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:


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


Corsellis JAN, Goldberg GJ, Norton AR: "Limbic encephalitis" and its association with


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

Graus F, René R: Clinical and pathological advances on central nervous system

Glantz MJ, Biran H, Myers ME, Gockerman JP, Friedberg MH: The radiographic diagnosis and

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


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:

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:
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:


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:


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:


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:


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:


