24th Annual Diagnostic Slide Session - 1983

Case 1

Several discussants raised the question of whether this was a xanthoma related to hypercholesteremia or histiocytosis X. The patient had a normal serum cholesterol and electron microscopy failed to reveal Birbeck-Langerhans granules or phagocytosis of plasma cells by histiocytes. The presenter's diagnosis was xanthogranuloma in systemic Weber-Christian disease.

1. Pick P, Jean E, Horoupian D, Factor S. Xanthogranuloma of the dura in systemic Weber-Christian disease. Neurol 1983; 33:1067-1070.

Case 2

The diagnosis was pleomorphic xanthoastrocytoma.

1. Kepes JJ, Rubinstein LJ. Malignant gliomas with heavily lipidized (foamy) tumor cells: A report of 3 cases with immunoperoxidase study. Cancer 1981; 47:2451-2459.
2. Kepes JJ, Rubinstein LJ, Eng LF. Pleomorphic xanthoastrocytomas: A distinctive meningocerebral glioma of young subjects with relatively favorable prognosis. Cancer 1979; 44:1839-1852.

Case 3

There was no general agreement on the diagnosis in this case. Most commentators felt that this was a neuroectodermal dysplasia related to tuberous sclerosis. Others, however, thought that this was a reactive gliosis secondary to a destructive process occurring in a malformed brain. A diagnosis of gliomatosis was also suggested.

Case 4

The diagnosis was subacute sclerosing panencephalitis .

1. Liss L, Oven K, Funkhauser JW, Hardman R, Singla P. Neurofibrillary change in subacute sclerosing panencephalitis. Proc. IX International Congress of Neuorpathology 1982; p.264 (#II-94).
2. Mandybur TI, Nagpaul AS, Pappas Z, Niklowitz WJ. Alzheimer neurofibrillary change in subacute sclerosing panencephalitis. Ann Neurol 1977; 1:103-107.

Case 5

The discussion centered around the question of whether this case should be considered inflammatory or neoplastic. Suggested diagnoses were cerebral infectous mononucleosis, lymphoid granulomatosis, and lymphoma with plasmacytoid features. The presenter favored a diagnoses of lymphoma.

Case 6

The diagnosis was toxoplasmosis.

1. Wongmongkolrit T, McPherson SL, El-Naggar A, et al. Acute fulminant toxoplasma meningioencephalitis in a homosexual man. Acta Neuropath 1983; 60:305-308.
2. Post MJD, Chan JC, Hensley GT, et al. Toxoplasma encephalitis in Haitian adults with aquired immunodeficiency syndrome: A clinical-pathologic-CT correlation. Amer J Neuroradiol 1983; 4:155-162.

Case 7

The diagnosis was congenital hypothalamic hamartoblastoma. This differs from hypothalamic ganglionic hamartoma in that the latter is less cellular and shows good neuronal differentiation. When combined with the other abnormalities which this patient showed it is called the Hall-Pallister Syndrome.

1. Hall JG, Pallister PD, Clarren SK, et al. Congential hypothalamic hamartoblastoma, hypopituitarism, imperforate anus, and post-axial polydactyl - a new syndrome? Amer J Med Genet 1980; 7:47-74.
2. Clarren SK, Alvord Jr EC, Hall JG. Congenital hypothalamic hamartoblastoma, hypopituitarism, imperforate anus and post-axial polydactyly-neuropathological considerations. Amer J Med Genet 1980; 7:75-83.

Case 8

The diagnosis was suprasellar granular cell tumor. The discussion centered around the cell of origin which most commentators felt was in the pituitary system. The GFAP stain was negative but others reported variable staining with this technique. It was suggested that these tumors represent a hetrogenous class. One discussor reported seeing such a tumor in the spinal subarachnoid space.

1. Becker DH, Wilson CB. Symptomatic parasellar granular cell tumors. Neurosurg 1981; 8:173-180.
2. Vagnero J, Leunda G, Cabezuda, et al. Granular pituicytoma of the pituitary stalk. Acta Neuropath 1981; 59:209-215.
3. Symon L, Ganz JC, Chir B, Burston J. Granular cell myoblastoma of the neurohypophysis. J Neurosurg 1971; 35:82-89.
4. Liss L, Kahn EA. Pituicytoma. Tumor of the sella turcica. J Neurosurg 1958; 15:481-488.

Case 9

The diagnosis was neuronal intranuclear hyaline inclusion disease. The inclusions were autofluorescent and had a filamentous structure by EM. The disease presents clinically as a mulitsystem atrophy. A similar case is presented in abstract 143.

1. Michaud J, Gilbert JJ. Multisystem atrophy with neuronal intranuclear hyaline inclusions. Acta Neuropath 1981; 54:113-119.

2. Sung JH, Ramirez-Lassepas M, Mastri AR, Larkin SM. An unusual degenerative disorder of neurons associated with a novel intranuclear hyaline inclusion. J Neuropath Exp Neurol 1980; 39:107-130.
3. Jonata I. Widespread intranuclear neuronal corpuscles (marinesco bodies) associated with a familial spinal degeneration with cranial and peripheral nerve involvment. Neuropath Appl Neurobiol 1979; 5:311-317.
4. Schuffler MD, Bird TD, Sumi SM, Cook A. A familial neuronal disease presenting as intestinal pseudo-obstruction. Gastroenterology 1978; 75:889-898.

Case 10

The diagnosis was Alexander's Disease. It was pointed out that in the original description, enlargement of the brain was a feature, and the condition should be considered in the differential diagnosis of brain stem gliomas.

1. Goebel HH, Bode G. Bulbar palsy with Rosenthal fiber formation in the medulla of a 15-year-old girl. Localized form of Alexander's disease? Neuropediatrics 1981; 12:382-91.

2. Soffer D, Horoupian, DS. Rosenthal fibers formation in the central nervous system. Its relation to Alexander's disease. Acta Neuropathol 1979; 47:81-84.