ECVP/ESVP Summer School in Veterinary Pathology

Marie Curie Training Courses

Summer School 2006 – Animal Modles 1768/96

1768/96
Cat, Korat

Cerebral cortex (possibly at the level of striated nuclei). The grey matter and nuclei of striated bodies are moderately and diffusely expanded by an abundant number of variably enlarged neuronal bodies. Distinct neuronal layers of the cortex are no longer evident. The enlarged neurons (up to 50 microns in diameter) have a rounded and distinct cell outline, abundant finely granular to foamy, faintly eosinophilic cytoplasm, and peripherally displaced oval and vesicular nuclei. Occasionally nuclei are shrunken, hyperchromatic and crescent shaped (pyknotic). Intermingled with neuronal nuclear groups and scattered within both the surrounding white and gray matter there are numerous spherical structures (spheroids) of variable loosely granular to densely eosinophilic appearance (axonal swellings due to accumulation of tubules and vesicles). In addition, a moderate number of disseminated glial cells is evident throughout the entire section, possibly consistent with hyperplastic and hypertrophic astrocytes (oval vesicular nuclei) and oligodendrocytes. Occasionally small cells, scattered within the white and grey matter and possibly consistent with glial elements, have also a moderately expanded densely eosinophilic to finely granular cytoplasm (stored material). Multifocal areas show numerous irregular empty vacuoles (spongy state) associated with loss of cytoplasmic margins of the swollen neuronal bodies [only some sections]. Perivascular empty spaces are frequently evident (edema), often associated with numerous vacuolated to foamy macrophages.
Brainstem. Diffusely, similar, severe neuronal swelling and spheroid formation is evident.
Cerebellum. Similarly to other sections, neurons of the Purkinje and granular layers are diffusely and variably enlarged; occasional loss of cytoplasmic margins is present. Multifocally, small cells consistent with glial cells also feature cytoplasmic eosinophilic swelling. Disseminated spheroids are evident. Multifocal to diffuse severe enlargement of the molecular and granular layers is evident due to clear irregular spaces and vacuoles (edema and loss of neurons).

Morphological diagnosis:
Cerebrum, brain stem and cerebellum: neuronal vacuolation, cytoplasmic, diffuse, moderate to severe, with multifocal intraglial cytoplasmic vacuolization.

Etiologic Diagnosis: Hereditary lysosomal neuropathy

Name of disease: GM1 gangliosidosis

Cause: Autosomal recessive lysosomal beta-galactosidase deficiency

Differential Diagnosis: Other lysosomal storage diseases (definitive diagnosis requires biochemical tests of stored material and/or ultrastructural examination to identify the accumulated material and/or enzymatic tests).