CASE 1

The diagnosis in this case depends upon the interpretation of the intracytoplasmic astrocytic inclusions. The presenter thought that they were Rosenthal fibers and that this was a case of Alexander's disease. Identical structures were seen in case 2 of 1988 presented by Dr. Hedley-Whyte (Mass. General Hospital). She has an additional case. Dr. ZuRhein (University of Wisconsin) has two additional cases, one associated with Aicardi's syndrome. Neither think that these are Rosenthal fibers. A case associated with Aicardi's syndrome has also been reported. The authors also did not think that the inclusions were Rosenthal fibers.


CASE 2

Rosenthal fibers were present throughout the neocortex and white matter as well as the brain stem. There were a few senile plaques in the neocortex and plaques, tangles, and granulovacuolar change in the hippocampus. These were considered to be within normal limits for the age of the patient. The diagnosis was adult onset Alexander's disease. A companion case was presented in 1988 (4) by Dr. Jans Muller. Here the changes were confined to the spinal cord and brain stem. Dr. Muller did not think that the present case belonged in the same category.

CASE 3

The diagnosis was Intestinal Gangliomatosis.


The diagnosis was autosomal dominant spheroid body myopathy. No actin could be demonstrated in the bodies but they were surrounded by alpha B crystalline and ubiquitin.


The diagnosis was primary amebic meningoencephalitis, secondary to Naegleria fowleri. The infection was probably acquired in California.


The Purkinje cells showed eosinophilic cytoplasmic inclusions ultrastructurally these had a core of granular or filamentous material with peripheral buds of bullet-shaped viral nucleocapsids. A direct fluorescent antibody reaction for rabies, performed on unfixed tissue, was positive. Culture of this tissue produced a lyssavirus characteristic of the silver-haired bat. The patient had had contact with bats. The diagnosis was rabies encephalitis.


CASE 7

The cerebellar peduncle showed severe demyelination and rare cells with intranuclear Cowdry type A inclusions. These inclusions were also seen in the inferior olivary nucleus. Immunohistochemistry and in situ hybridization were positive for HSV. Electron microscopy showed intranuclear viral particles consistent with herpes virus. Viral cultures from material taken at the time of autopsy grew Herpes Simplex Type I. These cultures were then used to inject with known susceptibilities to central spread of peripheral herpes. The susceptible strains died within 8 days after intraperitoneal injections. The brains showed brain stem and cerebellar vacuolization and HSV positive cells by immunocytology. The diagnosis was herpes simplex encephalitis.


CASE 8

The hippocampal formation shows mesial temporal sclerosis, a common lesion associated with temporal lobe epilepsy. The subcortical white matter shows a hamartomatous malformation. The presenters objected to this term because ἁμάρτια in koine Greek means "sin" (Rom 5:12). They preferred the word for "separate," ἄφις, and named the lesion a choristoma.

CASE 9

The diagnosis was plasma cell granuloma (inflammatory pseudotumor). The fusiform cells stained positively for muscle specific actin and were myofibroblasts. The MRI is a gadolinium enhanced image. EM revealed degranulated eosinophiles as well as plasma cells, lymphocytes and myofibroblasts. The authors proposed that this is the result of IGs released from plasma cells and is associated with the release of MBP, EDN and TGF beta which results in mesenchymal proliferation.


CASE 10

The cells stained positively for vimentin, desmin, and muscle specific actin and negatively for BMA. The diagnosis was angioleiomyoma arising from the leptomeninges. This tumor has not been previously reported in the CNS.

