AABP & AASRP Joint Sessions

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Chronic Wasting Disease of North American Cervids

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Abstract

Chronic wasting disease (CWD) is a transmissible spongiform encephalopathy that has been reported in captive and free-ranging mule deer (*Odocoileus hemionus hemionus*), white-tailed deer (*Odocoileus virginianus*), Rocky Mountain elk (*Cervus elaphus nelsoni*) and moose (Alces alces shirasi). CWD has been reported in free-ranging cervids in 11 states and two Canadian provinces. CWD has been found in captive cervids in eight states and two Canadian provinces, but all of these herds have been depopulated except for three herds in Colorado. The mode of transmission of CWD is not known, but thought to be associated with ingestion of contaminated food or water from a contaminated environment. The primary clinical signs in cervids with CWD include weight loss and excessive salivation. Primary gross lesions include emaciation with loss of body fat. The hallmark histological lesion of CWD is spongiform encephalopathy. Diagnosis can only be confirmed with examination of the brain and/or lymphoid tissues with immunohistochemical staining. ELISA tests are used for screening large numbers of samples, for example during hunter kill surveys.

Résumé

La maladie débilitante chronique (MDC) est une encéphalopathie spongiforme transmissible qui a été signalée chez le cerf mulet (*Odocoileus hemionus hemionus*), le cerf de Virginie (*Odocoileus virginianus*), le wapiti des Rocheuses (*Cervus elaphus nelsoni*) et l'élan du Yellowstone (*Alces alces shirasi*), chez des sujets en captivité et en liberté. La MDC a été signalée chez des cervidés en liberté de 11 états américains et de deux provinces canadiennes. La MDC a été décelée chez des cervidés en captivité dans huit états américains et deux provinces canadiennes, mais tous ces troupeaux ont été dépeuplés à l'exception de trois troupeaux du Colorado. Le mode de transmission de la MDC n'a pas été élucidé, mais on croit qu'il serait associé à l'ingestion d'aliments contaminés ou d'eau provenant d'un milieu contaminé. Les principaux signes cliniques de la MDC chez les cervidés comprennent la perte pondérale et l'hypersalivation. Les lésions macroscopiques primaires incluent l'émaciation avec perte de tissus adipeux, tandis que la lésion histologique classique de la MDC est l'encéphalopathie spongiforme. Le diagnostic ne peut être confirmé que par l'examen du cerveau et/ou par coloration immunohistochimique des tissus lymphoïdes. Les tests ELISA sont utilisés pour l'analyse d'échantillons en grand nombre, par exemple lors d'enquêtes sur les animaux tués à la chasse.

General Comments

Chronic wasting disease (CWD) is a transmissible spongiform encephalopathy and has been reported in captive and free-ranging mule deer (*Odocoileus hemionus hemionus*), white-tailed deer (*Odocoileus virginianus*) and Rocky Mountain elk (*Cervus elaphus nelsoni*). CWD was thought to be first observed as early as 1965 in captive mule deer facilities maintained by researchers from Colorado State University, Fort Collins Colorado and then by personnel with the Colorado Division of Wildlife in the early 1970s. CWD was identified as a spongiform encephalopathy by Drs. Williams and Young in 1978 in mule deer from the research facilities maintained by the Colorado Division of Wildlife. CWD has been reported in free-ranging cervids in Colorado (1981-elk, 1984-MD, 1985-WTD), Wyoming (1985-MD, ??-elk), Nebraska (2000-MD), South Dakota (2001-WTD), Wisconsin (2002-WTD), New Mexico (2002-MD), Illinois (2002-WTD), Utah (2003-MD), New York (2005-WTD), West Virginia (2005-WTD) and Kansas (2006-WTD). CWD has also been found in two Canadian provinces: Saskatchewan (2000-MD; 2001-WTD) and Alberta (2006-MD). CWD has been found in captive cervids in South Dakota (1997-elk), Nebraska (1998-elk, 2002-WTD), Oklahoma (1998-elk), Montana (1999-elk), Colorado (2000-elk), Kansas (2001-elk), Wisconsin (2002-WTD) and Minnesota (2002-elk). CWD has

also been found in captive cervids in two Canadian provinces: Saskatchewan (1996-elk, 2002-WTD) and Alberta (2002-elk).

Etiology

The etiology of CWD is thought to be the alteration of a normal cellular protein (PrP^{C}) of the body. This protein (PrP^{C}) is thought to be located in nearly all tissues, but is especially abundant in lymphoid and neural tissues. Its function is not presently known, but is thought to be associated with neural transmission at synaptic junctions. The dogma at the present time is that the agent that causes CWD (PrP^{CWD}) enters the body by ingestion and is then sequestered by lymphoid cells of the head and digestive tract. The PrP^{CWD} then alters the PrP^{C} in the animal. This same activity next occurs in the brain. The accumulation of this extremely stable isoform (PrP^{Sc}) of the normal cellular prion protein (PrP^{C}) with the inability of the body to break it down or metabolize it is thought to be the pathogenesis of the TSEs. PrP^{CWD} first accumulates in the dorsal motor nucleus of the vagus nerve, then spreads to the hypothalamic nuclei throughout the thalamus. Next PrP^{CWD} seems to spread throughout the brain. The cerebellum is the last neuroanatomical region to accumulate PrP^{CWD}.

The infectivity or stability of PrP^{CWD} is thought to be similar to scrapie. However, investigation into the infectivity and stability has not been done.

Susceptible Host

The primary natural hosts for CWD include mule deer (*Odocoileus hemionus hemionus*), white-tailed deer (*Odocoileus virginianus*) and Rocky Mountain elk (*Cervus elaphus nelsoni*). CWD has recently been observed in three moose (Alces alces shirasi) in north central Colorado. Non-natural hosts include ferrets, cattle, sheep, squirrel monkeys, goats, hamsters and to some degree, mice. These hosts were inoculated via the intracerebral route.

Transmission

The transmission of CWD is not known, but is thought to be associated with environmental contamination with oral ingestion of the prion on feed material.

Clinical Signs

The incubation period for CWD is probably two to five years, depending on the species and genetics of the species. The incubation was approximately two years in MM elk, three and one-half years in M/L elk and four to five years in L/L elk in one experiment in which 10 grams of brain homogenate was fed. Clinical signs are usually not observed until the terminal stages of the disease. Clinical signs include slight to moderate behavioral changes, excessive salivation, grinding of the teeth (especially in elk) and weight loss. Animals usually maintain a good appetite.

Post Mortem Lesions/Histopathology

The most striking gross lesion in animals in the terminal stage of CWD is emaciation with total loss of subcutaneous body fat. Even the adipose tissues behind the eyes and within the spinal column will have undergone serous atrophy. Occasional aspiration pneumonia can be found in deer and elk. In captive mule deer the rumen often contains an excessive amount of water; however, this lesion is rare in free-ranging deer. This lesion is also rare in captive and free-ranging elk. Rarely, abomasal ulcers are present.

The classical histological lesion is a spongiform encephalopathy. The lesion begins in the dorsal motor nucleus of the vagus nerve, then spreads to the hypothalamic nuclei. Next, the spongiform degeneration affects the thalamus, basal nuclei and spinal cord, followed by involvement of the cerebrum. The cerebrum is the last neuroanatomical region to be affected. The only other consistent histological lesion is a generalized depletion of lymphocytes in internal and peripheral nodes and spleen. In terminal cases, PrP^{CWD} has been found in other organs besides the brain and lymphoid tissues. These tissues include the adrenal gland, islets of Langerhans, myoenteric plexus, cardiac muscle, skeletal muscle and retina of eye. No histological lesions are found in the organs except for a few vacuolated neurons in the ganglia cell layer of the retina in elk.

Diagnosis

A diagnosis of CWD cannot be made on the basis of physical examination. First, a physical examination is difficult to do on a cervid anyway, but many conditions can cause chronic wasting in cervids. At the present time there are two live animal tests available with some degree of accuracy. The first is a palatine tonsil biopsy and the second is a rectal mucosal biopsy. Both of these biopsied tissues are then stained with immunohistological techniques with monoclonal antibody specific for PrP^{CWD}. It seems that the palatine tonsil technique is more difficult to perform, but is more accurate that the rectal test. One advantage to the rectal biopsy is that the instruments used for the biopsy can be discarded because they are inexpensive, whereas the tonsilar biopsy instruments are extremely expensive and must be sterilized between uses. Sterilization of instruments for prions is problematic. To confirm a

case of CWD, the brain and/or lymphoid tissues (preferably retropharyngeal lymph node and palatine tonsil) are examined via immunohistochemistry. ELISA techniques are often used for screening large numbers of animals, but all suspected positive cases are confirmed with immunohistochemistry.

Treatment

There is no known treatment for CWD in cervids.

Prevention and Control

CWD has been a devastating disease to free-ranging cervids and in the captive elk and white-tailed deer industries. An estimated 12-14,000 captive elk have been killed in the last six or seven years in attempts to contain CWD in captive herds. Several thousand free-ranging mule deer, white-tailed deer and elk also have been killed in attempts to reduce the disease in the wild. This killing of captive cervids appears to have reduced or at least slowed down the spread of CWD in these ranch-raised situations, but has had little to no effect in free-ranging herds.

Captive elk and deer cannot be ruled out as sources of infection of CWD in both free-ranging and ranch-raised cervids, therefore eradication of CWD in captive herds is a critical component of any strategy to limit the spread of CWD.

Suggested Reading

Williams ES: Chronic wasting disease. *Vet Path* 42:530-549, 2004.