36th ANNUAL DIAGNOSTIC SLIDE SESSION
1995

CASE 1

The diagnoses was primary hypokalemic periodic paralysis. Biopsy is not typically employed in the clinical diagnosis of this disease since it may be normal.

1. Hoffman EP. Voltage-gated ion channelopathies: Inherited disorders caused by abnormal sodium, chloride, and calcium regulation in skeletal muscle. Ann Rev Med 1995; 45:431-441.

2. Jurkat-Rott K, Lehmann-Horn F, Elbaz A, et al. A calcium channel mutation causing hypokalemic periodic paralysis. Hum Mol Genet 1994; 3:1415.

3. Engel AG. Periodic paralysis. In: Myology: Basic and Clinical. Engel AG, Banker BQ, eds, M°Craw Hill, New York, NY 1986, p.1843.

CASE 2

The diagnosis was solvent vapor abuse leukoencephalopathy. The perivascular macrophages contained trilaminar inclusions by electron microscopy. In this condition, as in adrenoleukodystrophy, there is a biochemical abnormality of long chain fatty acids. There was involvement of the cerebral hemispheres and brain stem. Axon cylinders were prominent in the lesions.

1. Kornfeld M, Moser AB, Moser HW, et al. Solvent vapor abuse leukoencephalopathy. Comparison to adrenoleukodystrophy. J. Neuropath Exp Neurol 1994; 53:389-398.

2. Rosenberg NL, Kleinschmidt-DeMasters BK, Davis KA, et al. Toluene abuse causes diffuse CNS white matter changes. Ann Neurol 1988; 23:611-614.

3. Hormees JT, Filley CM, Rosenberg NL. Neurologic sequelae of chronic solvent vapor abuse. Neurology 1986; 36:698-702.

CASE 3

The diagnosis was pellagra-like changes in alcoholic encephalopathy.

1. Victor M. Alcoholic dementia (Review article). Can J Neurol Sci 1994; 21:88-89.

2. Hauw JJ, De Baecque C, Hausser-Hauw C, Serdaru M. Chromatolysis in alcoholic encephalopathies. Pellagra-like changes in 22 cases. Brain 1988; 111:843-857.

3. Ishii N, Nishimara Y. Pellagra among chronic alcoholics: Clinical and pathological study of 20 necropsy cases. J Neurol Neurosurg Psych 1981; 44:209-215.

CASE 4

The diagnosis was multiple system atrophy, also called Shy-Dragger Syndrome and Parkinsonism plus Syndrome. Oligodendroglia inclusions were especially prominent. These stained positively with silver and were ubiquitin positive.

1. Wenning GK, Ben-Shlomo Y, Magalhaes M, et al. Clinicopathological study of 35 cases of multiple system atrophy. J Neurol Neurosurg Psych 1995; 58:160-166.

2. Lantos PL, Papp MI. Cellular pathology of multiple system atrophy: A review. J Neurol Neurosurg Psych 1994; 57:129-133.

3. Quinn N. Multiple system atrophy: the nature of the beast. J Neurol Neurosurg Psych 1989 Supp; 78-89.

CASE 5

The diagnoses was polyglucosan body disease.

1. Caffenty MS, Lovelace RE, Hays AP, et al. Polyglucosan body disease. Muscle and Nerve 1991; 14:102-107.

2. Lossos A, Barash V, Stoffer D, et al. Hereditary branching enzyme dysfunction in adult polyglucosan body disease: A possible metabolic cause in two patients. Ann Neurol 1991; 30:655-662.

3. Robitaille Y, Carpenter S, Karpati G, Dimausos S, et al. A distinct form of adult polyglucosan by body disease with massive involvement of central and peripheral neuronal processes and astrocytes. Brain 1980; 103:315-336.

CASE 6

The diagnosis was cerebral mycobacteriosis. The perivascular macrophages contained acid fast bacilli which were also PAS positive and stained with luxol fast blue. In addition the patient showed giant cells, which stained positively for HIV-1, and cytomegalic inclusion disease.

Anders KH, Goerra WF, Tomiyasu U, et al. The neuropathology of AIDS: UCLA experience and review. Am J Path 1986; 124:537-558.

CASE 7

The diagnosis was leiomyoma of dura and skin. The tumor was S-100 negative and muscle specific actin positive. Electron microscopy showed cells with muscle characteristics. Epstein-Barr virus was not demonstrated.

1. M°Clain KL, Leach CT, Jenson HB, et al. Association of Epstein-Barr virus with leiomyosarcomas in young people with AIDS. N Eng J Med 1995; 332:12-18.

2. Steel TR, Pell MF, Turner JJ, Lim GHK. Spinal epidural leiomyoma occurring in an HIV-infected man. J Neurosurg 1993; 79:442-445.

3. Chadwick EG, Connor EJ, Hanson CG, et al. Tumors of smooth muscle origin in HIV-infected children. JAMA 1990; 263:3182-3184.

CASE 8

The granules in the tumor cells were PAS positive. The cells stained positively for S-100 and vimentin. Electron microscopy showed coarse cytoplasmic granules. The diagnosis was malignant granular cell tumor.

1. Albuquerqe L, Pimentel J, Costa A, Christina L. Cerebral granular cell tumors: Report of a case and a note on their nature and expected behavior. Acta Neuropathol 1992; 84:680-685.

2. Gambini C, Ruelle A, Palladino M, Baccandro M. Intracerebral granular cell tumor. Pathologica 1990; 82:83-88.

3 Sakurama N, Matsukado Y, et al. Granular cell tumor of the brain and its cellular identify. Acta Neurochir 1981; 56:81-94.

CASE 9

The diagnosis was congenital glioblastoma multiforme with an angioblastic component.

1. Roosen N, Deckert M, Nicola N, et al. Congenital anaplastic astrocytoma with favorable prognosis. Case report. J Neurosurg 1988; 69:604-609,

2. Janisch W, Haas JF, Schreiber D, Gerlach H. Primary CNS tumors in stillbirths and infants. J Neuro Onc 1984; 2:113-116.

3. Sabet LM. Congenital glioblastoma multiforme associated with heart failure. Arch Pathol Lab Med 1982; 106:31-34.

CASE 10

The tumor was vimentin positive and negative for EMA, S-100, SMA and CD34. Electron microscopy showed spindle cells most of which had the characteristics of fibroblasts but some resembled myofibroblasts. The diagnosis was intracranial fibromatosis. The patient is doing well two years after surgery.

1. Mitchell A, Scheithauer BW, Ebersold MJ, Forbes GS. Intracranial fibromatosis. Neurosurg 1991; 29:123-126.

2. Dolman CL, Crichton JU, Jones EA, Lepointe J. Fibromatosis of dura presenting as infantile spasms. J Neurol Sci 1981;49:31-39.

3. Friede RL, Pollak A. Neurosurgical desmoid tumors: Presentation of four cases with a review of the differential diagnosis. J Neurosurg 1979; 50:725-732.

CASE 11

The diagnosis was meningioma with involvement of skull and subcutaneous tissue and pronounced hyperostosis (Tower skull).

1. Burger PC, Schiethauer BW, Vogel FS. Skull and related soft tissues. In: Surgical pathology of the nervous system and its coverings, 3rd ed. Churchill Livingstone, New York, NY, 1991, Chapter 1 p1-66.

2. Cushing H, Eisenhardt L. The hyperostosing tumors. In: Meningiomas. Charles C. Thomas, Springfield, IL and Baltimore, MD 1938, Chapter XXI, p463-502.