CASE 1

The diagnosis was Infective Vasculitis due to Bacillus cereus occurring in an immunocompromised host. This organism is a large gram positive rod which occurs in soil and is not considered to be a pathogen.

Koneman EW. The aerobic gram positive bacilli. Koneman EW, Allen SD, Janda WM, et al eds., Diagnostic Microbiology, Philadelphia, J. B. Lippincott 1992, pp 467-518.

Bryce EA, Smith JA, Tweeddale M, et al. Dissemination of Bacillus cereus in an intensive care unit. Infect Control Hosp Epidemiol 1993;14:459-462.

Drobniewski PA. Bacillus cereus and related species. Clin Microbiol Reviews 1993;6:324-338.

CASE 2

The gross lesion had a rubbery consistency. The infiltrating cells were polyclonal and included plasma cells and giant cells. Silver stains revealed spirochetes and the presenter's diagnosis was gumma. The question of the concomitant presence of lymphoma was raised.

Morgello S, Laufer H. Quaternary neurosyphilis in a Haitian man with HIV infection. Hum Pathol 1989;20:808-811.

Holowitz HW, Valsamis MP, Wicher V, et al. Cerebral syphilitic gumma confirmed by polymerase chain reaction in a man with HIV infection. NEJM, in press.

CASE 3

The diagnosis was Varicella-zoster encephalitis. Although the lesion resembles an infarct, it contains cells with Cowdry Type A intranuclear inclusions. Electron microscopy revealed virions consistent with herpes. Immunohistochemistry was positive only for VZV. The Bielschowsky stain showed some preservation of axons.

Morgello S, Block GA. Varicella-zoster virus leukoencephalitis and cerebral vasculopathy. Arch Path Lab Med 1988;112:173-177.

Rosenblum MK. Bulbar encephalitis complicating trigeminal zoster in AIDS. Hum Path 1989; 20:292-295.

Gray F, Mohr M, et al. Varicella-zoster virus encephalitis in AIDS: Report of four cases. Neuropath Appl Neurobiol 1992;18:502-514.

CASE 4

The diagnosis was Post-transplant Lymphoproliferative Disorder. The lymphocytes were predominantly beta cells. In situ hybridization was strongly positive for Epstein-Barr virus.

Nelesnik MA, Jaffe R, Starzl TE, et al. The pathology of post-transplant lymphoproliferative disorders occurring in the setting of cyclosporine A-prednisone immunosuppression. Am J Path 1988;133:173-192.

Randhawa PS, Jaffe R, Demetris AJ, et al. The systemic distribution of Epstein-Barr virus genomes in fatal post-transfusion lymphoproliferative disorders. Am J Pathol 1991;138:1027-1033.

Swerdlow SH. Post-transplant lymphoproliferative disorders: a morphologic, phenotypic and genotypic spectrum of disease. Histopathology 1992;20:373-385.

CASE 5

The presenter's diagnosis was Familial Mitochondrial Encephalopathy. The discussion centered around the question of whether or not this is a form of Leigh's disease.

Dussieres LM, I'Flugfelder IW, et al. Exercise responses after cardiac transplantation in mitochondrial myopathy. Am J Cardiol 1993;71:1003-1006.

Santorelli FM, Shancke S, et al. The mutation at nt8993 of mitochondrial DNA is a common cause of Leigh's syndrome. Ann Neurol 1993;34:827-834.

Sparaco M, Bonilla E, et al. Neuropathology of mitochondrial encephalopathies due to mitochondrial DNA defects. J Neuropath Exp Neurol 1993;52:1-10.

CASE 6

The diagnosis was Choreoacanthcytosis.

Feinberg TE, Cianci CD, Morrow JS, et al. Diagnostic tests for choreoacanthocytosis. Neurology 1991;41:1000-1006.

Haedie RJ, Pull on HWH, Owen JS, et al. Neuroacanthocytosis: A clinical, hematological and pathological study of 19 cases. Brain 1991;114:13-49.

Bosman CJCGM, Bartholomeus IGP, DeGrip WJ, Horstink MWIM. Erythrocyte anion transporter and antibrain immunoreactivity in chorea-acanthocytosis. A contribution to etiology, genetics, and diagnosis. Brain Res Bull 1994;33-5:523-528.

CASE 7

The perivascular material did not stain with Congo Red but was positive with beta A4 antibody and electron microscopy revealed filaments consistent with amyloid. The presenter's diagnosis was amyloid angiopathy and intraparenchymal amyloid deposition. The hippocampus showed granulovacuolar degeneration. The silver stain showed no plaques in the cortex but numerous tangles were seen in small neurons stained with the tau antibody. The question of a variant of Alzheimer's disease was raised but most commentators did not think that this diagnosis was justified.

Heffner RR Jr, Porro RS, Olson ME, Earle KM. A demyelinating disorder associated with cerebrovascular amyloid angiopathy. Arch Neurol 1976;33:501-506.

Gray F, Dubas F, Roulett E, Escourolle R. Leukoencephalopathy in diffuse hemorrhagic cerebral amyloid angiopathy. Ann Neurol 1985;18:54-59.

Vinters HV. Cerebral amyloid angiopathy. A critical review. Stroke 1987;18:311-324.

Cohen M, Lanska D, Roessman U, et al. Amyloidoma of the CNS. I Clinical and pathological study. Neurology 1992;42:2019-2023.

CASE 8

The diagnosis was cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL). The radiology and histopathology are that of Binswanger's disease.

Mas JL, Dilouya A, de Recondo J. A familial disorder with subcortical ischemic strokes, dementia, and leukoencephalopathy. Neurology 1992;42:1015-1019.

Baudrimont M, Dubas F, Joutel A, et al. Autosomal dominant leukoencephalopathy and subcortical ischemic stroke. Stroke 1993;24:122-125.

Bousser MG, Tournier-Lasserve E. Summary of the Proceedings of the First International Workshop on CADASIL. Stroke 1994;25:704-707.

CASE 9

The diagnosis was Galloway-Mowat Syndrome. This is an autosomal recessive migrational disorder.

Choi BH, Matthias SC. Cortical dysplasia associated with massive ectopia of neurons and glial cells within the subarachnoid space. Acta Neuropath (Berl) 1987;73:105-109.

Choi BH. Developmental events during the early stages of cerebral cortical neurogenesis in man: a correlative light, electron microscopic, immunohistochemical and Golgi study. Acta Neuropathol (Berl) 1988;75:441-447.

Kozlowski PB, Sher JH, Nicastri AD, Rudelli RD. Brain morphology in the Galloway syndrome. Clin Neuropathol 1989;8:85-91.

Cooperstone BG, Friedman A, Kaplan BS. Galloway-Mowat syndrome of abnormal gyral patterns and glomerulopathy. Am J Med Genet 1993;47:250-254.

Norman MG, McGillivary BC, Kalousek DK, et al. Congenital malformations of the brain. Oxford Univ Press, in press.

CASE 10

This is an example of displacement of cerebellar tissue from herniated cerebellar tonsils. It is generally seen only in patients who have been on a respirator.