GS, Schuster FL, Martinez AJ. *Balamuthia mandrillaris*, N.G., N. Sp., agent of amebic meningoencephalitis in humans and other animals. J Eukaryotic Microbiol 1993; 40:504-514.

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:

1. Hanson P, et al. Acute corticosteroid myopathy in intensive care patients. Muscle Nerve 1997; 20:1371-1380.

2. Paps EC, Bird SJ, Hansen-Flaschen J. Prolonged muscle weakness after neuromuscular blockade in the intensive care unit. Critical Care Clinics 1994; 10:799-813.

3. Ramsay DA, et al. A syndrome of acute severe muscle necrosis in intensive care unit patients. J Neuropathol Exp Neurol 1993; 52:387-398.

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:

1. Kyle RA. Monoclonal proteins in neuropathy. Neurology Clinics. Peripheral Neuropathy: New Concepts and Treatment, Vol. 10, No.3, August, 1992.

2. Midroni G, Bilbao JM. Biopsy Diagnosis of Peripheral Neuropathy, Chapter 14; Dysproteinemic neuropathy, pp. 263-282. Butterworth-Heinemann, 1995.

3. Chassande B, Leger JM, Younes-Chennoufi AB, Bengoufa D, Maisonbe T, Bouche P, Baumann N. Peripheral neuropathy associated with IgM monoclonal gammopathy. Muscle Nerve 1998; 21:55-62.

4. Ropper AH, Gorson KC. Current concepts: Neuropathies associated with paraproteinemia. N Engl J Med 1998; 338:1601-1607.

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:

1. Sparaco M, Bonilla E, DiMauro S, Powers JM. Neuropathology of mitochondrial encephalomyopathies due to mitochondrial DNA defects. J Neuropathol Exp Neurol 1993; 52:1-10.

2. Ohno K, Yamamoto M, Engel AG, Harper CM, Roberts LR, Tan GH, Fatourechi V. MELAS- and Kearns-Sayre-type co-mutation [corrected] with myopathy and autoimmune polyendocrinopathy. Ann Neurol. 1996; 39:761-6.

3. Brockington M, Alsanjari N, Sweeney MG, Morgan-Hughes JA, Scaravilli F, Harding AE. Kearns-Sayre syndrome associated with mitochondrial DNA deletion or duplication: a molecular genetic and pathological study. J Neurol Sci 1995; 131:78-87.

CASE 1998-10

Submitted by: Drs. Martha Quezado, 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:

1. Carney, JA. Carney complex: the complex of myxomas, spotty pigmentation, endocrine overactivity, and Schwannomas. Seminars in Dermatology 1995; 14:90-98.