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Diagnostic Exercise From the Latin Comparative Pathology Group and the Davis-Thompson Foundation

Coenurosis in sheep

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Clinical history:

Cases of neurologic disease occurred in a sheep farm in late 2023, with a surge in cases in July 2024. The disease affected lambs more frequently. Clinical signs lasted from 7 days to 1 month and included blindness, ataxia, lateral deviation of the head, circling, and reflux of ruminal contents through the nose. One animal had exophthalmos. There were several dogs on the farm. The total number of sheep on the farm was 100 and 50 were in the group where the disease occurred. Six of those 50 had clinical signs and died. All sheep were free in the pasture.

Autopsy findings:

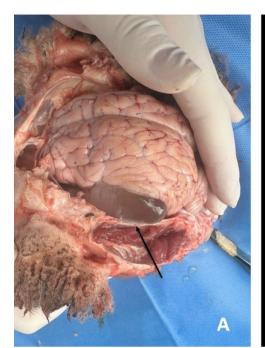
A 2.5-year-old sheep was autopsied. A 5 cm in diameter cyst compressed the right cerebral hemisphere. The overlying membranous skull bone was marked thinned due to compression by the cyst (Fig. 1A). The cyst was formed by a thin transparent membrane and was filled with clear fluid; numerous white protoscolices internally adhered to the cyst membrane (Fig. 1B). Due to the space-occupying effect of the cyst, there was flattening of the cerebral gyri and caudal displacement of part of the occipital lobe of the brain under the tentorial space and into the caudal fossa (subtentorial herniation), with consequent compression of the cerebellum (Fig.2).



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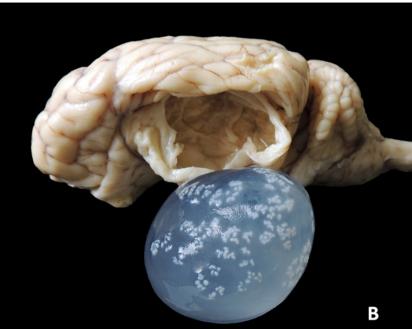


Figure 1. Brain, sheep, coenurosis. A. The right cerebral hemisphere is expanded by a 5 cm in diameter cyst that compresses the brain. At this site (*arrow*), the membranous bone of the skull is thin due to compressive atrophy. B. The pyriform lobe has an empty area from where the parasitic cyst was extirpated. The cyst was formed by a thin transparent membrane and filled with a clear translucent fluid; numerous white scolices adhered internally to the membrane of the cyst.



Figure 2. Brain, sheep, coenurosis. Due to the space-occupying effect of the cyst, there is caudal displacement of part of the occipital lobe (*) under the tentorial space and into the caudal fossa (subtentorial herniation), with consequent compression of the cerebellum. The cerebral gyri are flat.

Follow-up questions?

- Morphologic diagnosis?
- Etiology?
- Name of the condition?

ANSWERS

Microscopic description:

The inner surface of the neuroparenchyma, previously in contact with the cyst, consisted of eosinophilic amorphous material and cellular debris. Beneath this layer, there was a layer of epithelioid macrophages and multinucleated giant cells with a palisade arrangement. The surrounding neuroparenchyma was infiltrated by lymphocytes and plasma cells (Fig. 3). Evaginated from the cyst wall there were numerous parasitic structures (Fig. 4).

• Morphologic diagnosis:

Brain, parasitic cyst with severe, locally extensive, lymphoplasmacytic and granulomatous encephalitis.

• Etiology:

Coenurus cerebralis (larval stage of Taenia multiceps)

Name of the condition:

Coenurosis

Comments:

Coenurosis is a disease of the central nervous system (CNS), primarily of young sheep (6), caused by *C. cerebralis*, the larval form (metacestode) of *Taenia multiceps* (Box 1). The adult parasite inhabits the small intestine of dogs or wild canids, the definitive hosts (9). *T. multiceps* eggs contaminate the environment and are ingested by the intermediate host, which develops the cyst in the brain and, less often, spinal cord. Goats, cattle (5), wild ruminants (10), and humans (1) can occasionally or rarely develop coenurosis (Box 1).

In most cases of coenurosis in southern Brazil, the dog-sheep route of infection is the main transmission pathway (8).

Understanding the life cycle of *T. multiceps* is crucial in assessing the relevance of coenurosis in sheep and establishing strategies for its control (10). The cycle is as follows (Fig. 5):

- 1. The 40 to 100-cm-long tapeworm *T. multiceps* inhabits the intestines of dogs. The mature proglottids containing embryonated eggs (each gravid proglottids contains 32,000-37,000 eggs) leave the intestine and fall onto the grass.
- 2. The grass contaminated with the proglottids and embryonated eggs is ingested by sheep or other intermediate hosts.

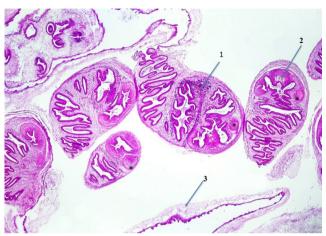
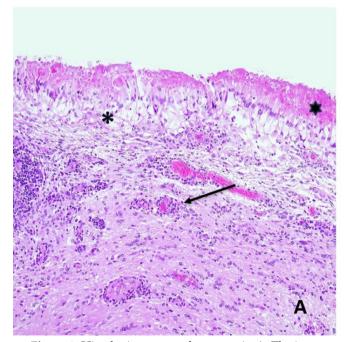


Figure 4. Structure of the scolex. The acoelomate scolex is characterized by the absence of a digestive tract and numerous oval basophilic structures (calcareous bodies) randomly distributed throughout the parenchyma (1). The circular structure composed of muscles with fibers in a radial arrangement are the suckers (*), and part of the rostellum with hooks (2) is at the anterior pole of the scolex. The wall of the cyst consists of two layers (3).



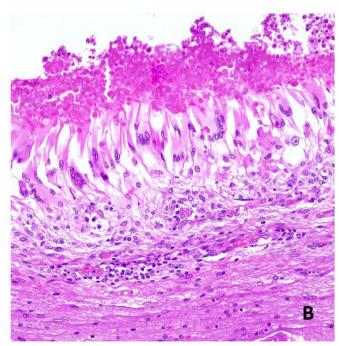


Figure 3. Histologic aspects of coenurosis. A. The inner surface of the neuroparenchyma, previously in contact with the cyst, is covered by eosinophilic amorphous material with cellular debris (*star*). Beneath this layer, there is a discreet layer of epithelioid macrophages (asterisk) and multinucleated giant cells with a palisade arrangement. The surrounding neuroparenchyma is infiltrated by lymphocytes and plasma cells (*arrow*). B. Higher magnification from the image in A.

- 3. In the intermediate host intestine, eggs lose their shell release oncospheres (embryos) into the bloodstream.
- 4. Oncospheres reach the CNS and grow slowly for about 3 months. The mature cysts (*C. cerebralis*) are filled with clear fluid and have a thin, transparent wall containing numerous (400-500) protoscolices.

Box 1. Major information about Taenia multiceps (9)

Class: Cestoda Family: Taeniidae

Length of adult tapeworm: 40-100 cm

Size of the head of the tapeworm: 0.8 mm in diameter.

Number of suckers: 4

Size of the eggs: $29\text{-}37~\mu m$ in diameter Rostellar hooks: two rings of 22-32

Larval form or metacestode: Coenurus cerebralis Condition produced by C. cerebralis: coenurosis Predilection sites: Definitive hosts (small intestine); intermediate hosts (brain and spinal cord)

Definitive hosts: Dog, fox, coyote, jackal, wolf

Intermediate hosts: Sheep, cattle, goat, deer, pig, horse, humans Distribution: worldwide; absent in USA and New Zealand

Life cycle of Taenia multiceps

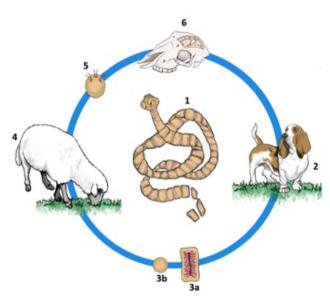


Figure. 5. Cycle of *Taenia multiceps*. The adult *T. multiceps* (1) resides in the small intestine of the definitive host (2). Proglottids(3a) and eggs (3b) or pregnant are shed in the feces of the definitive host into the environment. After ingestion, the eggs hatch in the small intestine of the intermediate host (4) and release the oncospheres (5) that penetrate the intestinal wall and migrate via the bloodstream to the CNS, where the oncosphere encysts and develop over several months into a mature infective *Coenurus cerebralis* in the brain of the intermediate host (6). If dogs ingest infected CNS tissue (1), protoscolices reach the canine intestine and transform into *T. multiceps*, completing the cycle (*Drawing by Dr. Mario Assis Neto*)

5. If dogs ingest infected CNS tissue, protoscolices reach the intestines and transform into adult *M. multiceps*, completing the cycle.

The epidemiology of coenurosis depends on the geographic area of the world where it occurs. In southern Brazil, coenurosis is the most important disease of CNS in sheep, accounting for 25% of all ovine CNS neurologic diseases (7).

Morbidity data are variable. In 16 outbreaks of chronic coenurosis, the authors reported morbidity rates of 0.1%-10%, whereas case fatality rates were 100%. The age of affected animals varied from 5 to 30 months (8).

Two forms of coenurosis can occur in sheep: 1) the acute form, which appears approximately one month after the larval invasion of the CNS, results from traumatic lesions produced the migration of the embryos (oncospheres) of *T. multiceps* in the CNS (2,3,4,6); and 2) the chronic form, associated with the well-formed parasitic cyst-acting as a space-occupying lesion (6), which develops two to four months after infection. In the acute form there is ataxia, muscle tremors, hyperesthesia, hypermetria, and recumbency (2). The chronic form is the most commonly observed, and the main clinical signs are isolation from the flock, depression, blindness, circling, head deviation, and incoordination (10).

Chronic coenurosis is one of the most common nervous diseases of sheep in Brazil (8). However, as in other countries (2,3,4,6), it is infrequently reported and mostly underdiagnosed.

Gross lesions of *acute coenurosis* (6) consist of suppurative meningoencephalitis mainly in the cerebral convexity, the base of the brain, around the optic chiasma, and in the interpeduncular fossa. Meningeal lesions may be the only pathologic change, but in coronal sections of the brain, single or multiple areas of inflammation can occur (6). These tracks mark the trajectory of the oncospheres through the brain, and embryos may be found at the end of the tracks (6).

Histologically (2,6), lesions consist of areas with fibrin, hemorrhage, and neutrophils, which are surrounded by broad bands of degenerated neutrophils lying in a mesh of edematous tissue. These lesions may be bordered by a more predominantly lymphocytic or eosinophilic infiltration. Suppurative changes extend into the brain tissue from the sulci. At the borders between the lymphocytic and the necrotic zones, there may be a palisading of fibroblasts, epithelioid macrophages, and nests of multinucleated giant cells. Areas of necrosis may contain numerous foamy macrophages (6). Evidence of extra-neural migration of oncospheres may be observed as caseous tracts associated to larvae in the kidney, liver, heart, diaphragm, and skeletal muscle (4).

It is estimated that 20 (6) to 35% (4) of lambs that recovered from acute coenurosis develop chronic disease.

Chronic coenurosis occurs in growing sheep 9–18 months of age and is rarely reported in sheep older than 3 years old (10). Clinical signs are a consequence of the

space-occupying effect of the cyst in the brain or spinal cord (10).

Mature cysts (5-6 cm in diameter) can be seen within 7-8 months of the infection in sheep with chronic coenurosis. The distribution of *C. cerebralis* cysts in the brain of infected sheep is primarily parietal and occipital lobes (41.3% of cases), frontal lobe (24% of cases), temporal lobe (17%), and cerebellum (17%) (8). Fewer cases are reported in the spinal cord (6). Hydrocephalus may result from a cyst located in a ventricle (6).

Histologically, the cysts found in chronic consist of two weakly eosinophilic membranes, with dilatations of the internal membrane, from which numerous spherical scolices of C. cerebralis, approximately 1 mm in diameter, emerge (8). The scolices had the parenchyma surrounded by tegument, were acoelomate and lacked a digestive tract. Numerous oval basophilic bodies were observed distributed throughout the parasite parenchyma (calcareous bodies). In some sections, circular structures composed of muscles fibers in a radial arrangement (sucking apparatus, or rostellum with hooks) are at the anterior pole of the scolex. The hooks are birefringent under polarized light. Adjacent to the cyst wall, focally extensive areas of necrosis and infiltrations of epithelioid macrophages in a palisade disposition associated with multinucleated giant cells, and deposition of basophilic amorphous granular material (mineralization) are observed. In the perilesional areas, mild to severe perivascular lymphohistiocytic infiltrate occur amid fibroblasts and vessels. Compression, and edema of the adjacent parenchyma are observed. Foamy macrophages may occasionally be present in large numbers (8).

The clinical signs of coenurosis are not specific and may occur in other neurologic diseases of sheep, including oestrosis (nasal and infection by larval form of *Oestrus ovis*), CNS abscesses, scrapie, neurolisteriosis, congenital copper deficiency, and polioencephalomalacia.

The geographic location, the age of the affected sheep, and the flock history will help in the

differential diagnosis, but confirmation should rely on autopsy and histology.

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