Pathologic Quiz Case
An 86-Year-Old Woman With Draining Sinuses of the Tibia
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The patient was an 86-year-old woman who presented with draining sinuses of the midportion and upper part of the right tibia and fractures of the right tibia and fibula. The osteomyelitis of the tibia had been present for several years, and debridement had been performed. The patient lived on a farm nearly her entire life and had significant contact with goats and sheep. Radiologic examination (plain and magnetic resonance imaging) revealed the presence of extensive complex cystic changes in addition to the fractures. Because of frequent septicemic episodes and the failure of debridement and antibiotic therapy to resolve the condition, a right above-knee amputation was performed. Gross examination of the tibia showed a ragged anterior surface with multiple cavities filled with red-brown necrotic material and soft light-yellow and rubbery white cystlike structures (Figure 1). There was proximal expansion of the tibia and fibula with fractures.

Microscopy revealed acellular laminated membranes with a germinial layer within the medullary cavity of the tibia and fibula. The membranes were further highlighted with the periodic acid–Schiff and Gomori methenamine silver stains (Figures 2 through 4). Rare degenerating scolices and hooklets were sometimes seen. The laminated structures were surrounded by an intense acute and chronic inflammatory reaction that focally destroyed the trabecular and eroded cortical bone.

What is your diagnosis?
Pathologic Diagnosis: Echinococcosis (Hydatid Cyst) of the Bone

Echinococcosis (hydatid disease) is an infection by the larva of the tapeworms of the genus *Echinococcus*. Infections by *Echinococcus granulosus* are the most common, but *Echinococcus multilocularis* and *Echinococcus oligarthrus* also infect humans. In North America, the adult worm of *Echinococcus* lives in the small intestine of dogs, coyotes, and wolves. The gravid segment of the adult tapeworm breaks off and disintegrates in the large bowel, releasing hundreds of infective eggs, which then pass out with the feces. Sheep, goats, deer, moose, and other animals and humans acquire the larva stage by ingesting the egg. The oncosphere hatches in the small intestine and reaches tissue by the hematogenous route. Once in the tissue, the larva secretes a hyaline membrane and becomes surrounded by it. This membrane eventually differentiates into an outer acellular laminated structure and an inner germinal layer. The inner surface of the germinal layer gives rise to protoscolices (the stage infective to the definitive host).

Humans are almost always intermediate hosts of hydatid disease, and the cyst develops mainly in the liver (60%), lungs (20%), and brain (3%). The hydatid cyst requires several months to mature, and when fully grown, it can contain thousands of protoscolices. For the life cycle to be completed, the hydatid cyst and its content must be ingested by a canine host. In the United States, human infections are rare, with occasional cases seen among Indians in the southwestern United States and Indians and Eskimos in the northern regions of North America. This patient had only bone disease, although involvement of the bones is rare (1% to 2% of cases). The most common site for cases with bone disease is the lower vertebrae, accounting for approximately 50% of these cases.

The clinical manifestations of hydatid disease are due to the space-occupying nature of the slowly expanding cysts and thus are directly related to the number, anatomical location, and rate of growth of the cysts. Rupture of cysts may occur in 20% of cases, producing fever, urticaria, and occasionally anaphylactic shock due to the release of antigenic cyst contents. Cyst rupture may also lead to dissemination of infection due to the release of thousands of protoscolices, each with the capability to produce a new cyst. The cysts of the bone may result in pathologic fractures, and cysts in the lower vertebrae may lead to spinal cord compression and paraplegia. Over time, cysts may die and become calcified.

Diagnosis of hydatid disease is difficult and depends primarily on clinical, radiologic, and serologic findings. Obtaining an accurate case history is essential to the diagnosis of hydatid disease. Ownership of dogs, life on a farm even during childhood, and travel to endemic areas are important aspects of history that point to the possibility that a patient might have hydatid disease. Definitive diagnosis is made by identifying the hydatid cyst that is usually first detected by radiologic studies. Although the aspiration of cyst contents and demonstration of hydatid sand (consisting of free daughter cysts and free scolices) may be diagnostic, it is contraindicated because of the risk of anaphylaxis and dissemination of infection. Serologic testing may be useful, but results are negative in 10% to 40% of infections. Histopathologically, the demonstration of cyst wall with hematoxylin-eosin stains establishes the diagnosis. The walls are finely laminated and amorphophilic, and their appearance is so characteristic that a diagnosis can be made even when only a small fragment is identified in a section. Surgical excision of hydatid cysts is the therapy of choice for symptomatic patients. If surgery is contraindicated, medical therapy with high-doses of albendazole or praziquantel may be effective.

References