28th Annual Diagnostic Slide Session - 1987

Case 1

The diagnosis was leptomeningitis due to Cryptococcus neoformans (with giant forms) and Prototheca wickerhamii. Although its occurence in skin is well recognized, this is the first reported instance of an algal organism causing meningitis in the human. This organism was isolated from the central nervous system but not from any other organ at post mortem.

Tyler DE, Lorenz MD, Blue JL, et al. Disseminated protothecosis with CNS involvement in a dog. JAVMA 1980;176:987-993.

Connor DH, Neafie RC. Protothecosis. In: Binford CH, Connor DH, eds. Pathology of tropical and extraordinary diseases. Washington DC:AFIP, 1976;2:684-689.

CASE 2

The diagnosis was hemimegalencephaly. Several discussants reported favorable clinical results following surgical resection.

King M, Stephenson JBP, Ziervogel M, Doyle D, Gailbraith S. Hemimegalencephaly - a case for hemispherectomy? Neuropediatrics 1985;16:46-55.

Manz HJ, Phillips TM, Rowden G, McCulloch DC. Unilateral megalencephaly, cerebral cortical dysplasia, neuronal hypertropy, and heterotopia: Cytomorphometric flurometric cytochemical, and biochemical analysis. Acta Neuropathol 1979;45:97-103.

Case 3.

The diagnosis was mucopolysacchrosis type VIB. (Maroteaux-Lamy syndrome).

Scully RE, Mark EJ, McNeely BU. Case records of the Massachusetts General Hospital. N Engl J Med 1983;309:1109-1119.

Young R, Kleinman G, Ojemann RG, Kolodny E, Davis K, Halperin J, Zalneraitis E, DeLong GR. Compressive myelopathy in Maroteaux-Lamy syndrome: Clinical and pathologic findings. Ann Neurol 1980;8:336-340.

Case 4.

The neocortex showed spongioform change but the hippocampal formation was normal. Western blots prepared from forzen brain showed a positive reaction against an antiserum prepared from scrapie-infected hamster brain. This is characteristic of Creutzfeldt-Jakob Disease.

Brown P, Coker-Vann M, Pomeroy K, Franko M, Asher DM, Gibbs CJ, Gajduseck DC. Diagnosis of Creutzfeldt-Jakob disease by western blot identification of marker protein in human brain tissue. N Engl J Med 1986;314:547-551.

Brown P, Rodgers-Johnson P, Cathala, Gibbs CJ, Gajdusek DC. Creutzfeldt-Jakob disease of long duration: Clinicopathological characteristics, transmissibility, and differential diagnosis. Ann Neurol 1984;16:295-304.

Case 5

The infiltrating cells were demonstrated by immunologic methods to be histocytes. the diagnosis was differentiated histiocytosis (histocytosis x).

McMillan E, et al. Analysis of histocytosis x infiltrates with monoclonal antibodies directed against cells of histiocytic, lymphoid, and myeloid lineage. Clin Innuniol Immunopath 1986;38:296-301.

Kepes JJ. Xanthomatous lesions of the CNS: Definition, classification and some recent observations. In: Zimmerman HM, ed. Progress in Neuropathology. New York:Raven Press, 1979;4:179-213.

Case 6

The patient had a four year history of systemic lupis erythematosis. There was no evidence of residual tumor at autopsy and the heart was normal. The presenter's diagnosis was multiple infarcts secondary to lupis vasculitis. Some observers doubted the presence of a primary vasculitis. There was no inflammation in the basilar artery.

DeWitt LD, Kistler JP, Miller DC, Richardson EP, Buonanno FS, NMR-neuropatholgic correlation in stroke. Stroke 1987;18:342-351.

Aisen AM, Gabrielson TO, McCune WJ. MRI of SLE involving the brain. AJNR 1985;6:197-201.

Case 7

The diagnosis was multiple sulphlatase deficiency (mucosulfatidosis).

Burk RD, Valle D, Thomas GH, Miller C, Moser A, Moser H, Rosenbaum KN. Early manifestations of multiple sulfatase deficiency. J Pediatr 1984;104:574-578.

Austin J. Studies in metachromatic leukodystrophy. XII. Multiple sulphastase deficiency. Arch Neurol 1973;28:258-264.

Case 8

Herpes simplex, type II was isolated from the rash. The same organism was demonstrated by immunologic methods and electron microscopy of the spinal cord. The diagnosis was acute necrotizing myelitis due to HSV II.

Wiley CA, Van Patten PD, Carpenter PM, Powell HC, Thai LJ. Acute ascending necrotizing myelopathy caused by Herpes simplex virus type II. Neurology (in press).

Tucker T, Dix RD, Katzen C, Davis RL, Schmidley JW. Cytomegalovirus and Herpes Simplex virus ascending myelitis in a patient with AIDS. Ann Neurol 1985;18:74-79.

Case 9

The diagnosis was systemic Whipple's disease with skeletal muscle involvement. The patient had worked as a sales manager in the jewelry business. Exposure to mineral dusts might have contributed to impairment of macrophage function. He was HIV antigen negative.

Adams M, Rhyner PA, Day J, DeArmond SJ, Smucker EA. Whipple's disease confined to the CNS. Ann Neurol 1987;21:104-108.

Feldman M. Southern Internal Medicine Conference: Whipple's disease. Am J Med Sci 1986;291:56-67.

Case 10

The histologic pattern of this tumor has been well recognized for many years and there is general agreement that it carries a good prognosis. The controveristy has been in defining its histogensis. The presenter showed clear electron microscopic evidence that the cells are of ependymal origin. The diagnosis was ependymoma, clear cell variant. One of the discussors cited a recent paper showing that childhood hemispheric gliomas which present as mural nodules have a good prognosis regardless of their histology.

Tomita T, McLone DG, Naidich TP. Mural tumors with cysts in the cerebral hemispheres of children. Neurosurgery 1986;19:998-1005.

Kawano N, Yada K, Aihara M, Yagishita S. Oligodendroglioma-like cells (clear cells) in ependymoma. Acta Neuropathol 1983;62:141-144.

Case 11

The diagnosis was kinky hair syndrome (Menkes' Disease)

Yoshimura N, Kudo H. Mitrochondrial abnormalities in Menkes' kinky hair disease: Electron microscopic study of the brain from an autopsy case. Acta Necropathol 1983;59:295-303.

Ghatak NR, Hirano A, Poon TP, French JH. Triochopoliodystrophy:II Pathologic changes in skeletal muscle and nervous system. Arch Neurol 1972;26:60-72.

Case 12

The diagnosis was multiple system atropy (Shy-Drager syndrome).

Pastakia B, Polinsky R, DiChiro G, Simmons JT, Brown R, Wener L. Multiple system atropy (Shy-Drager Syndrome): MRI. Radiology 1986;159:499-502 (see also the preceeding paper in the same journal (p. 493-498).

Sung JH, Mastri AR, Segal E. Pathology of Shy-Drager syndrome. J Neuropathol Exp Neurol 1979;38:353-368.