Recurrent spinal hydatidosis in North America

Case report and review of the literature

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Spinal hydatidosis is an uncommon manifestation of the parasite *Echinococcus*, affecting fewer than 1% of patients with hydatid disease. The authors report on a 34-year-old Turkish woman who presented with recurrent primary spinal hydatid disease. The patient originally presented with progressive numbness and paraparesis that was reversed after T5–6 laminectomy and cyst removal. Pathological findings indicated parasitic infection and she underwent treatment for cysticercosis. Nevertheless, she returned 4 years later with back pain, numbness, and monoparesis. Neuroimaging studies revealed spinal cord compression with multiple cysts that were again resected. Pathological findings were consistent with *Echinococcus*.

Although this disease is uncommon, particularly in North America, the authors conclude that spinal hydatidosis should be considered in the differential diagnosis of any patient who has lived or traveled within endemic areas and who presents with spine lesions and cord compression. The authors review the literature pertaining to the epidemiological features, presentation, diagnosis, neuroimaging characteristics, recommended treatments, and overall prognosis of spinal hydatidosis.

**KEY WORDS** • hydatid cyst • *Echinococcus* • spinal infection • myelopathy

Hydatidosis affecting the spine comprises less than 1% of the total cases of hydatid disease and is particularly uncommon in North America. Often the diagnosis is difficult to make by either neuroimaging or immunohistochemical modalities, yet successful management relies on precise diagnosis in combination with appropriate and thorough treatment. Without this combination, recurrence is generally predictable. We report such a case of recurrent hydatid disease of the thoracic spine, which was partly due to misdiagnosis and, consequently, inadequate treatment.

**CASE REPORT**

**History and Examination.** This 34-year-old Turkish woman presented on March 21, 2000, with a 4-month history of tingling in both feet, numbness from her waist to her ankles, and bilateral lower-extremity weakness. She reported back pain, a burning sensation in her hips, and sore calf muscles. On examination, she had mild paraparesis with a sensory level at T-5 as well as tenderness at that level, 1+ bilateral patellar and Achilles reflexes, and an unsteady gait.

**Neuroimaging.** Admission MR imaging demonstrated an epidural mass at T-5 that was projecting 1.5 cm into the spinal canal and eroding the left pedicle and portions of the body, lamina, and proximal T-5 rib head (Figs. 1 and 2). At least five separate cystic areas were evident. The principal working diagnosis was spinal cord tumor.

**Operation and Subsequent Treatment.** Surgery involved a thoracic laminectomy of T5–6 and resection of spinal cord and T-5 nerve root decompression. The surgical specimen measured 1.5 × 2 cm and was described as a whitish, pearlike, semitranslucent, cystic material, which was thought to be parasitic. Results of pathological testing were not definitive, but indicated there were features of hydatid cyst (*Echinococcus granulosus*), or cysticercosis. The decision made at that time was to treat her according to the presumed diagnosis of cysticercosis.

**Repeated Presentation and Examination.** The patient returned in early 2004; her chief symptom was back pain that had continued for 3 months, associated with numbness in her left breast, axilla, and bilateral lower extremities. Clinical findings included 4/5 motor strength in the right lower extremity causing a limping gait. She had a limited range of motion because of pain.

**Findings on Repeated Neuroimaging.** On repeated MR imaging and CT scanning, a 3.8 × 1.8 × 3-cm multiloculated cystic structure was demonstrated at the T4–5 levels (Figs. 3 and 4). The preoperative diagnosis was an epidural paracystic mass at T-5.

**Second Operation and Postoperative Course.** Surgery involved an extracavitary approach with excision of the
head of the fifth rib and the transverse process for resection of the epidural mass at T-5. The mass was composed of multiple cysts of varying sizes containing scoles (Fig. 5). Postoperatively, the patient was able to move all four extremities well. When she was discharged home in 7 days, she was ambulatory and had complete resolution of her numbness. Her discharge diagnosis was spinal hydatidosis consistent with Echinococcus. Her follow-up visits have been routine, with no recurrence of her symptoms; she is being followed closely in the Infectious Disease Clinic. She is currently receiving a minimum 6-month course of albendazole.

**DISCUSSION**

Hydatid disease is caused by two forms of the parasite *Echinococcus: E. granulosus*, and less commonly *E. multilocularis*, the latter primarily causing alveolar echino-
coccosis. Bidloo* reported osseous hydatidosis in 1708, in a case involving the humerus. In 1807, Chaussier* reported the first case of spinal hydatid disease, and Reydellet* is believed to have performed the first surgical intervention for spinal hydatidosis in 1819. Lloyd* reported the first North American case of vertebral hydatidosis in 1896.

The definitive hosts of *Echinococcus* are dogs, wolves, and other carnivorous animals, in which the adult parasite lives within the intestine and the ova are subsequently passed in the stool. Intermediate hosts, such as sheep, cattle, horses, and hogs, ingest the ova, which then hatch into embryos (hexacanth) in the duodenum. The embryos reproduce asexually and form multiloculated cysts. Humans contract the disease by contamination through direct contact with the definitive host or its feces, or by ingesting food infected with ova.

The parasite is most commonly found in livestock-raising areas in Mediterranean countries, Africa, South America, New Zealand, and Southern Australia, but has a worldwide distribution. Although rare in North America, cases have been reported in California, Utah, the Lower Mississippi valley, Alaska, and northwest Canada. In countries where the disease is endemic, it is a major public health concern, a fact reflected by the 21,303 cases of hydatid disease reported between 1987 and 1994 in Turkey.

The most common sites of infection are the liver (75%), lungs (15%), and brain (2–4%). Bone involvement is uncommon, but when present it affects the vertebrae 44% of the time. In total, however, only 0.5 to 1% of hydatid disease cases involve the spine. Vertebral hydatid disease is more likely to be caused by a primary process than a secondary one, although there have been case reports of local spinal invasion from pulmonary hydatidosis. Spinal involvement has been classified by Braithwaite and Lees into five types: 1) primary intramedullary hydatid cyst; 2) intradural extramedullary hydatid cyst; 3) extradural intraspinal hydatid cyst; 4) hydatid disease of the vertebrae; and 5) paravertebral hydatid disease. Of these five, the first three types are considered rare. In our review of 232 reported cases, the vertebral level involved was documented in only 129 patients, establishing thoracic involvement as the most common level (52%), followed by lumbar (37%), and then cervical and sacral involvement (5.5% each). There was one case of thoracic involvement stemming from the seventh rib.

Spinal involvement is believed to occur through vertebral–portal venous anastomosis. Infestation of the spine is described to progress as a multivesicular infiltration of cancellous bone that involves the vertebral bodies, pedicles, and laminae to varying extents. The intervertebral discs, however, are usually spared because the cyst growth is confined within the periosteum.

Histologically, *E. granulosus* can be identified by its multilayered cyst wall containing hooklet-bearing scolices. The thick outer laminated wall may calcify and is composed of layers of chitin. The innermost germinal layer produces hydatid fluid and may contain numerous embryonal scolices termed “hydatid sand.”

A review of reported cases of spinal hydatidosis reveals a greater rate of infection in men (62%) than in women.
The patients’ age ranged from 6 to 74 years, with a mean age of 36 years. This disease is found primarily in adults because of its slow progression; however, cases in children have been reported. The most common signs and symptoms include paraparesis (62%) or paraplegia (26%), back or radicular pain (55%), sensory loss or disturbance (36%), and sphincter disturbance (30%).

Diagnosis is usually difficult, and often is not made...
techniques have been used to confirm the diagnosis. Use of sorbent assay to detect serum levels of indirect hemagglutination or the enzyme-linked immunosorbent assay may be difficult, the best approach probably consists of a detailed history and clinical evaluation of the patient combined with precise neuroimaging. The neuroimaging methods of choice are CT scanning and MR imaging. Plain x-ray films are nonspecific and can be dangerous because of possible dissemination of the disease intradurally; it is also limiting because it only shows the mass effect on the dural sac. The CT scanning modality is useful for clearly demonstrating destructive bone changes and paraspinal soft-tissue involvement. It has an advantage over MR imaging in demonstrating subtle osteolytic changes, but CT scanning is limiting because it cannot be used to distinguish between cystic lesions and the dural sac. Multiple cysts create additional difficulties, because larger cysts may disguise smaller ones. The CT modality is most useful combined with either MR imaging or myelography.

Magnetic resonance imaging is the best single neuroimaging modality for precise determination of the anatomical relationship of the cystic lesions and their exact level, as well as allowing imaging of the entire neuraxis. Fahl, et al., have described the characteristics of hydatid cysts on MR images, reporting that cysts generally have two dome-shaped ends, have no debris in the lumen, and usually look like flattened sausages, with thin, regular walls without septations. Intradural cysts may be single or multiple; extradural cysts are always multiple and involve the bone. The T1-weighted MR images are particularly sensitive for identifying cystic lesions and their relationships to surrounding structures. Tekkok and Benli, using proton density–weighted images, were able to characterize a viable cyst as one containing low-intensity fluid with iso- to mildly hyperintense cyst walls. A dead cyst was identified by a decrease in hyperintensity and an increase in hypointensity from collapsed cyst walls. Postoperative MR imaging is invaluable for determining the completeness of resection as well as for documenting any recurrent or newly developing cysts.

Surgery is the most common initial treatment for hydatid disease, with total removal of the cyst(s) the primary goal. For spinal hydatidosis, laminectomy with simple decompression is used most frequently, yet it is not without accompanying risks. Complications often arise due to excessive local mass effect on critical neural structures, combined with a cyst growth rate of up to 7 mm per month. Also, the cyst’s thin walls may easily rupture, either spontaneously or due to trauma or surgery, resulting in recurrence of multiple cysts, or the development of an anaphylactic reaction secondary to contact with intracystic fluid. Rupture occurs most frequently in spinal hydatidosis because the cysts are contained in narrow spaces within bone boundaries, making removal particularly difficult. Rupture rates have been reported to be as high as 44.4%, particularly if the cyst is in an extradural location. No specific surgical technique has been found to avoid this problem completely, although the operating microscope can be helpful.

Chemotherapy is often used in conjunction with surgery to prevent recurrence and to protect the patient from dissemination of a ruptured cyst. Albendazole and mebendazole are the two most commonly used anthelmintic drugs. Studies have indicated that albendazole is more effective because it is better absorbed and has increased efficacy. Albendazole acts by blocking glucose uptake and depleting the glycogen stores of the parasite. No long-term studies have documented potential side effects of albendazole or mebendazole, although albendazole may cause a temporary, minor elevation in liver enzymes that usually does not require discontinuation of the drug if the patient’s condition is closely monitored. The appropriate duration of antibiotic therapy has not been established, but studies show a mean recommended course duration of 3 to 4 months. In one study investigators suggested at least 1 year of albendazole therapy after neurodecompression.

Reports also indicate that irrigating the wound with hypertonic saline or a diluted Betadine solution after cyst removal helps osmotically destroy and disrupt the parasites, although this remains unproven. In a few studies, researchers have detailed treatment alternatives for inoperable cases or patients in whom surgery is contraindicated. Lam, et al., describe a case of inoperable vertebral hydatidosis treated with albendazole and praziquantel that resulted in a reversal of imminent paralysis. Patients have also been reported to have been successfully treated using CT-guided needle aspiration and hypertonic saline irrigation.

Recurrence remains a major problem in spinal hydatidosis; the literature cites rates of 30 to 100%. Given this
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high rate of recurrence, spinal hydatidosis has a poor prognosis, and has been compared to spinal malignancy.18 Reported mortality rates vary, with a range from 3% to more than 50%.18,24,49 In one study researchers suggest a mean life expectancy of 5 years after onset of spinal involvement.22 Nevertheless, with improved neuroimaging, aggressive resection, and more extensive experience with current anthelmintic drugs, the recurrence rates may decline.

CONCLUSIONS

We report on a patient with a recurring thoracic extradural hydatid cyst; the recurrence was presumably due to misdiagnosis and therefore inadequate treatment. A literature review concerning presentation, diagnosis, neuroimaging, treatment, and prognosis of spinal hydatidosis is presented. Hydatid disease of the spine is rare, especially in the US, but should be considered in the differential diagnosis of patients presenting with progressive myelopathy secondary to intraspinal masses and who have had contact with areas in which hydatidosis is endemic. The diagnosis must be clearly established, appropriate surgical therapy instituted, and anthelmintic chemotherapy administered with vigilant follow-up review maintained to achieve optimal and lasting results.

References

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