

32nd ANNUAL DIAGNOSTIC SLIDE SESSION - 1991

Case 1 & 2

Both cases stained positively for amyloid and case 2 was also positive on EM and was found on chemical analysis to be lambda 2-5 subtype. Both cases were cerebral amyloid presenting as a mass lesion and the diagnosis was amyloidoma.

Hori A, Kitamoto T, Tateishi J, Friede RL. Focal intracerebral accumulation of a novel type of amyloid protein. Acta Neuropath 1988; 76:212-215.

Townsend JJ, Tomiyasu U, MacKay A, Wilson CB. Central nervous system amyloid presenting as a mass lesion. J Neurosurg 1982; 56:439-442.

Spaar FW, Goebel HH, Volles E, Wickboldt J. Tumor-like amyloid formation (amyloidoma) in the brain. J Neurol 1981; 224:171-182.

Case 3

The material in this case stained negatively for amyloid and had a granular appearance on EM. It was strongly PAS positive, but diastase resistant, oil red O positive and alcian blue negative. Several discussants reported similar cases but there was no consensus as to pathogenesis or of the nature of the proteinaceous material except that it was not amyloid. The presenter's diagnosis was pseudoamyloid. The reference cited describes a lesion which is grossly and microscopically similar to the present case. The authors did not prove either histologically or radiologically that this was a colloid cyst.

Bentalanffy H, Kretzschmar H, et al. Large colloid cyst in lateral ventricle simulating brain tumor. Acta Neurochir 1990; 104:151-155.

Case 4

The diagnosis was Pseudo-Tangier Disease. No previous cases have been reported.

Yao JK, Natarajan V, Dyck PJ. The sequential alterations of endoneurial cholesterol and fatty acid in Wallerian degeneration and regeneration. J Neurochem 1980; 35:933-940.

Dyck PJ, Effefson RD, Herbert PN. Adult-onset of Tangier Disease: 1. Morphometric and pathologic studies suggesting delayed degradation of neutral lipids after fiber degeneration. J Neuropath Exp Neurol 1978; 37:119-137.

Thomas PK, King RHM. The degeneration of unmyelinated axons following nerve section: an ultrastructural study. J Neurocytol 1974; 3:497-512.

Case 5

Most commentators thought that the muscle changes were those of Werdnig-Hoffman Disease. Many did not find evidence of neuronal loss in the spinal cord sections. The presenter showed definite cell loss in the cranial nerve nuclei. These were associated somatic abnormalities and the diagnosis was Pena-Shokeir Syndrome.

Abe J, Nemoto K, Ohnishi Y, et al.: Case report: Pena-Shokeir Syndrome: A comparative pathologic study. Am J Med Sci 1989; 297:123-127.

Hall JG: Invited editorial comment. analysis of Pena-Shokeir Phenotype. Am J Med Genet 1986; 25:99-117.

Pena SFJ, Shokeir MHK. Syndrome of camptodactyly, multiple ankyloses, facial abnormalities and pulmonary hypoplasia: a lethal condition. J Ped 1974; 85:373-375.

Case 6

The slide showed largely subpial and subependymal inflammation with intranuclear inclusions and a few PML-like white matter lesions. The presenter's diagnosis was atypical papova virus infection, probably SV-40.

Lackner AA, Dandekar S, et al. Neuropathology of similar and feline immunodeficiency virus infections. Brain Pathol 1991; 1:202-212.

Holmberg CA, Gribble DH, et al. Isolation of simian virus 40 from rhesus monkeys with spontaneous progressive multifocal leukoencephalopathy. J Infect Dis 1977; 136:593-596.

Gribble DH, Haden CC, et al. Spontaneous PML in macaques. Nature 1975; 254:602-604.

Case 7

The diagnosis was sarcoidosis. The disease was also present in the mediastinal lymph nodes and on the skin of the left foot.

Clark WC, Acker JD, et al. Presentation of CNS sarcoidosis as intracranial tumors. J Neurosurg 1985; 63:851-856.

Powers WJ, Miller EM. Sarcoidosis mimicking glioma: case report and review of intracranial sarcoid mass lesions. Neurology 1981; 31:907-910.

Case 8

Marker studies showed both T & polyclonal B lymphocytes. No organisms were demonstrated by staining or EM. The diagnosis was idiopathic hypertropic pachymeningitis.

Berger J, Snodgrass S, et al. Multifocal fibrosclerosis with hypertropic intracranial pachymeningitis. Neurology 1989; 39:1345-1349.

Masson C, Henin D, et al. Pachymeningites craniennes de cause indeterminee. Rev Neurol 1989; 145:16-23.

Feringa E, Weatherbee L. Hypertropic granulomatous cranial pachymeningitis causing progressive blindness in a chronic dialysis patient. J Neurol Neurosurg Psychia 1975; 38:1170-1176.

Case 9

The presenter's diagnosis was mixed neurilemmoma and astrocytoma. Several commentators, however, thought the astrocytic component was reactive rather than neoplastic.

Stefanko SZ, Vuzevski VD, et al. Intracerebral malignant schwannoma. Acta Neuropath 1986; 71:321-325.

Kasantikul V, Brown WJ, Cahan LD. Intracerebral neurilemmoma. J Neurol Neurosurg Psychia 1981; 44:1110-1115.

Case 10

The diagnosis was primitive neuroectodermal tumor with ependymal differentiation. The ependymal area was reticulum positive. This tumor apparently has a favorable prognosis.

Ross GW, Rubenstein LJ. Lack of histopathologic correlation of malignant ependymomas with postoperative survival. J Neurosurg 1989; 70:31-36.

Sime PJ, et al. Differentiation in medulloblastomas and other primitive neuroectodermal tumors. Brit J Neurosurg 1989; 3:89-100.

Case 11

The diagnosis was Lhermitte-Duclos Disease.

Milbouw G, Born JD, et al. Clinical and radiological aspects of dysplastic gangliocytoma: a report of two cases with review of the literature. Neurosurgery 1988; 1:124-128.

Shiurba RA, Gessaga EC, et al. Lhermitte-Duclos Disease. An immunohistochemical study of the cerebellar cortex. Acta Neuropath 1988; 5:474-480.

Case 12

On H & E this tumor looks like an oligodendroglioma. The Bodian stain showed rosettes and the tumor stained positively for synaptophysin and neuron specific enolase. It showed neuronal characteristics on EM. The diagnosis was intraventricular central neurocytoma. The prognosis is favorable.

Kubota T, Hayashi M, et al. Central neurocytoma: immunohistochemical and ultrastructural study. Acta Neuropath 1991; 81:418-427.

Barbosa MD, Balsitis M, et al. Intraventricular neurocytoma: A clinical and pathological study of three cases and review of the literature. Neurosurg 1990; 26:1045-1054.

Hassoun J, Gambarelli D, et al. Central neurocytoma: An electron microscopic study of two cases. Acta Neuropath 1981; 56:151-156.