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Case Report Open Access

A Rare Case of Primary Spinal Cystic Echinococcosis

Vacaras V, Maior T, Rahovan I, Marias D, Muresanu DF

Department of Neuroscience, University of Medicine and Pharmacy, Cluj-Napoca, Romania

Abstract

Background: Hydatidosis is an infection disease caused mostly by the tapeworm *Echinococcus granulosus* and continues to be a significant health problem in some regions. Spinal involvement is uncommon and found in less than 1% of all cases. Spinal hydatid disease is an important cause of spinal cord compression, especially in endemic countries. Common presenting symptoms are chronic back pain, radiculopathy and lower limb weakness. Early diagnosis, surgical decompression with total removal of the hydatid lesion, when possible, and drug therapy with benzimidazole derivatives is considered to be the most effective therapy.

Case presentation: The purpose of this article is to report a rare case of primary spinal hydatidosis, with more than 50 surgically excised cysts, who admitted to our clinic because of lumbar pain of increasing severity and progressive difficulty with walking, which had lasted for one year.

Conclusion: Despite a correct management, spinal echinococcosis is associated with a high degree of morbidity and mortality. Long-term follow-up is critical, with regular clinical, radiologic and serologic examinations.

Keywords: *Echinococcus granulosus;* Spinal hydatid cyst

Introduction

The first description of spinal hydatidosis was made by Churrier in 1807 [1]. Hydatid disease remains a serious public health issue in several countries and regions, even if worldwide incidence and prevalence have fallen over the past several years. The greatest prevalence of cystic echinococcosis in human and animal hosts is found in countries of the temperate zones [2]. Being a cestode, *Echinococcus granulosus* has both intermediate and definitive hosts. Sheep are the most common intermediate hosts, while dogs are usually the definitive hosts.

Humans function as accidental hosts, because they are usually a dead-end for the parasitic infection cycle [3]. Hydatid disease is generally asymptomatic for a long period, often being discovered incidentally on a routine X-ray or ultrasound [4]. Few cases of primary spinal echinococcosis have been reported in the literature. Hydatidosis of the bone occurs in 0.5-3% of all the cases and the vertebral column is involved in 50% of these [5]. Ultrasonography, computed tomography scanning, magnetic resonance imaging and serological assays are necessary tools for achieving a correct diagnosis. When the cysts do not show characteristic features, the differential diagnosis can be difficult [6]. Spinal cystic echinococcosis (spinal CE- involvement of the spinal cord, the spine, or both structures) is associated with a high degree of morbidity, disability, and mortality and the prognosis has often been compared to that of malignancies, with a case fatality rate reported to exceed 50% within 5 years after onset of symptoms [7]. We present a case of primary hydatid disease of the thoracic spine with no other organ involvement at the moment of the diagnosis. Case management consisted of surgical and medical treatment with anthelminthic agent.

Case Report

A 43-year-old woman, with right laterality and no history of any chronic medical illnesses, presented to our department, with complaints of persistent middle back pain for a year, unresponsive to usual conservative treatment, associated with progressive paraparesis. There was no history of fever, trauma, altered sensorium, or loss of consciousness. She resides in a rural area with history of contact with sheep in the early childhood and permanent contact with dogs. On neurological examination, the patient presented without any changes in the sphere of the cranial nerves, she had increased tone in both lower limbs and moderate paraparesis (motor deficit grade 2/5), increased knee and ankle reflexes bilateral, Babinski sign present on the right side, sensory loss from D8. Complete blood work was done. Notable laboratory values included mild inflammatory syndrome (erythrocyte sedimentation rate=27 mm/h and the C- reactive protein=0.87 mg/dL), iron deficiency anemia (serum iron=38 μg/dL), low levels of folic acid (folic acid=2.93 ng/ml). Antibodies against echinococcosis antigen were found positive, using enzyme immunoassay (EIA) method. The additionally performed blood tests for different rheumatological/autoimmune diseases (rheumatoid factor, antinuclear antibodies, anti-neutrophil cytoplasmic antibodies, anti-cyclic citrullinated peptide antibodies, anti- cardiolipin antibodies, and anti-double stranded DNA antibodies), as well as Borrelia burgdorferi and HIV screening serologies were all negative. An electromyography (EMG) investigation revealed a bilateral radiculopathy and myelopathy syndrome with involvement of the D8 to D10 roots. Thoracic computerized tomography (CT) showed an expansive dorsal process from D1 to D10 with invasive aspect to spinal canal and bone erosions. Magnetic resonance imaging (MRI) of the vertebral column revealed multiple cystic lesions in the paravertebral thoracic region as well as prevertebral space, compressing the spinal cord in the D7-D8 region (Panel B- arrow), invading the vertebral body at this level (Figure 1). Furthermore, an abdominal CT scan completed the investigations, to exclude other sites of the infection, but the examination did not reveal any abdominal organ involvement. The patient underwent surgery to excise the cysts and had a laminectomy and foraminotomy performed through the posterior approach for neurologic decompression at the level of spinal involvement (D7-D8), then

***Corresponding author:** Vitalie V, Department of Neuroscience, University of Medicine and Pharmacy, Cluj-Napoca, Romania, Tel: +40 264 598 523; E-mail: vvacaras@umfcluj.ro

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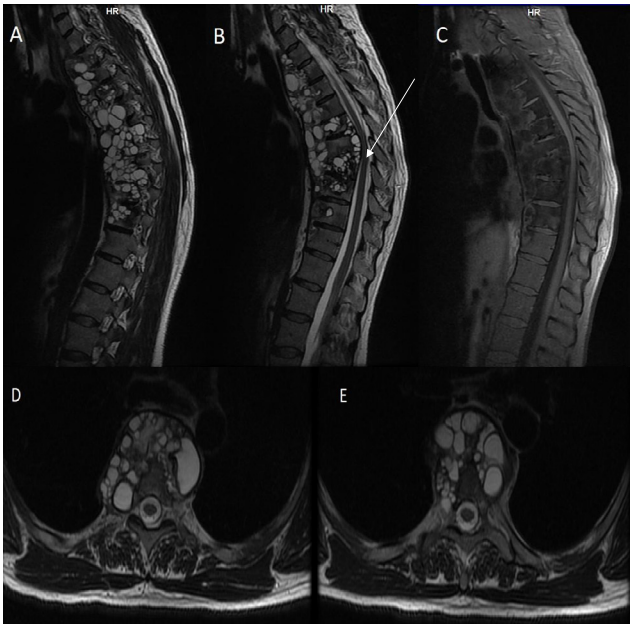

Figure 1: Pre-operative sagittal (A, B, C) and axial (D, E) cross-section MRIs, T2 (A, B, D, E) and T1+ FS (C) weighted images showing multiple hydatid cysts, some of them compressing the spinal cord (B-white arrow).

D7 vertebrolaplasty with acrylic cement. Over 50 cysts were excised, and the surgical region was washed off using 20% hypertonic saline solution. No perioperative antiparasitic therapy was used. Post-operative thoracic CT-scan was performed with no evidence of pulmonary cystic lesions. The patient's postoperative evolution was unremarkable. The histopathological examination of the excised material confirmed the diagnosis of cystic echinococcosis. Pharmacotherapy with Albendazole 400 mg twice a day was started after surgery. Complete blood count as well as liver enzymes were tested before starting the treatment and monthly thereafter. The patient was followed up 3 months postoperatively with clinical examination, liver function tests (serum aspartate aminotransferase and alanine aminotransferase), kidney function, complete blood count, hydatid serological examination and MRI. The paraperesis almost fully recovered, and blood tests were in normal ranges. The antibodies for *Echinococcus granulosus* and multilocularis IgG were positives. MRI of spine (Figure 2) revealed multiple microvesicular lesions from D6 to D10 with infiltration of the bone (osteolytic lesions) at these levels (Panel F, white arrows). Multiple other cysts were observed in the pre- and paravertebral space (extra pulmonary), from D1 to D11, with inhomogeneous content. In addition, a solitary cystic lesion (69 mm/29 mm/44 mm) was situated in the posterior side of the left costodiaphragmatic recess.

Discussion

Hydatid disease is characterized by cystic lesions that are mainly found in the liver (75%) followed by the lungs (15%), brain (2-4%), and genitourinary tract (2-3%) [8]. Spinal involvement is uncommon and found in less than 1% of all cases [9]. Spinal hydatid cysts are located most frequently at the thoracic level (52%), followed by the lumbar (37%) and then the sacral and cervical levels [8,10]. In our study, we presented a rare case of primary cystic echinococcosis of the spine. In order to support this diagnosis, screening investigations (pulmonary and abdominal CT-scan) for extraspinal CE were made before and 24 hours after surgery. We excluded any other organ involvement.

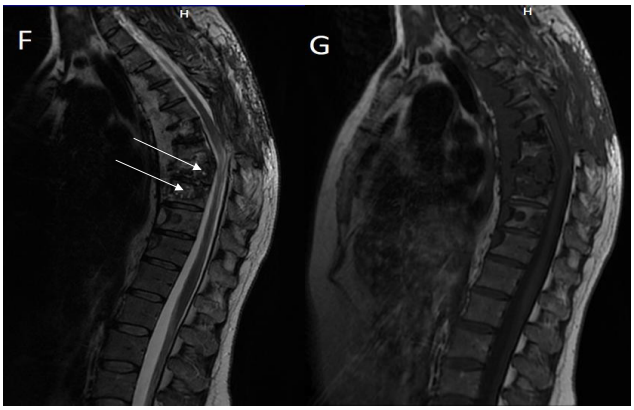

Figure 2: Post-surgery sagittal cross-section MRI images, T2 weighted (F) and T1 weighted (G).

Cystic echinococcosis is still endemic in several countries especially in temperate zones (the Mediterranean regions, southern and central parts of Russia, central Asia and China), South America, Australia, north and east Africa [2]. An epidemiological survey of cystic echinococcosis in cattle and sheep (8569 animals) conducted for two years in endemic areas of northeastern and southern Romania showed the hyperendemic presence of *Echinococcus granulosus* in this regions. The hydatid cyst was present in 49.87% of the sheep and in 32.34% of the cattle analyzed, showing the necessity of sustainable surveillance and control strategies both in animals and humans [11]. It is estimated that about 2-3 million people around the world suffer from echinococcosis and the incidence of hospitalized cases ranges between 0-32/100.000. Its frequency increases with age and is more common in women [12].

Braithwaite and Lees classified the spinal hydatid disease in five types:

A) Intramedullary hydatid cyst;

B) Intradural extramedullary hydatid cyst;

C) Extradural intraspinal hydatid cyst;

D) Spinal hydatid cyst;

E) Paravertebral hydatid cyst [13].

Hydatid disease continues to be a significant health problem, often causing spinal cord compression syndrome [2,14]. There are no pathognomonic signs or symptoms for spinal echinococcosis, but it might be the cause of continuous back pain, weakness, and numbness in both lower limbs. The differential diagnosis is important, especially for those living in endemic areas, as it may easily be misdiagnosed [15]. This disease can be fatal, due to the anaphylactic response caused by cyst rupture. In our case, spinal cord was under significant pressure, causing severe neurological deficit. Antibody detection by serology is useful, although sensitivity is only maximum 85% (higher for hepatic cases) and a negative test does not exclude the diagnostic of echinococcosis [16]. CT scan is playing a complementary role to MRI because efficiently demonstrates erosions of the bone. Osteolytic bone states like metastasis or tuberculosis are important for the differential diagnosis [17]. Magnetic resonance imaging (MRI) is the preferred imaging modality in the diagnosis of hydatid cysts. MRI is essential for evaluating the extension of the disease because of its superior soft tissue resolution. The use of diffusion-weighted MRI proved to help for the differential diagnosis. The cysts are hypointense on T1W images and hyperintense on T2W images. The utility of myelography is limited nowadays [18].

In our case, MRI showed the invasive character of the lesion and this was crucial in deciding the most suitable surgical approach. The only medical treatment option for spinal CE is antiparasitic therapy with benzimidazole compounds. Albendazol (ABZ) is the drug of choice, for a minimum of 3-6 months, 400 mg twice daily (10-15 mg/kg/day) [19]. The reported efficacy is the radiological decrease of lesions in 30%-50% of cases [16]. Adverse effects of ABZ include severe leucopenia, thrombocytopenia, hepatotoxicity, alopecia. During the treatment, monitoring the liver function is mandatory, once a month [20]. Chemotherapy is contraindicated for large cysts that are at risk to rupture and for inactive or calcified cysts [21]. Treatment response depends on cyst stage, size, the presence of daughter cysts and bone involvement.

Surgical early decompression is the treatment of choice for spinal hydatid disease with posterior or anterior approach by laminectomy and stabilization of the compromised column. The size and the location of the lesions determine the surgical approach. Stability of the anterior column must be ascertained. The aim of surgery is total resection of the cysts, but often this is not possible because of the surrounding structures near the lesion [22]. During the intervention, rupture of cysts may provoke various hypersensitivity reactions like urticaria, rash, edema, bronchospasm and more severe, anaphylactic shock that can be fatal [23]. In order to prevent these adverse effects, perioperative treatment with albendazol should be initiated. The role of percutaneous treatment and radiotherapy is still uncertain [22]. In our case, radical surgery was not possible because of the high degree of the bone erosion. In the large literature review regarding spinal hydatid disease, by Neumayr et al. it appears that primary spinal CE is more frequent than secondary spinal CE. In CE arising from vertebral bone, growth of the parasite is generally slow and produces, in time, aggressive bone infiltration. In CE arising from the spinal or paraspinal soft tissues, the growth pattern is primarily eccentrically spherical. Recurrence rates are higher in cases with vertebral bone involvement. Local recurrence of disease has been reported to occur up to 29 years after surgery for spinal CE. Previous surgery for extraspinal CE appears to be more frequently associated with thoracic vertebral involvement [24].

Conclusion

Primary spinal echinococcosis is a rare condition, with significant morbidity. Surgery combined with anthelminthic therapy can stop the progression of the disease and prevent recurrence. The compliance of the patient is very important in the management of the case. The relapse rate is high, so patients should be checked regularly. In this case report, the combination of surgical resection and postoperative chemotherapy achieved a very good recovery of the neurological deficit, but a secondary cystic lesion was discovered in the left costodiaphragmatic recess, three months after surgery, despite the continuous administration of 800 mg albendazole/day. Because of the high recurrence rate cited in literature, we consider that additional research is required for new therapeutic approach of hydatid cysts.

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