37th ANNUAL DIAGNOSTIC SLIDE SESSION

1996

CASE 1996-1

Submitted by: Eric Huang, M.D., Ph.D., and Richard L. Davis, M.D., UCSF

Diagnosis: Polyarteritis nodosa

Comment: The systemic arteries were also involved at post mortem, except for those of the lung.

References:

Graf WD et al.: Stroke and mixed connective tissue disease. J. Child Neurol 8:256-259, 1993.

Greenan TJ et al.: Cerebral vasculitis: MR imaging and angiographic correlation. Radiology 182:65-72, 1992.

Harris KG et al.: Diagnosing intracranial vasculitis: the roles of MR and angiography. AJNR Am J Neurorad 15:317-330, 1994.

Hurst RW, Grossman RI: Neuroradiology of central nervous system vasculitis. Sem Neurol 14(4):320-340, 1994.

Moore PM, Cupps TR: Neurological complications of vasculitis. Ann Neurol 14:155-167, 1983.

Moore PM, Fauci AS: Neurologic manifestations of systemic vasculitis: a retrospective and prospective study of clinicopathologic features and responses to therapy in 25 patients. Am J Med 71:517-524, 1981.

Rosenberg MR et al.: Central nervous system polyarteritis nodosa. Western J Med 153:553-556, 1990.

Sigal LH: The neurologic presentation of vasculitic and rheumatologic syndromes: a review. Medicine 66(3):157-180, 1987.

CASE 1996-2

Submitted By: Cheryl Ann Palmer, M.D., University of Alabama at Birmingham

Diagnosis: Paraneoplastic (limbic) encephalitis

Comment: At autopsy, there was metastatic squamous cell carcinoma in lymph nodes. A primary tumor was not found. Serum studies during life for anti-Hu antibodies were negative,

but they were not done in CSF. Virus antigens were negative in brain, by immunocytochemistry. Lesions (microglial nodules, perivascular lymphocytes) were also present in brainstem and spinal cord.

References:

Brierley JB, Corsellis JAN, Hierons R, Nevin S: Subacute encephalitis of later adult life, mainly affecting the limbic areas. Brain 83(3):357-370, 1960.

Henson RA, Hoffman HL, Urich H: Encephalomyelitis with carcinoma. Brain 88:449-468, 1965.

Corsellis JAN, Goldberg GJ, Norton AR: "Limbic encephalitis" and its association with carcinoma. Brain 91:481-496, 1968.

Glaser GH, Pincus JH: Limbic encephalitis. J Nervous Mental Disease 149(1):59-67, 1969.

Brennan LV, Craddock PR: Limbic encephalopathy as a nonmetastatic complication of oat cell lung cancer: its reversal after treatment of the primary lung lesion. Am J Med 75:518-520, 1983.

Kodama T, Numaguchi Y, Gellad FE, Dwyer BA, Kristt DA: Magnetic resonance imaging of limbic encephalitis. Neuroradiology 33:520-523, 1991.

Graus F, René R : Clinical and pathological advances on central nervous system paraneoplastic syndromes. Rev Neurol 148:496-501, 1992.

Glantz MJ, Biran H, Myers ME, Gockerman JP, Friedberg MH: The radiographic diagnosis and treatment of paraneoplastic central nervous system disease. Cancer 73(1):168-175, 1994.

CASE 1996-3

Submitted by: Brian Harding, B.M., D.Phil., FRCPath, Great Ormond Street Hospital for Children, London

Diagnosis: Alpha-fucosidosis

Comment: Appearances in this disorder similar to Alexander's leukodystrophy have been documented on two previous occasions. The patient's younger brother had a bone marrow transplant at age 7 months, with normalization of enzyme levels; the brother remains well at age four years.

References:

Labrisseau A, Brochu P, Jasmin G: Fucosidosis de Type 1. Étude anatomique. Arch Fr Pédiatr 36:1013-25, 1979.

Garcia CP, McGarry PA, Duncan CM: Fucosidosis and Alexander's leukodystrophy. J. Neuropathol Exp Neurol 39:353, 1980.

Lake BD: Lysosomal and peroxisomal disorders. In: Greenfield's Neuropathology, 5th ed., Adams JH, Duchen LW, eds, Edward Arnold, London, 1992, pp. 709-810.

CASE 1996-4

Submitted by: Thomas C. Cannon, M.D., Richard W. Leech, M.D., and Roger A. Brumback, M.D., University of Oklahoma

Diagnoses: Alzheimer's disease, congophilic amyloid angiopathy, laminar cortical necrosis, and subcortical system degeneration, with involvement of basal ganglia, thalamus, and cerebral white matter

Comment: The audience mostly favored a diagnosis of Creutzfeldt-Jakob (prion) disease of unusual severity, with Alzheimer's disease.

Reference:

Carota et al.: A panencephalopathic type of Creutzfeldt-Jakob disease with subacute lesions of the thalamic nuclei in two Swiss patients. Clin Neuropathol 15:125-134, 1996.

CASE 1996-5

Submitted by: Hans H. Goebel, M.D., Johannes Gutenberg-Universität, Mainz

Diagnosis: Hypoplasia of nerve fascicles (nerve) owing to aplasia of myelinated axons

References:

Thomas PK: Hereditary sensory neuropathies. Brain Pathol 3:157-164, 1993.

Schröder JM, Heide G, Ramaekers V, Mortier W: Subtotal aplasia of myelinated nerve fibers in sural nerve. Neuropediatrics 24:286-291, 1993.

Ferrière G, Guzzetta F, Kulakowski S, Evrard P: Nonprogressive type II hereditary sensory autonomic neuropathy: a homogeneous clinicopathological entity. J Child Neurol 7:364-370, 1992.

CASE 1996-6

Submitted by: Caterina Giannini and Joseph E. Parisi, Mayo Clinic

Diagnosis: Erdheim-Chester Disease

Comment: The vertebral artery lesion was a xanthoma. The disorder in this patient was generalized, involving the long bones, lung and retroperitoneum. S-100 immunoreactivity in the histiocytic cells was negative, a distinguishing feature from histiocytosis X.

References:

Chester W: Über lipoidgranulomatose. Virchows Arch [A] 279:561-602, 1930.

Pertuiset E, Laredo JD, Liote' D, et al.: Erdheim-Chester disease: report of a case, review of the literature, and discussion of relationships with Langerhans cell histiocytosis. Rev Rheum (Engl Ed) 60:504-11, 1993.

Fink MG, Levinson DJ, Brown NL et al.: Erdheim-Chester disease. Case report with autopsy findings. Arch Pathol Lab Med 115:619-23, 1991.

Fukazawa T, Tsukishima E, Sasaki H et al.: Erdheim-Chester disease and slowly progressive cerebellar dysfunction. J Neurol Neurosurg Psych 58:238-40, 1995.

CASE 1996-7

Submitted by: Dr. J.M. Bilbao, St. Michael's Hospital, Toronto

Diagnosis: Primary embryonal rhabdomyosarcoma of cerebrum in an adult

Comment: Cells are positive for myoglobin and desmin. The inclusions are cytoplasmic bodies, as seen in skeletal muscle. The patient's father recently developed a cerebral tumor, which was a giant cell glioblastoma, with many histiocytes.

References:

Dropcho EJ, Allen JC: Primary intracranial rhabdomyosarcoma: case report and literature review. J Neurooncol 5:139-150, 1987.

Russell DS, Rubinstein LJ: Pathology of Tumors of the Nervous System, 5th ed, Williams and Wilkins, Baltimore, 1989, p. 689.

Bradford R, Crockard, Isaacson PG: Primary rhabdomyosarcoma of the central nervous system: case report. Neurosurgery 17:101-104, 1985.

Olson JJ, Menezes AH, Godersky JC, Lobosky JM, Hart M: Primary intracranial rhabdomyosarcoma. Neurosurgery 17:25-34, 1985.

CASE 1996-8

Submitted by: Amyn M. Rojiani, M.D., Ph.D., University of Florida College of Medicine

Diagnosis: Malignant fibrous histiocytoma, pleomorphic/storiform variant, presenting as a second intracranial malignancy following chemotherapy

Comment: The tumor cells were positive for α-1-antichymotrypsin.

References:

Paulus W, Slowij F, Jellinger K: Primary intracranial sarcomas: histopathological features of 19 cases. Histopathology 18:395, 1991.

Enzinger F, Weiss S: Malignant fibrous histiocytoma. In: Soft Tissue Tumors, 3rd ed, Mosby-Year Book, 1995.

Smith LM, Donaldson SS: Secondary malignancies in patients with retinoblastoma and Ewing's sarcoma. Oncology-Huntingt 5(5):135, 1991.

Zarrabi MH, Rosner F: Second neoplasms in Hodgkin's disease: current controversies. Hematol Oncol Clin North Am 3:303-318, 1989.

CASE 1996-9

Submitted by: John J. Kepes, M.D., and Michael S. Handler, M.D., University of Kansas Medical Center (presented by Dr. E. T. Hedley-Whyte, for Dr. Kepes)

Diagnosis: Metastatic carcinoma from lung, with sarcomatoid metaplasia

References:

Humphrey, PA, Scroggs MW, Roggli VL, Schelburne JD: Pulmonary carcinomas with a sarcomatoid element: an immunocytochemical and ultrastructural analysis. Hum Pathol 19:155-165, 1988.

Colby TV, Koss NM, Travis WD: Tumors of the lower respiratory tract. AFIP Atlas of Tumor Pathology, 3rd Series, Fascicle 13, AFIP, Washington, D.C., 1995, pp 411-416.

Weiss SW, Brathauer GL, Morris PA: Postirradiation malignant fibrous histiocytoma expressing cytokeratin. Implications for the immunodiagnosis of sarcoma. Am J Surg Pathol 12:544-558, 1988.

Litzky LA, Brooks JJL: Cytokeratin immunoreactivity in malignant fibrous histiocytoma and spindle cell tumors: comparison between frozen and paraffin-embedded tissues. Modern Pathol 5:30-38, 1992.

Hirose T, Kudo E, Hasegawa T, Abe J-I, Hizawa K: Expression of intermediate filaments in malignant fibrous histiocytomas. Hum Pathol 20:871-877, 1989.

CASE 1996-10

Submitted by: Meena Gujrati, M.D., John M. Lee, M.D., Ph.D., Keith Izban, M.D., Chinnamma Thomas, M.D., Loyola University Medical Center, Maywood, Illinois

Diagnosis: Blastomycosis (meningoencephalitis)

Comment: The organism (*Blastomyces dermatitidis*) was cultured from the lesion in the fourth ventricle, at autopsy. North American blastomycosis has broad-based budding, and it causes a combination of acute and chronic inflammation, as seen in this case.

References:

Wheat J: Endemic mycoses in AIDS: a clinical review. Clin Microbiol Rev 8(1):146-159, 1995.

Witzig RS, Hoadley DJ, Greer DL, Abriola KP, Hernandez RL: Blastomycosis and human immunodeficiency virus: three new cases and review. Southern Med J 87(7):715-719, 1994.

Pappas PG, Pottage JC, et al.: Blastomycosis in patients with the acquired immuno-deficiency syndrome. Ann Int Med 116:847-853, 1992.

Roos KL, Bryan JP, Maggio WW, Jane JA, Scheld WM: Intracranial blastomycoma. Medicine 66(3):224-235, 1987.

Mirra SS, Trombley IK, Miles ML: Blastomycoma of the cerebellum. Acta Neuropathol 50:109-114, 1980.