33RD ANNUAL DIAGNOSTIC SLIDE SESSION

1992

CASE 1

The diagnosis in this case depends upon the interpretation of the intracytoplasmic astrocytic inclusions. The presenter thought that they were Rosenthal fibers and that this was a case of Alexander's disease. Identical structures were seen in case 2 of 1988 presented by Dr. Hedley-Whyte (Mass. General Hospital). She has an additional case. Dr. ZuRhein (University of Wisconsin) has two additional cases, one associated with Aicardi's syndrome. Neither think that these are Rosenthal fibers. A case associated with Aicardi's syndrome has also been reported. The authors also did not think that the inclusions were Rosenthal fibers.

Abe H, Yagishita S, Itoh K, Hamano S. Novel eosinophilic inclusions in astrocytes. Acta Neuropath 1992; 83:659-663.

CASE 2

Rosenthal fibers were present throughout the neocortex and white matter as well as the brain stem. There were a few senile plaques in the neocortex and plaques, tangles, and granulovaciolar change in the hippocampus. These were considered to be within normal limits for the age of the patient. The diagnosis was adult onset Alexander's disease. A companion case was presented in 1988 (4) by Dr. Jans Muller. Here the changes were confined to the spinal cord and brain stem. Dr. Muller did not think that the present case belonged in the same category.

CASE 3

The diagnosis was Intestinal Gangliomatosis.

Pham BN, Villanueva RP. Ganglioneuromatous proliferation associated with juvenile polyposis eoli. Arch Path Lab Med 1989;113:91-94.

Mendelsohn G, Diamond MP. Familial ganglioneuromatous polyposis of the large bowel. Am J Surg Pathol 1984;515-520.

DeSchryver-Kecskemet K, Clonse RE, Goldstein MN, et al. Intestinal Ganglioneuromatosis. NEJM 1983;308:635-639.

CASE 4

SPPI

The diagnosis was autosomal dominant spheroid body myopathy. No actin could be demonstrated in the bodies but they were surrounded by alpha B crystaline and ubiquitin.

Halbig L, Goebel HH, Hopf HC, Moll R. Autsomal dominant "spheroid body myopathy." Rev Neurol 1991;147:300-307.

Dickoff DJ, Hayes AP, Uncini A, et al. Autosomal dominant cytoplasmic body myopathy. Clin Neuropathol 1989;8:226.

Gobel HH, Muller J, Gillen HW, Merritt AD. Autosomal dominant "spheroid body myopathy." Muscle and Nerve 1978;1:14-26.

CASE 5

The diagnosis was primary amebic meningoencephalitis, secondary to Naegleria fowleri. The infection was probably acquired in California.

Ma P, Visesvara GS, Martinez AJ, et al. Naegleria and Acanthamoeba infections: renew. Reviews of Infectious Diseases 1990;12:490-513.

Marciano-Cabral F. Biology of Naegleria spp. Microbiological Reviews 1988;52:114-133.

John D. Primary amebic meningoencephalitis and the biology of Naegleria fowleri. Ann Rev Microbiol 1982;36:101-123.

CASE 6

The Purkinje cells showed eosinophilic cytoplasmic inclusions ultrastructurally these had a core of granular or filamentous material with peripheral buds of bullet-shaped viral nucleocapsids. A direct fluorescent antibody reaction for rabies, performed on unfixed tissue, was positive. Culture of this tissue produced a lyssavirus characteristic of the silver-haired bat. The patient has had contact with bats. The diagnosis was rabies encephalitis.

Smith JS, Fishbein DB, Rupprecht CE, Clark K. Unexplained rabies in three immigrants in the United States. A virological investigation. NEJM 1991;324:205-211. (Comments 324:1890-1891).

Smith JS. Rabies virus epitopic variation: Use in ecological studies. Advances in virus research 1989;36:215-253.

Dupont JR, Earle KM. Human rabies encephalitis. A study of 49 fatal cases with a review of the literature. Neurology 1965;15:1023-1034.

CASE 7

The cerebellar peduncle showed severe demyelination and rare cells with intranuclear Cowdrytype A inclusions. These inclusions were also seen in the inferior olivary nucleus. Immunohistochemistry and in situ hybridization were positive for HSV. Electron microscopy showed intranuclear viral particles consistent with herpes virus. Viral cultures from material taken at the time of autopsy grew Herpes Simplex Type 1. These cultures were then used to inject with known susceptibilities to central spread of peripheral herpes. The susceptable strains died within 8 days after intraperitoneal injections. The brains showed brain stem and cerebellar vacuolization and HSV positive cells by immunocytology. The diagnosis was herpes simplex encephalitis.

Kastrukoff LF, Lau AS, Leung GY, Walker D, Thomas EE. Herpes simplex virus type I (HSVI)-induced multifocal central nervous system (CNS) demyelination in mice. J Neuropath Exp Neurol 1992;51:432-439.

Martin JR, Holt RK, Webster H. Herpes-simplex-related antigen in human demyelinative disease and encephalitis. Acta Neuropath 1988;76:325-327.

Lach B, Atack E. Disseminated hemorrhagic leukoencephalomyelitis with localized herpes simplex brain stem infection. Acta Neuropath 1988;75:354-361.

Kastrukoff LF, Lau AS, Kim SU: Multifocal CNS demyelination following peripheral inoculation with herpes simplex virus type I. Ann Neurol 1987;22:52-59.

Roman-Campos G, Toro G: Herpetic brain stem encephalitis. Neurology 1980;30:981-985.

CASE 8

The hippocampal formation shows mesial temporal sclerosis, a common lesion associated with temporal lobe epilepsy. The subcortical white matter shows a hamartomatous malformation. The presenters objected to this term because ἁμάρτια in koine Greek means "sin" (Rom 5:12). They preferred the word for "separate," χωρις, and named the lesion a choristoma.

Burton CJ: The neuropathology of temporal lobe epilepsy. New York, Oxford University Press. 1988.

CASE 9

The diagnosis was plasma cell granuloma (inflammatory pseudotumor). The fusiform cells stained positively for muscle specific actin and were myofibroblasts. The MRI is a gadolinium enhanced image. EM revealed degranulated eosinophiles as well as plasma cells, lymphocytes and myofibroblasts. The authors proposed that this is the result of IGs released from plasma cells and is associated with the release of MBP, EDN and TGF beta which results in mesenchymal proliferation.

Mirra SS, Tindall SC, Check IJ, et al. Inflammatory meningeal masses of unexplained origin. An ultrastructural and immunological study. J Neuropath Exp Neurol 1983;42:453-458.

Pincus SH, Ramesh KS, Wyler D. Eosinophiles stimulate fibroblast DNA synthesis. Blood 1987;70:572-574.

Bahad M, Liebow AA. Plasma cell granulomas of the lung. Cancer 1973;31:191-208.

CASE 10

The cells stained positively for vimentin, desmin, and muscle specific actin and negatively for EMA. The diagnosis was angioleiomyoma arising from the leptomeninges. This tumor has not been previously reported in the CNS.

Freedman AM, Meland NB. Angioleiomyomas of the extremities: report of a case and review of the Mayo Clinic experience. Plast and Recon Surg 1989;83:328-331.

Katenkamp Von D, Kosmehl H, Langbein L. Angiomyoma: Eine pathologisch-anatomische Anayse von 229 Fallen. Zentralbl allg Path Anat 1988;134:423.

Hachisuga T, Hasimoto H, Enjoy, M. Angioleimyoma: A clinicopathologic appraisal of 562 cases. Cancer 1984;54:126-130.