Title

Soft Tissue Echinococcosis

Journal

Clinical Orthopaedics and Related Research, 385(&NA;)

ISSN

0009-921X

Authors

Cannon, Christopher P
Nelson, Scott D
Panosian, Claire B
et al.

Publication Date

2001-04-01

DOI

10.1097/00003086-200104000-00029

Peer reviewed

CLINICAL ORTHOPAEDICS AND RELATED RESEARCH
Number 385, pp. 186–191

Soft Tissue Echinococcosis

A Report of Two Cases and Review of the Literature

Christopher P. Cannon, MD*; Scott D. Nelson, MD**; Claire B. Panosian, MD†; Leanne L. Seeger, MD††; Frederick R. Eilber, MD§; and Jeffrey J. Eckardt, MD*

Echinococcosis (hydatid cyst disease) is a zoonotic infection caused by the parasitic tapeworm Echinococcus. The larval stage of this parasite can implant in many organs of the body, most commonly the liver, and create internal budding cystic masses. Echinococcal cysts also can implant in soft tissues; however, a review of the literature revealed no published case with the patient initially presenting with a soft tissue mass. Two such cases are reported in the current study. Physicians who evaluate soft tissue masses, particularly in patients from Echinococcus-endemic areas, need to include echinococcosis in their differential diagnoses. The current treatment of choice for soft tissue echinococcosis is wide resection combined with perioperative medical therapy.

From the Departments of *Orthopaedic Surgery; **Pathology and Laboratory Medicine; †Infectious Diseases; ††Radiological Sciences; and §Surgery, University of California, Los Angeles School of Medicine, Los Angeles, CA.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article. No funds were received in support of this study.

Reprint requests to Jeffrey J. Eckardt, MD, Department of Orthopaedic Surgery, University of California, Los Angeles School of Medicine, 10833 Le Conte Avenue, CHS 76-134, Los Angeles, CA 90095.

Received: March 22, 2000.

Revised: July 28, 2000.

Accepted: August 22, 2000.

Echinococcosis (hydatid cyst disease) is a zoonotic infection most commonly caused by the larvae of Echinococcus granulosus, and the larvae of Echinococcus multilocularis, Echinococcus vogeli, and Echinococcus oligarthrus.[8,12] Adult tapeworms of these species live in the small intestine of carnivorous canines, usually dogs. Tapeworm eggs passed in the feces of definitive hosts subsequently are ingested by grazing animals, usually sheep. Humans who inadvertently ingest tapeworm eggs after contact with infected dogs also can serve as intermediate hosts.

Tapeworm eggs hatch in the gastrointestinal tract of the intermediate host and release oncospheres, which penetrate the intestinal mucosa and enter the general circulation. The oncospheres disseminate to various tissues, where they implant and develop into hydatid cysts. Most (60%) cysts develop in the liver; other sites of development are the lung (20%), and less frequently the spleen, heart, kidney, central nervous system, bone, and soft tissue.[2,7,11] The life cycle is completed when the dog or other definitive host ingests a hydatid cyst in the infected viscera of the intermediate host.

Echinococcosis is widespread throughout sheep-producing regions of southern Europe, Asia, Australia, Africa, and the Middle East. Although rare in North America, imported and

autochthonous cases have been reported in Utah, California, Arizona, New Mexico, the Mississippi Valley, and Canada.[7] In one recent series from southern California, 25 of 29 (89%) patients with disease diagnosed at two teaching hospitals were foreign born, primarily from the Middle East.[5] In the current report, two foreign-born patients with soft tissue echinococcosis presenting to the authors' institution are described.

CASE REPORTS

Case 1

A 29-year-old man, who recently emigrated from Turkey, had pain and swelling in his left gluteal region after a fall while playing volleyball. The pain from the fall slowly resolved, but the mass persisted. A computed tomography (CT) scan was obtained that revealed the presence of a large, multilobulated mass of fluid density between the gluteus maximus and medius muscles (Fig 1). There was enhancement of the periphery of the mass and internal septations, with an appearance of cysts within cysts. The differential diagnoses included echinococcal cyst disease, tuberculous abscess, and myxoid liposarcoma. A fine needle aspiration yielded 40 mL of fluid with negative Gram stain, culture, and microscopic examination. The mass persisted, and the patient subsequently underwent surgical excision of the mass. Intraoperative spillage of the cyst occurred, which was managed with copious irrigation. No adverse clinical effects occurred at the time of cyst spillage. Final pathologic examination revealed echinococcal cysts. An ultrasound of the liver obtained after surgery revealed a 4×5 cm cyst consistent with echinococcal infection. After surgery, the patient received a 4-week course of praziquantel and several 4-week cycles of albendazole. The patient later moved to another state to complete his graduate studies. Fifteen months after surgery, written communication with the patient's infectious disease physician indicated the patient was doing well with no local recurrence. At the time of most recent communication, the patient's liver cyst had not been surgically addressed.

Case 2

A 53-year-old man, who was raised on a farm in the Basque region of Spain and emigrated to the United States at 19 years of age, presented with a painless mass in his left calf of 8 months' duration. Radiographs revealed no bony lesions; magnetic resonance imaging (MRI) revealed a large, multicystic calf mass within the soleus muscle (Fig 2). Results of the remaining workup, including chest radiograph, bone scan, complete blood cell count, erythrocyte sedimentation rate, and serum chemistry tests, were normal except for mildly elevated liver enzyme levels. A CT-guided biopsy was done that revealed echinococcal cyst wall and characteristic hooklets of an embryonic tapeworm scolex. Chest and abdominal CT scans were done to iden-

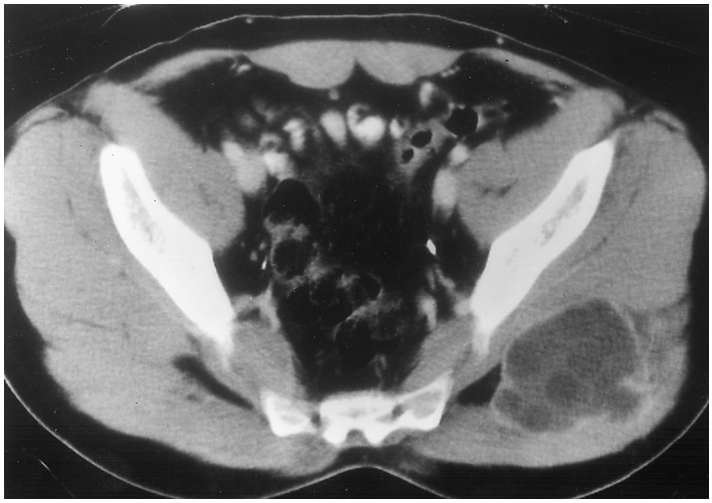

Fig 1. Contrast enhanced CT scan, soft tissue window. A large, multilobulated mass of fluid density is present between the gluteus maximus and medius muscles. There is intense enhancement of the rim.

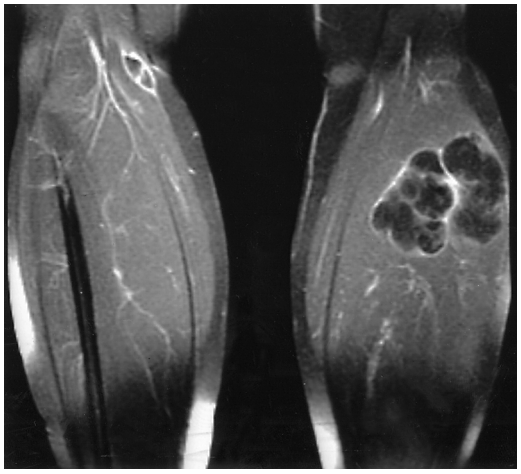

Fig 2. Magnetic resonance imaging scan showing a multicystic calf mass within the soleus muscle. Coronal fat-supressed, T1-weighted image (SE 600/20) after intravenous gadolinium. There is very subtle enhancement of a portion of the mass centrally, and there is enhancement of the rim.

tify possible lung or liver cysts. No such cysts were found. The patient was placed on albendazole 400 mg for two 1-month cycles and underwent wide resection of the calf lesion, ensuring no cyst spillage (Fig 3). The biopsy tract was removed in continuity with the specimen. Pathologic examination confirmed the diagnosis of echinococcal cyst (Figs 4, 5). After surgery, the patient was treated with an additional 1-month course of albendazole. The patient's perioperative and postoperative courses have been unremarkable, with no immediate anaphylaxis or soft tissue recurrence by physical examination at 6 months.

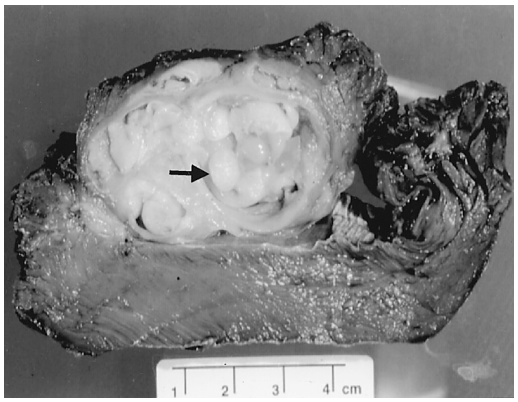

Fig 3. Gross photograph showing resected specimen of hydatid cyst with numerous daughter cysts (arrow).

DISCUSSION

Echinococcosis (hydatid cyst disease) is widespread throughout sheep-producing regions of southern Europe, Asia, Australia, Africa, and the Middle East. However, it is a relatively rare disease in the United States, and in at least one series, most commonly was found in foreign-born patients.[5] In humans, the liver is the most common site of cyst development (60%), followed by the lungs (20%), and much less commonly, the kidney, spleen, brain, and soft tissue.[2,7,11] Infections involving bony sites, including the spine, pelvis, and hip, also have been reported.[1,10,14] The possible occurrence of soft tissue echinococcosis is mentioned in pathology texts and review articles.[7,11] However, a review of the English language literature from the last 25 years failed to reveal reports of echinococcosis presenting as a soft tissue mass. Thus, it is impossible to characterize the typical presentation of soft tissue echinococcosis. In the two cases presented, a painless mass led the patients to seek medical attention.

The diagnosis of echinococcosis usually is based on the identification of a hydatid cyst in tissue. Imaging modalities such as ultrasound, CT, or MRI may reveal a calcified cyst wall and microcalcifications within daughter cysts and varying fluid densities between cysts and surrounding organs.[7] Accurate diagnosis by CT was made in 96% of 157 patients with hepatic or other parenchymal cysts. Computed tomography or ultrasound-guided needle biopsies also are helpful in diagnosis.[3,6,9] Concerns over microscopic spillage at the time of needle biopsy do not appear warranted, especially if patients receive subsequent medical treatment and biopsy tracts are resected at the time of surgery.[3,12] Various serologic tests can assist in diagnosing echinococcosis, including latex

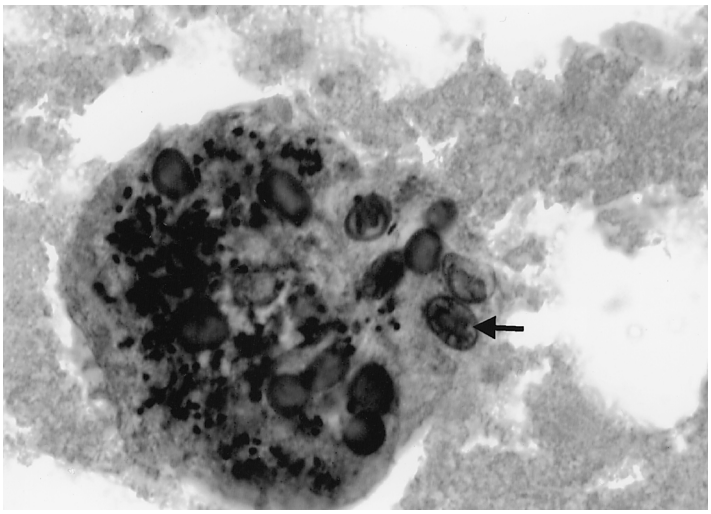

Fig 4. Echinococcal daughter cyst with daughter embryonic forms (arrow) (Stain, hematoxylin and eosin; original magnification, ×100).

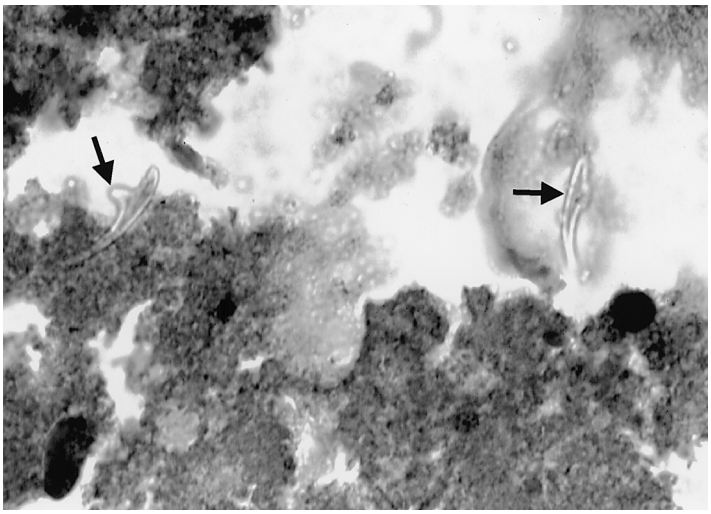

Fig 5. High power view showing hooklets (arrows) present within amorphous debris (Stain, hematoxylin and eosin; original magnification, ×1000).

agglutination, indirect hemagglutination assays, enzyme-linked immunosorbent assays, enzyme-linked immunodiffusion assays, and immunoblotting.[7] However, serology alone is insufficient to diagnose echinococcosis.[7,8] False negative test results occur in as many as 50% of patients with solitary lung cysts.[8] The sensitivity and specificity of serologic tests in patients with soft tissue echinococcosis is not known.

In Patient 1, the cyst within a cyst appearance on CT scan was consistent with echinococcosis. However, initial fine needle aspiration did not reveal hydatid cyst structures. After the diagnosis was confirmed by final surgical pathologic examination, a second review of the fine needle aspiration confirmed the presence of cyst wall in the original specimen. Serologic studies were not done on the patient. In Patient 2, the multicystic appearance to the patient's calf mass on MRI was considered most consistent with a soft tissue

sarcoma, and echinococcocis was not considered in the initial differential diagnosis. The CT-guided biopsy did show hydatid cyst wall and characteristic hooklets, thereby establishing the diagnosis.

Complete surgical resection, including the biopsy tract, and perioperative medical therapy is the preferred treatment for isolated echinococcosis.[7] At the time of surgery, rupture or spillage of cysts should be avoided to prevent local or distant dissemination and immediate anaphylaxis. Cyst spillage did occur in Patient 1 but was managed with copious irrigation and subsequent medical therapy using praziquantel and albendazole. Anaphylaxis was not observed. The overall recurrence rate for surgical treatment of hydatid cyst disease is from 2% to 10%.[8] The recurrence rate in soft tissue disease is not known.

Historically, surgical management of echinococcosis has included sterilization of cyst contents using protoscolicidal chemical solutions such as formalin, ethanol, or hypertonic saline injected into the cyst just before excision. However, the efficacy of these protocols has never been proven, and given the known potential complications of these agents, the practice of perioperative injection of cysts should not be done as a routine procedure.[8,12]

A more important development in recent years has been the growing use of imidazole compounds such as albendazole and mebendazole either as sole or perioperative adjunct therapy in echinococcosis. Treatment with the antihelminthic albendazole (10 mg/kg/day for multiple periods of 4 weeks with intervals of 14 days) or mebendazole (40 to 50 mg/kg/day continuously) has resulted in significant regression of cyst size in 40% to 70% of patients and a complete cure in as many as ⅓ of patients.[4] The drugs also have proven to be an effective adjunct to surgery, allowing safer manipulation of the cysts and minimizing the chance of secondary recurrence in the event of an intraoperative spillage.[12,15] Because of its superior pharmacokinetics, albendazole is the preferred drug.[8,12,13] Although less well studied in humans, weekly doses of praziquantel 40 mg/kg also may prevent recurrence of echinococcosis after intraoperative spillage.[12]

Given its relative rarity, making the diagnosis of soft tissue echinococcosis requires a high index of suspicion. Based on the authors' experience, the infection should be considered in the differential diagnosis of any soft tissue mass that occurs in a foreign-born patient with past exposure to sheep producing regions. Recommendations for the management of hydatid cyst disease should be in a manner analogous to a tumor. Radiologic imaging and CT-guided biopsy should safely and accurately establish the diagnosis. Additional workup must be done to exclude concurrent disease in sites such as the liver and lung. Wide resection, with careful attention to include the biopsy tract and avoid cyst spillage, minimizes the risk of recurrence or an immediate anaphylactic reaction. Preoperative and postoperative treatment with albendazole also is beneficial, minimizing the risk of recurrence and allowing safer handling of the cyst at the time of surgery.

References

1. Agarwal S, Shah A, Mohammed S, Rooney R: Hydatid bone disease of the pelvis. Clin Orthop 250:251–255, 1992.
2. Booz MK: The management of hydatid disease of bone and joint. J Bone Joint Surg 54B:698–709, 1972.
3. Bret PM, Fond AF, Bretagnolle M, et al: Percutaneous aspiration and drainage of hydatid cysts in the liver. Radiology 168:617–620, 1988.
4. Davis A, Dixon H, Pawlowski Z: Multicentre clinical trials of benzimidazole carbamates in human cystic echinococcosis (phase 2). Bull World Health Organ 67:503–508, 1989.
5. Donovan SM, Mickiewicz N, Meyer RD, Panosian CB: Imported echinococcosis in southern California. Am J Trop Med Hyg 53:668–671, 1995.
6. Filice C, Di Perri G, Strosselli M, et al: Parasitologic findings in percutaneous drainage of human hydatid liver cysts. J Infect Dis 161:1290–1295, 1990.
7. Fraiji EK, Connor DH: Echinococcosis. In Connor DH, Chandler FW (eds). Pathology of Infectious Diseases. Vol 2. Stamford, Appleton & Lange 1405–1413, 1997.
8. Kammerer WS, Schantz PM: Echinococcal disease. Infect Dis Clin North Am 7:605–618, 1993.
9. Munzer D: New perspectives in the diagnosis of echinococcus disease. J Clin Gastroenterol 13:415–423, 1991.
10. Rao S, Parikh S, Kerr R: Echinococcal infestation of

the spine in North America. Clin Orthop 271:164–169, 1991.
11. Schaefer JW, Khan MY: Echinococcosis (hydatid disease): Lessons from experience with 59 patients. Rev Infect Dis 13:243–247, 1991.
12. Schantz PM: Echinococcosis. In Guerrant RL, Walker DH, Weller PF (eds). Tropical Infectious Diseases. New York, Churchill Livingstone 1005–1025, 1999.
13. Szypryt EP, Morris DL: Combined chemotherapy and surgery for hydatid bone disease. J Bone Joint Surg 69B:141–144, 1987.
14. Voutsinas S, Sayakos J, Smyrnis P: Echinococcus infestation complicating total hip replacement. J Bone Joint Surg 69A:1456–1458, 1987.
15. Wilson JF, Rausch RL, McMahon BJ, Schantz PM: Parasiticidal effect of chemotherapy in alveolar hydatid disease: Review of experience with mebendazole and albendazole in Alaskan Eskimos. Clin Infect Dis 15:234–249, 1992.