38th ANNUAL DIAGNOSTIC SLIDE SESSION

1997

CASE 1997-1

Submitted by: Dr. Juan M. Bilbao, St. Michael's Hospital, Toronto, Ontario, CANADA

Diagnosis: **Chloroquine neurotoxicity (neuropathy)**, in a patient with plasma cell dyscrasia, longstanding monoclonal gammopathy (IgG) and history of systemic lupus erythematosus treated with chloroquine.

Comment: Lamellar inclusions are seen within the myelinated fibers.

References:

Leger JM, Pnifouilloux H, Dancea S, Hauw JJ, Bouche P, Rougemont D, Laplane D: Neuromyopathies á le chloroquine: 4 cas á cours d'une prophylaxie anti-paludeene. Revue Neurologique 1986; 142:745-752.

Estes ML, Ewing-Wilson D, Chou SM, et al: Chloroquine neuromyotoxicity. Clinical and pathologic perspective. Am Jour Med 1987; 82:447-455.

Tegner R, Tomé FM, Godeau P, Lhermitte F, Fardeau M: Morphological study of peripheral nerve changes induced by chloroquine treatment. Acta Neuropathol 1988; 75:253-260.

CASE 1997-2

Submitted by: Drs. Yves Robitaille, Stéphane Ledoux, and Neil Cashman, Ste. Justine and Montreal Neurological Hospitals, Montreal, Québec, CANADA

Diagnosis: Intranuclear inclusion body disease, with atypical features

Comment: Some neuronal loss was apparent in the basal ganglia and substantia nigra, although not in the cerebral cortex.

References:

Michaud J, and Gilbert JJ: Multiple system atrophy with neuronal intranuclear hyaline inclusions: report of a new case with light and electron microscopic studies. Acta Neuropathol 1981; 54:113-119.

Palo J, Haltia M, Carpenter S et al: Neurofilament sub-unit-related proteins in neuronal intranuclear inclusions. Ann Neurol 1984; 15:322-328.

Tateishi J, Nagara H, Ohta M et al: Intranuclear inclusions in muscle, nervous tissue and adrenal gland. Acta Neuropathol 1984; 63:24-32.

Funata N, Maeda Y, Koike M et al: Neuronal intranuclear hyaline inclusion disease: report of a case and review of the literature. Clin Neuropathol 1990; 9:89-96.

Weidenheim K, Dickson DW: Intranuclear inclusion bodies in an elderly demented woman: a form of intranuclear inclusion body disease. Clin Neuropathol 1995; 14:93-99.

Case 1997-3

Submitted by: Drs. Maryam Mohammadkhani, Kathy L. Newell, and E. Tessa Hedley-Whyte, Massachusetts General Hospital, Boston, MA.

Diagnosis: Rupture of massive mitral annulus calcification with fatal systemic embolization

Comment: In retrospect, tiny calcifications could be appreciated on CT scan of the brain. Grossly at autopsy, the mitral annulus was chalky-white and fluid-like. There were emboli in multiple systemic organs.

References:

Lin CS, Schwartz IS, Chapman I: Calcification of mitral annulus fibrosus with systemic embolization: a clinicopathologic study of 16 cases. Arch Pathol Lab Med 1987; 111:411-414.

Benjamin EJ, Plehn JF, D'Agostino RB, et al: Mitral annular calcification and the risk of stroke in an elderly cohort. N Engl J Med 1992; 327:374-379.

Case 1997-4

Submitted by: Dr. Jeanne Bell, Western General Hospital, Edinburgh, UK.

Diagnosis: New variant Creutzfeldt-Jakob disease

Comment: The pathology is quite uniform throughout the cerebral cortex, and hence a

cerebral biopsy is likely to disclose the characteristic plaques with spongiform change. In addition, PrP^{res} can be demonstrated in biopsy of the tonsil.

References:

Budka H. et al: Tissue handling in suspected Creutzfeldt-Jakob disease (CJD) and other spongiform encephalopathies (prion disease). Brain Pathol 1995; 5:319-322.

Will RG, Ironside JW, Zeidler M, Cousens SN, Estibeiro K, Alperovitch A, Poser S, Pocchiari M, Hofman A, Smith PG: A new variant of Creutzfeldt-Jakob disease in the UK. Lancet 1996; 347:921-925.

Bell JE. Gentleman SM, Ironside JW, et al: Prion protein immunocytochemistry: UK five centre consensus report. Neuropathol Appl Neurobiol 1997; 23:26-35.

Case 1997-5

Submitted by: Drs. Hindi N. Al-Hindi and Clayton A. Wiley, Presbyterian University Hospital, Pittsburgh, PA, and Dr. William E. Ballinger Carolinas Medical Center, Charlotte, NC.

Diagnosis: Severe meningoencephalitis associated with JC virus and HIV infection

Comment: There is extensive necrosis. In atypical cases of this sort, it is usual that only rare cells can be demonstrated to contain JC virus. In addition, herniation has been reported to occur in cases of PML.

References:

Vazeux R, et al: Severe encephalitis resulting from coinfections with HIV and JC virus. Neurology 1990; 40:944-948.

Aksamit AJ, et al: AIDS-associated progressive multifocal leukoencephalopathy (PML): comparison to non-AIDS PML with in situ hybridization and immunohistochemistry. Neurology 1990; 40:1073-1078.

Hair LS, et al: Progressive multifocal leukoencephalopathy in patients with human immunodeficiency virus. Human Pathol 1992; 23:663-667.

Case 1997-6

Submitted by: Dr. Anthony T. Yachnis, University of Florida, Gainesville, FL.

Diagnosis: Microsporidiosis involving the central nervous system

Comment: Organisms can be well demonstrated by the use of plane polarized light. The actual organism in this case was *Trachipleistophora hominis*. Treatment is with albendazole.

References:

Yachnis AT, Berg J, Martinez-Salazar A, et al: Disseminated microsporidiosis especially infecting the brain, heart, and kidneys: report of a newly recognized pansporoblastic species in two symptomatic AIDS patients. Am J Clin Pathol 1996; 106:535-543.

Mertens RB, Didier ES, Fishbein et al: *Encephalitozoon cuniculi* microsporidiosis: Infection of the brain, heart, kidneys, trachea, adrenal glands, and urinary bladder in a patient with AIDS. Mod Pathol 1997; 10:68-77.

Weber R, Deplazes P, Flepp M, et al: Cerebral microsporidiosis due to *Encephalitozoon cuniculi* in a patient with human immunodeficiency virus infection. New Engl J Med 1997; 336:474-478.

Case 1997-7

Submitted by: Dr. Leila Chimelli, School of Medicine of Ribeirão Preto, BRAZIL

Diagnosis: Cerebral trypanosomiasis (Chagas disease).

Comment: The diagnosis of *Trypanosoma cruzii* infection was confirmed by immunocytochemistry.

References:

Gluckstein D, Ciferri F, Ruskin J: Chagas' disease: another cause of cerebral mass in the acquired immunodeficiency syndrome. Am J Med 1992; 92:429-432.

Rocha A, Meneses ACO, Silva AM, et al: Pathology of patients with Chagas' disease and acquired immunodeficiency syndrome. Am J Trop Med Hyg 1994; 50:261-268.

Chimelli L, Scaravilli F: Trypanosomiasis. Brain Pathol 1997; 7:599-611.

Case 1997-8

Submitted by: Drs. James M. Henry and Alan L. Morrison, Armed Forces Institute of Pathology, Washington, DC

Diagnosis: Rabies.

(Also, microabscess formations, multifocal, consistent with *Candida*.)

Comment: The hippocampal section was loaded with Negri bodies, but the cerebellar section had few Purkinje cells, making recognition of them more difficult. Lack of inflammation in the hippocampus is characteristic. Nucleotide sequence analysis of the rabies strain in this case was similar to that found in the Mexican freetailed bat.

References:

Earle KM, Dupont J-R: Negri bodies in human rabies. Texas Reports of Biology and Medicine 1966; 24:317-325.

Mrak RE, Young L: Rabies encephalitis in a patient with no history of exposure. Hum Pathol 1993; 24:109-110.

Mrak RE, Young L: Rabies encephalitis in humans: pathology, pathogenesis and pathophysiology. J Neuropathol Exp Neurol 1994; 53:1-10.

Esiri MM: Viruses and rickettsiae. Brain Pathol 1997; 7:695-709.

Case 1997-9

Submitted by: Drs. Brian Summers and Alexander de Lahunta, College of Veterinary Medicine, Cornell University, Ithaca, and Dr. John Speciale, Rochester, NY.

Diagnosis: Feline ischemic encephalopathy, a syndrome of cerebral infarction in the domestic cat recently associated with *Cuterebra* sp. myiasis.

Comment: This case is unusual, since the remains of the parasite (the fly *Cuterebra*) are still present. The disease in cats is most common in the summer months.

References:

Feline ischemic encephalopathy and feline CNS cuterebra:

Cook JR, Levesque DC, Nuehring LP: Intracranial Cuterebral myiasis causing acute lateralizing meningoencephalitis in two cats. Journal of the American Animal Hospital

Association 1985; 21:279-284.

Summers BA, Cummings JC, de Lahunta A: Veterinary Neuropathology. Mosby, 1995.

Cuterebra myiasis in man:

Baird JK, Baird CR, Sabrosky CW: North American cuterebrid myiasis: report of seventeen new infections of human beings and review of the disease. J Am Acad Dermatol 1989; 21:763-772.

Glasgow BJ, Maggiano JM: *Cuterebra* ophthalmomyiasis. Am J Ophthalmol 1995; 119:512-514.

Case 1997-10

Submitted by: Drs. Hans H. Goebel and J.K. Mellies, Mainz University, Mainz, and Municipal Hospital, Osnabruck, GERMANY

Diagnosis: CADASIL (cerebral autosomal-dominant arteriopathy with subcortical infarcts and leukoencephalopathy)

Comment: The specimen was a skin biopsy, with characteristic small granular inclusions in the basal lamina of blood vessels.

References:

Ruchoux M-M, Chabriat H, Bousser M-G, Baudrimont M, Tournier-Lasserve E: Presence of ultrastructural arterial lesions in muscle and skin vessels of patients with CADASIL. Stroke 1994; 25:2291 -2292.

Schröder JM, Sellhus B, Jörg J: Identification of the characteristic vascular changes in a sural nerve biopsy of a case with cerebral autosomal-dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL). Acta Neuropathol 1995; 89:116-121.

Goebel HH, Meyermann R, Rosin R, Schlote W: Characteristic morphologic manifestation of CADASIL, cerebral autosomal-dominant arteriopathy with subcortical infarcts and leukoencephalopathy, in skeletal muscle and skin. Muscle Nerve 1997; 20:625-627.