NEURODEGENERATIONS IN ANIMALS

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Neurodegenerations are characterized by primary degeneration and loss of certain cells and secondary degeneration of associated cells in specific regions and cell populations of the nervous system. Degenerative lesions are bilaterally symmetrical and often quite discrete; gliosis is often the only indication that a lesion is present. Many of these diseases are congenital hereditary, familial, or of suspected genetic background. Such diseases present as slowly progressive neurologic symptoms in immature animals. The underlying mechanism are often unknown. Several conditions affect primarily the neuronal cell body, others the axon. The most common ones are discussed in more detail here below.

**Cerebellar atrophy**

This disease occurs in different species especially dogs. More than 40 different entities have been reported. Macroscopically the cerebellum is almost normal in shape and size, but histologically the Purkinje cells are lacking to a great extent.

**Motor Neuron Disease (MND)**

Degeneration of motor neurons in the ventral horns of the spinal cord, and neurons of brain stem nuclei, have been observed in several species in a variety of conditions. In all these conditions, the lesions are very similar. The neurons are swollen, with chromatolysis and nuclear changes. There is Wallerian degeneration in the peripheral nerves and neurogenic muscle atrophy. Some of these conditions are hereditary (also called spinal muscular atrophy or SMA) e.g. SMA in Brown Swiss calves and Brown Swiss crosses, SMA in brittany spaniels and swedish lapland dogs. In man there are hereditary as well as sporadic forms of amyotrophic lateral sclerosis (ALS). Motor neuron degeneration can occur as a result of trace element deficiency in lambs (Sway back), goat kids and in piglets. Many motor neuron diseases are sporadic such as Equine motor Neuron Disease (EMND). Affected animals have low vit E serum levels and a history of limited access to pasture. Similar lesions have been found in young animals of a variety of other species including cats, cattle,
pigs, rabbits and zebras. In all these reports, a genetic defect had been suspected but not proven.

**Dysautonomia**

Diseases affecting the autonomous nervous system have been recognized in animals. Grass sickness in horses has been known for a long time in certain endemic regions in Europe. Dysautonomia occurs also in in cats and rarely in dogs. The disease appears to be endemic in wild living hares. The cause and pathogenesis of dysautonomia is not known. In grass sickness it is likely that a hitherto unrecognized plant poison is involved. Pathologically all these conditions are characterized by degeneration of the autonomic neurons in ganglia, spinal cord and brainstem. The degeneration leads to massive neuronal loss and proliferation of the supporting cells.

**Multisystemic neuron degeneration in Cocker Spaniels**

The first symptoms with seizures and dementia show in puppies at 2 - 6 months of age, rarely later up to 1 year. The lesions in the brain consist in status spongiosus of the cerebral white matter and various brain stem nuclei, gliosis, swollen axons and loss of neurons in affected nuclei. Cerebellar atrophy in the Kerry blue terrier is associated with degeneration of the globus pallidus; thus this is also a multisystemic degeneration.

**Hereditay ataxia in Jack Russell terriers and smooth haired Fox terriers**

Very little is known about the genetic background of this disorder. But as it almost exclusively affects those two closely related breeds a genetic basis seems probable. The histologic lesions never are spectacular. They consist fibre degeneration throughout the long tracts especially the dorso-lateral columns of the cord, the cerebral peduncles the brain stem as well. Degenerating axons are found in the trapezoid body. There are also degenerating fibers in the peripheral nervous system.

**Weaver Syndrome, Myeloencephalopathy in Brown Swiss cattle**

The symptoms start with progressive ataxia at the age of 5 - 6 months. An autosomal recessive hereditary trait is suspected. Pathologically the disease affects the white matter of the spinal cord, most severely in the thoracic segments. Axonal degeneration with spheroid formation and axonal loss (empty "swollen" myelin sheaths) as well as gliosis are seen. Similar but milder lesions
may be present in the brain stem, and scattered Purkinje cell degeneration is a consistent finding.

**Axonal dystrophy in Rottweiler dogs**
The first symptoms appear seldom before, but in most instances after 1 year of age. Autosomal recessive inheritance is postulated. The characteristic lesion consist in large numbers of swollen axons (spheroids) at the entries of nerve roots in the dorsal horns throughout the spinal cord, and certain brain nuclei such as the geniculate bodies in the latero-caudal thalamus. Similar axonal dystrophies have been described in other dog breeds (Scotch Terrier, Chihuahua, Collies), sheep and rarely in cats and horses.

**Equine Degenerative Myeloencephalopathy**
The disease affects mostly young horses of about 6 months, but the first symptoms may appear as late as 24 months of age. They present with disturbed general proprioception and upper motor neuron function. On histologic slides all funiculi show Wallerian degeneration and astrogliosis throughout the entire length of the spinal cord. But most affected are the dorsolateral (spinocerebellar) and ventromedial (motor) funiculi of cervical and thoracic segments. Neuronal necrosis can be found in Clarke's column dorsolateral to the central canal of the thoracic and anterior lumbar spinal cord and in the dorsal column nuclei. Vit. E deficiency appears to play a role. The disease is most common in the US. In Europe there have been very few reports.

**Idiopathic Myelopathy in large dog breeds**
This change has been described first in older German shepherds with paresis and abnormal gait in the hind quarters. Later it was observed in other large dog breeds as well. The lesions are restricted to the spinal cord and as rule most conspicuous in the thoracic segments. They consist in scattered ballooned myelin sheaths and an occasional swollen axons, but no inflammatory cells or macrophages. In most cases the rather mild histologic lesions do not correspond with the severe clinical deficits.