NEURONAL CEROID LIPOFUSCINOSIS IN A BEAR FROM KALVENE ZOO

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In the spring of 2013 a five year old bear kept in Kalvene Zoo (Nature Park of Riga Zoo) developed sudden motor disfunction, loss of coordination and difficulty to eat. During summer neurological deficits progressed and bear was killed in September of 2013. Postmortem necropsy perfomed at FVM showed no gross lesions in the brain, spinal cord or muscles. Microscopic examination revealed accumulation of large amount of fine eosinophilic-yellow vacuoles in nearly neurons of all levels of brain and spinal cord. Neurons were often fragmented, necrotic or lost. Vacuoles were intensively positive by LFB stain and moderately positive for PAS stain confirming lysosomal storage disease. The appearance of vacuoles and staining properties suggested neurodegenerative storage disease -- neuronal ceroid lipofuscinosis.

Neuronal ceroid lipofuscinosis (NCL) is a lysosomal storage disease characterized by accumulation of lipopigment within central nervous system and peripheral tissues. This occurs due to mutations in genes that code for a variety of proteins including lysosomal enzymes and membranes of various compartments. Accumulation of lipopigment is progressive and cumulative, resulting in disruption of neuronal integrity and imparing neuronal functions.

The bear affected by NCL was offspring of close sibling / parent matings several generations in a row which likely facilitated expression of this recessively inhereted disease. NCL is rare disease best characterized in humans but also affecting domestic animals – most commonly dogs, cats, sheep and cattle.