30TH ANNUAL DIAGNOSTIC SLIDE SESSION

CASE 1

The diagnosis was Hereditary Adult-Onset Leukodystropy. The condition is considered by Seitelbergeras a form of adult onset Pelizaeus-Merzbacher disease. Histologic similarities to Canavan's disease were mentioned by several observers.

Eldridge R, Anayiotos CP, Schlesinger S, Cowen D, Bever C, Patronas H, McFarland H. Hereditary adult-onset leukodystropy simulating chronic progressive multiple sclerosis. New Eng J Med 1984; 311:948-953.

Seitelberger F. Pelizaeus-Merzbacher disease. Vinken PJ, Bruyn GW, eds. In: Handbook of Clinical Neurology. Amsterdam, North Holland, 1970, Vol 10, pp 150-202.

CASE 2

There was no definite diagnosis. The material in the astrocytes showed some similarity to Rosenthal fibers but there were significant differences. A relationship to Alexander's disease was postulated.

Riggs JE, Schochet SS Jr, Nelson J. Asymptomatic adult Alexander's disease: entity or nosological misconception? Neurology 1988; 38:152-154.

Martin JJ, Martin L, Centerick C. Encephalopathy associated with lamellar residual bodies in astrocytes a new observation. Neuropadiatrie 1977; 8:181-189.

Towfighi J, Grover W, Gonatas NK. Mental retardation, hypotonia and generalized seizures associated with astrocytic "residual" bodies. An ultrastructual study. Human Pathology 1975; 6:667-680.

CASE 3

The diagnosis was melanotic schwannoma. The tumor is of uncertain malignant potential.

Carney JA, Gordon H, Carpenter PC, Vittal Shenoy B, Go VLW. The complex of myxomas, spotty pigmentation, and endocrine hyperactivity. Medicine 1985; 64:270-283.

CASE 4

The diagnosis was malignant schwannoma with rhabdomyosarcomatous differentiation. The patient has neurofibromatosis.

Enzinger FM, Weiss SW. Malignant tumors of peripheral nerve. In: Soft Tissue Tumors. CV Mosby, St. Louis, 1983, p 639-641.

Woodruff J, Chernik NL, Smith MC, Millett WB, Foote FW. Peripheral nerve tumors with rhabdomyosarcomatous differentiation (Malignant Triton tumor): Cancer 1973; 32:426-439.

CASE 5

The inclusions observed in this case of metastatic adenocarcinoma are identical to the inclusions origionally described by Russell as "organisms" which he thought were fungi. The term "Russell bodies" was subsequently used to describe gamma globulin deposits in the cytoplasm of plasma cells. By EM, true Russell bodies are hetrogenous consisting of intracytoplasmic lumina, cytoplasmic nuclear inclusions, apoptotic bodies, and large nucleoli.

Russell W: An address on a characteristic organism of cancer. Brit Med J 1890; 2:1356-1361.

Michels NA: Medullary and non-medullary erythropoesis with special reference to the plasma cell erythrophage or Russell body cell and to the erythrocatheretic (erythrolytic) function of lymph nodes and hemal nodes. Am J Anat 1935; 57:439-499.

CASE 6

The biopsy was obtained in 1982. The patient was given only antiepileptic medications. Her seizures are under control and 7 years later she is functioning in a managerial capacity. The diagnosis was chronic encephalitis. The plasma cells were polyclonal and contained "Russell bodies." No viral particles were demonstrated on EM.

CASE 7

The diagnosis was primary granulomatous angiitis with multiple small infarcts, and secondary acute zygomycotic vasculitis with ruptured mycotic aneurysm.

Case Records of the Massachusetts General Hospital. Primary (granulomatous) angiitis of central nervous system. N Engl J Med 1989; 320:514-524.

Benbow EW, Stoddart RW. Systemic zygomycosis. Postgrad Med J 1986; 62:985-996.

CASE 8

The diagnosis was systemic lymphoproliferative disorder with focal herpes encephalitis due to varicella zoster.

Mayo DR, Booss J. Varicella zoster-associated neurologic disease. Arch Neurol 1989; 46:313, 315.

Salahuddin SZ, Ablashi DV, Markham D, Josephs SF, Sturzeneggfer S, Kaplan M, Halligan G, Kramarsky B, Gallo RC. Isolation of a new virus, HBLV, in patients with lymphoproliferative disorders. Science 1986; 234:596-601.

CASE 9

The diagnosis was rhabdomyositis and cardiomyositis. Although structures resembling myxovirus were observed on EM, the viral culture was negative and a definite viral etiology could not be established. The possibility of this being an autoimmune disease was raised.

Polymyositis and Dermatomyositis. Dalakas MC, ed., Stoneham: Butterworths, 1988.

CASE 10

The diagnosis of the presenter was postictal cerebral and cerebellar hemiatropy: developing as a result of the release of cytotoxic neurotransmitters from the epileptogenic tissue. The CT scans clearly demonstrated that the hemiatropy was acquired not congenital. The presenter further stated that the sparing of the corpra striatum argued against an ischemic or vascular basis for the destructive lesions in the right hemisphere.

Olney JW, Collins RC, Sloviter RS. Excitotoxic mechanisms of epileptic brain damage. In: Advances in Neurology, Vol 44, Delgado-Escueta AV, Ward AA, Woodbury DM, Porter RJ, eds. Raven Press, N.Y., 1986, p857-877.

Tan N, Urich H. Postictal cerebral hemiatropy: with a contribution to the problem of crossed cerebellar atrophy. Acta Neuropath 1984; 62:332-339.