

Accidentally diagnosed multiple intradural extramedullary spinal hydatidosis in a young adult: a case report and review of the literature

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Abstract

In this paper, we report a rare case of asymptomatic multiple intradural, extramedullary spinal hydatidosis; accidentally diagnosed in a patient with signs and symptoms of a true protruded disc. Although quite rare, vertebral hydatidosis should always be considered as a differential diagnosis for spinal presentations, particularly in endemic areas.

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Key words: Echinococcus granulosus, Hydatid cyst, Spinal hydatidosis

Abstract
In this paper, we report a rare case of asymptomatic multiple intradural, extramedullary spinal hydatidosis; accidentally diagnosed in a patient with signs and symptoms of a true protruded disc. Although quite rare, vertebral hydatidosis should always be considered as a differential diagnosis for spinal presentations, particularly in endemic areas for echinococcosis.

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Introduction

Being a Greek term, hydatid means “watery cyst”. Hydatid disease is a zoonotic infection mainly caused by the larva form of *Echinococcus granulosus* ; a helminth of the Cestoda class¹²³. Echinococcosis is one of the 20 Neglected Tropical Diseases (NTDs) listed by the World Health Organization (WHO) and affects more than one million people worldwide imposing over three billion dollars on healthcare systems each year⁴. *Echinococcus granulosus* mainly affects the liver and lungs. Skeletal involvement is quite rare and occurs in only 0.2 to 1 % of all patients, while 50% of these cases are spinal hydatidosis⁴. In this paper, we aim to present a rare case of incidentally diagnosed multiple spinal hydatidoses and give a review of the literature about the disease.

Case Presentation

A 26-year-old male presented with low back pain that radiated to the lower limbs accompanied by bilateral anterior thigh hypoesthesia. The symptoms developed gradually one year ago and did not improve with medical treatments. Three years ago, he had cardiac surgery due to cardiac hydatidosis. He took albendazole for about one year and a half afterwards. He had no history of contact with dogs and livestock and no complaint of coughs, weight loss, night sweats, or other constitutional symptoms.

At examination, the force of the proximal lower limbs was 4/5 and the distal force was 5/5. Deep tendon, bulbocavernosus, and superficial anal reflexes were all intact. There was no sphincter dysfunction or hyperesthesia in the perineal area. He had hypoesthesia at L1/L2 dermatomes bilaterally. All other examinations The biochemical and hematological profiles were within normal limits and ESR and CRP levels were not elevated. Abdominopelvic ultrasonography and computed tomography (CT) scans of the chest and abdomen revealed no significant findings. In the next step, lumbosacral magnetic resonance imaging (MRI) was performed. As was predictable from the patient’s symptoms, there was an extruded disc at the T12/L1 level (Figure 1A). Surprisingly, we found three well-demarcated cystic lesions with different diameters at the level of the T12 vertebral body. Two other lesions with the same characteristics were present at the level of the L4/L5 vertebral bodies (Figure 1B). The lesions exhibited iso-signal intensity on T1-weighted and hyper-signal intensity on T2-weighted images. The cystic lesions were all intradural and possibly extramedullary at both levels. There were no signs of cord compression or bone destruction. MRI with intravenous contrast showed no enhancement.

With the pre-operation diagnosis of an extruded T12/L1 disc and multiple spinal hydatid cysts, the patient underwent surgery in two stages. During surgery, the L4/L5 laminectomy was performed, the dura was opened and two white pearl-like cystic masses were gently removed in order to avoid cyst rupture (Figure 2). The whole surgery field and surrounding regions were irrigated with 3% hypertonic saline. The wound was then stitched in layers.

At the next stage of the procedure, one week later, a T12/L1 discectomy and Pedicular Screw Fixation (PSF) for T12 and L1 vertebrae were performed. So, the dura was opened and three white intramedullary cysts were excised without rupture. The site was then washed with hypertonic saline. At that point, the wound was closed under continuous hypertonic saline irrigation.

As a result of the operation, the patient’s neurological symptoms improved, and he was discharged with anthelmintic treatment of albendazole 400 mg BD for the next six months.

At the follow-up session four months later, the patient was symptom-free and the neuraxis MRI showed no recurrence or remnant of the CNS hydatidosis (Figure 3).

Discussion

Occurring in all age groups and in both sexes, cystic echinococcosis (CE) accounts for a substantial disease burden globally. There is an estimated 2.2% death rate post operation and unfavorably the relapse rate is about 6.5%⁴⁵.

Cystic Echinococcosis is predominantly reported in sheep-raising areas while being considered highly endemic in the eastern part of the Mediterranean region, central Asia, southern and eastern Europe, northern Africa, southern America, Siberia and western China. This disease is not reported in Antarctica and has been eliminated through comprehensive control programs in New Zealand, Iceland, the Falkland Islands, Tasmania, and Cyprus⁶⁴.

Human beings act as accidental intermediate hosts for *Echinococcus granulosus*. Adult worms mature in the intestine of carnivorous animals such as dogs and wolves as definitive hosts, and the eggs are shed in their stool. Medium-sized herbivores, such as sheep and cattle ingest the eggs. Human beings contract the disease through the direct contact with the carnivorous animals and their feces or by ingestion of food infected with parasite eggs. Once ingested by the human or any other intermediate hosts, the oncosphere(true larva) hatches from the egg and burrows into the intestinal submucosa, then migrates through veins or lymphatic vessels to internal organs such as liver¹⁴.

The human liver acts as an effective barrier for most of the larvae. However, some may pass through the liver and enter the right side of the heart and then the lungs. If the larva is not lodged in the liver or lungs, it may virtually embed anywhere in the body, such as the spleen, peritoneum, heart, kidney, brain, spine, skeletal bones and muscles¹³. Ninety percent of the larvae are eliminated by the host reaction. However, if they survive, the metacestode or hydatid cyst develops in the affected organ over a course of years⁷.

Commonly involved organs are the liver (75%), lung (15%), brain (2–4%), and the genitourinary tract (2–3%)⁸. Only 0.2 to 1% of patients experience bone involvement, and about half of which occurs in the spine⁹. *Echinococcus* mainly infects the thoracic spine (52%), followed by the lumbar (37%) and then the cervical and sacral spine¹¹⁰.

Braithwaite and Lees had classified spinal hydatid lesions into five groups:(a) primary intramedullary hydatid cyst, (b) intradural extramedullary hydatid cyst, (c) extradural intraspinal hydatid cyst, (d) hydatid disease of the vertebra, and (e) paravertebral hydatid disease¹¹.The first three groups are quite uncommon. Comparing to extradural lesions, intradural hydatidosis mostly presents as a single cyst and at a younger age¹²⁵. In our young patient, there were multiple intradural and extra-medullary spinal cysts at two different levels. In addition, he had a history of prior cardiac involvement.

In a systematic analysis of 467 cases of spinal hydatidosis, Neumayr et al. reported that 78 cases (16.7%) had a history of surgical intervention for extraspinal hydatid cysts. They proposed that it is difficult to say whether spinal involvement in patients with a history of extraspinal echinococcosis results from simultaneous primary infection, or secondary hematogenous seeding or even a new exogenous contamination⁵.

Hydatid cysts are mostly misdiagnosed or missed in the early stages, since they insidiously grow for years before making any symptoms⁹¹³. The condition manifests itself as significant cord compression and/or bone damage¹⁴¹. Symptoms and signs such as radiculopathy, myelopathy, paresthesia, paraparesis, paraplegia, sphincter malfunction and deformity are all reported but none of them is pathognomonic of spinal hydatidosis¹⁵⁹. Spinal hydatidosis is associated with a high degree of morbidity and mortality¹⁰, and its prognosis is being compared to that of malignancies (*‘le cancer blanc’*)⁵. Interestingly, our patient presented with symptoms related to one extruded disc, but his spinal hydatid cysts were asymptomatic.

There are various conditions that resemble spinal echinococcosis, which causes challenges in preoperative diagnosis. These differential diagnoses include spinal tuberculosis (Pott’s disease), brucellosis, osteomyelitis, mycosis, arachnoid cyst, fibrous dysplasia, simple solitary or aneurysmal bone cysts, spinal abscess, malignancy, and vertebral metastases¹⁶¹⁷.

The sensitivity of serological tests to diagnose extra-hepatic echinococcosis is low³. X-rays and CT scans are nonspecific. In some cases, the intervertebral disc is intact, but there are cystic lesions and irregular destruction of the vertebral bodies¹⁷⁹. CT scans and ultrasonography are helpful tools for finding other lesions in the lungs, liver, and other organs. The evaluation of other organs in our patient revealed no involvement.

When it comes to spinal hydatidosis, MRI is the best diagnostic tool for determining the location of the cystic lesions, the spinal levels affected, and their relationship with surrounding organs¹⁶⁹¹⁸. MRI images show sausage-shaped lesions with thin walls and dome-shaped ends without septations or debris in the lumen. A spherical lesion may occur occasionally. CSF-like signal characteristics are found in hydatid cyst contents¹. On T1-weighted images, the parent cyst appears iso-intense or slightly less intense than its fillings. The T2-weighted images reveal homogeneous hyper-intense contents surrounded by a low-intense rim¹⁹¹⁹. The low-intense rim on MRI images results from reactive fibrosis and degeneration around the parasitic membrane and is in accordance with the histopathological results. The existence of a considerably hypo-intense cyst wall on T1 and T2-weighted MRI image sequences is characteristic of spinal hydatidosis. The T2-weighted images also may reveal the viability of the cysts where decreased high signal and increased low signal from collapsed cyst walls indicate a succumbed cyst¹⁹. MRI is also helpful in evaluating the effectiveness of medical therapy and early diagnosis of postoperative recurrence¹. Histopathological reports of the excised cyst may still confirm the diagnosis

For spinal hydatidosis, the primary treatment consists of surgical excision followed by anthelmintic therapy to achieve neural decompression and establish the diagnosis⁹. As patients usually present at advanced stages, treatment is difficult, and recurrence is common in most forms. The location of the cysts, the extent of bone involvement and the presence of spinal instability determine the type of operative procedure, the extent of resection, and the decision whether to perform spinal stabilization or not³ (Pamirl. 2002). Considerable care should be taken during the surgery to avert rupture of the cysts and spillage of their content, which can cause anaphylactic reactions and/or subsequent recurrence⁹. If cysts rupture during excision, the surgical field should be irrigated with hypertonic saline; but unfortunately recurrence is inevitable¹.

The preferred anthelmintic drug against spinal hydatidosis is albendazole, but its efficacy and appropriate duration of treatment are still controversial²⁰. The world health organization (WHO) recommends albendazole for visceral hydatid disease, with the dosage of 10-15mg/kg/day⁴. Recurrence is related to the location of the cysts and is quite uncommon in the intradural extramedullary form of spinal hydatidosis, where there is no intraoperative cyst rupture²¹.

MRI is the modality of choice in the follow-up of spinal hydatid disease as it enables early detection of recurrences¹⁶.

Conclusion

Although spinal hydatid disease is a rare condition, its burden is remarkably high. Vertebral hydatidosis should always be considered as a differential diagnosis for typical and atypical spinal presentations, particularly in endemic areas for echinococcosis.

Preoperative neurologic evaluation combined with MRI helps in localizing the lesion and planning a suitable surgical approach. The treatment consists of surgery and adjuvant anthelmintic therapy. The cysts should be excised carefully to avoid rupture and subsequent recurrence. Irrigation of the surgical field with hypertonic saline is also helpful.

Strict follow-up and regular MRIs are necessary to detect recurrence at early stages. Despite medical advances, many aspects of spinal hydatidosis are still vaguely understood. Therefore, further studies in this field are warranted.

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