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Letter to the Editor

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Secondary lumbosacral echinococcosis as presumptive sequelae of other primary locations

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Sir,

A 62-year-old patient attended the emergency department due to increased pain in the lumbosacral spine in the last 2 months. At the examination, she presented worsening of her previous motor and sensory deficits in the lower extremities. The patient presented with a medical history of interest was hydatidosis with pulmonary cysts that required surgery at the age of 12 years and later liver cysts operated on at the age of 30 years. About 20 years ago she was diagnosed with lumbosacral hydatidosis with intolerance to albendazole due to hepatic toxicity, requiring a total of 13 surgical interventions (including arthrodesis of L1-L2-L3 vertebrae). Despite all these interventions, she presented poor clinical evolution with chronic low back pain and incomplete paraplegia.

A computed tomography (CT) scan of the lumbosacral region was performed, which showed a known multicystic lesion from L4 to S1 compressing the sacral canal and conjunctival foramina. A magnetic resonance imaging of the lumbosacral spine was also conducted (Figure 1A and B), showing signs of osteolysis on L5 vertebrae with anterior displacement of the intersomatic devices, severe anterolisthesis and vertebral hydatidosis at L4-L5-S1. Clinical and radiological findings were consistent with progression of known lumbosacral hydatidosis, so the patient was admitted for surgery and intravenous (IV) treatment. Revision surgery of lumbosacral arthrodesis (L1-L3 vertebrae) and iliac screws was conducted. Unilocular cysts (Figure 1C) consistent with hydatid cysts were removed in the lumbar spine together with previous osteosynthesis material and lumbar arthrodesis. Samples were taken from the cysts (Figure 1D) for microbiology and pathological anatomy.

Protoscoleces and hooklets consistent with *E. granulosus*-complex were observed on saline wet mount (Figure 2C and D) in microbiology laboratory. Hyalinized and calcified fibrous wall with structures compatible with hydatidosis were also observed using hematoxylin and eosin stain (Figure 2A and B). She received 1200 mg/12 h of oral praziquantel with no associated albendazole (due to previous episodes of hepatotoxicity). The patient improved her condition, being discharged with 1200 mg/12 h of oral praziquantel for three months. The patient improved improved significantly after three months, returning to baseline symptoms and improvement of the previous motor and sensory deficits. A lumbosacral CT was conducted six months after the admission with significant decrease of the collection size in subcutaneous cellular tissue in this area.

Cystic echinococcosis (CE) or hydatid disease is a parasitic zoonosis caused in humans by the larval stage of *Echinococcus granulosus sensu lato,* which includes five species: *E. granulosus* sensu stricto, *Echinococcus equinus, Echinococcus ortleppi* or *Echinococcus canadensis,* belonging to the cestode class. This disease is considered one of the 20 neglected tropical diseases by the World Health Organization [1]. CE has a global distribution, with higher prevalence in Mediterranean countries, North and East Africa or China, among others. Casulli *et al.* reported the incidence of cases in many European countries in a recent article. In Spain, the incidence in the period 1997–2020 was one case per 100,000 inhabitants, while in the period 2017–2019 it was 0.56 cases per 100,000 inhabitants, with a total of 10,675 reported cases and a decreasing trend since 1997 [1]. In Spain, the number of cases of CE seem to be underdiagnosed for two reasons: (a) only symptomatic patients are studied; and (b) reporting of cases to the epidemiological surveillance systems remain suboptimal. The review by Zabala *et al.* highlights this deficiency of the current European and Spanish reporting systems due to the disparity of cases reported in the literature compared to those declared [2]. CE is transmitted within the domestic domain, with dogs, foxes or wolves participating as definitive hosts, small ruminants as intermediate hosts and humans as accidental hosts [3]. Transmission to humans can occur by hand-

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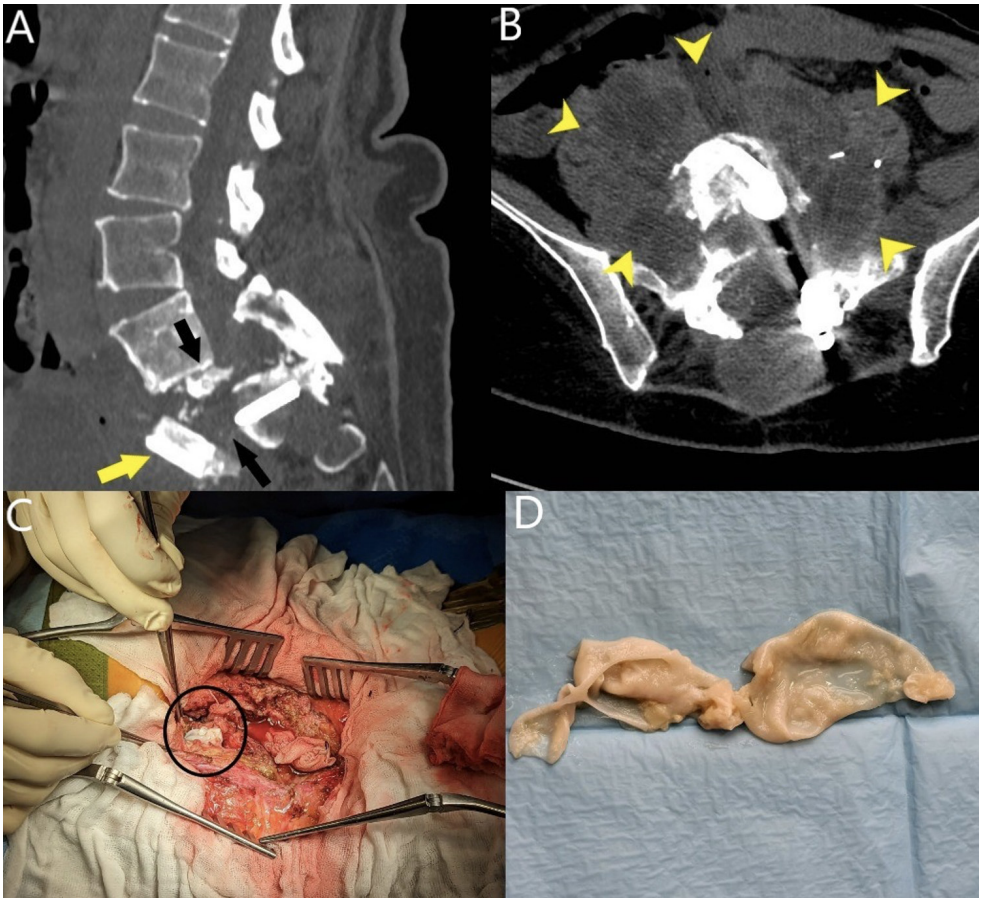

Figure 1 **A: Osteolysis of the L5 vertebral body (black arrows), spondylolisthesis and displacement of the intervertebral device (yellow arrow). B: Multiple perivertebral hydatid cysts (yellow arrow head). C: Removal of hydatid cysts by instillation of hypertonic saline. D: Macroscopic aspect of hydatid cyst removed during spinal surgery.**

to-mouth transfer or contamination with embryonated eggs of *E. granulosus sensu lato*.

Hydatid disease is usually asymptomatic until a complication occurs such as anaphylactic reaction after cyst rupture or mass effect on surrounding structures, and it is frequently an incidental finding. Most patients present with a single cystic lesion located in a single organ, most typically the liver (in up to 70% of cases) followed by the lungs as the second most common organ involved [4]. Rarely, *E. granulosus* can affect other organs such as abdominal or pleural cavities, kidney, brain or bones, among others [5, 6]. It can occur in primary form, exclusively in an extra-hepatic organ, concomitant with hepatic and pulmonary hydatidosis, or by peritoneal seeding secondary to rupture of an intra-abdominal hepatic hydatid cyst. Since the patient in this case had a history of previous hepatic and pulmonary hydatidosis, it is most likely that secondary seeding was present during some of the cystectomies. Spinal hydatidosis is rare, accounting for 0.5%-1% of total cases, however, these account for up to 45% of total cases of CE affecting bone sites [7], with lower dorsal and lumbar regions as the most frequently affected sites, while involvement of cervical and sacral vertebrae is anecdotal [8]. The usual presentation includes symptoms of spinal cord compression as radiculopathy or myelopathy [9], and/or local pain due to destructive bone lesions or pathological fracture [10]. Alveolar echinococcosis (AE) caused by *Echinococcus multilocularis* should be considered in the presence of cysts due to their similarities with CE. AE occurs mainly in northern hemisphere, including central and northern Europe, Central Asia, northern Russia, northern Japan, north America, among other countries. AE is very rare in Spain, in fact, only two cases have been reported in the literature [11]. *E. multilocularis* forms multivesiculated cystic mass and shows peripheral infiltration with central necrosis, which resembles invasive malignant tumors. Whereas *E. granulosus* grows slowly to form unilocular cyst that expand and crowd the affected organs and tissues [12], being consistent with this case.

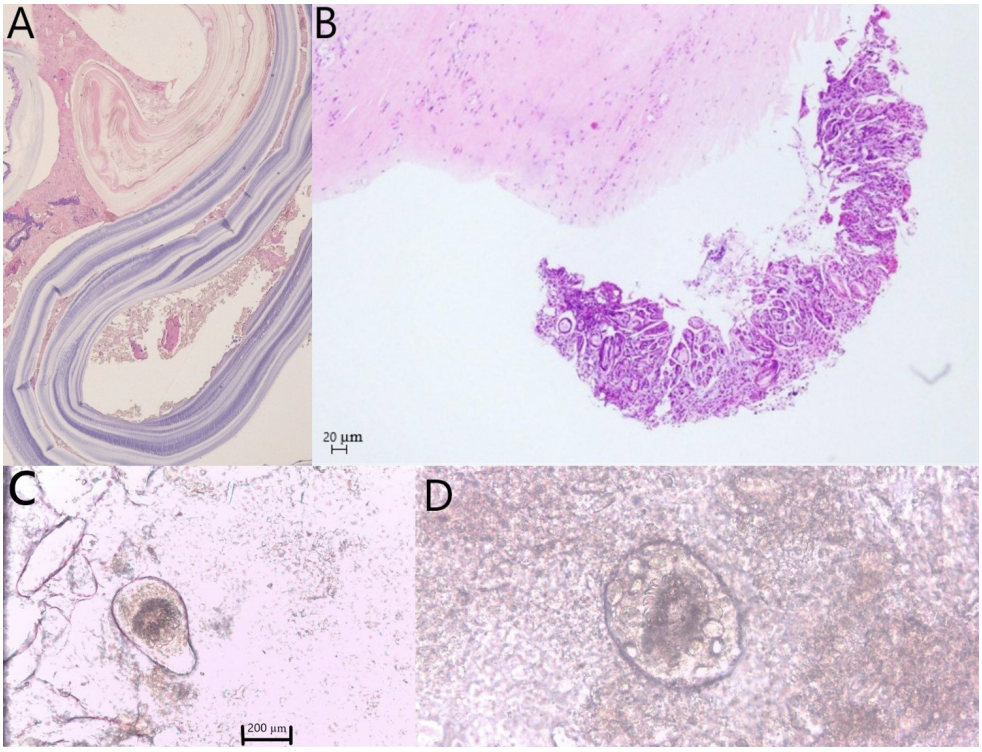

Figure 2 **A: hematoxylin and eosin stain (H&E, x 400). Example of the layers of the hydatid cyst, with intermingled fibrinohaematic tissue. On the right, the middle lamellar layer is identified and, loosely, disintegrated strips of the inner layer. B: 4x H&E stain. Histological section showing fragment nucleate germinal layer from which the brood capsule arises with proliferative vesicles artifacted by diathermy. C and D: multiple protoscoleces and hooklets consistent with the *E. granulosus* cestode were observed in saline wet amount from the clinical specimens.**

CE diagnosis is fundamentally based on epidemiological history, clinical findings, serological tests and imaging studies. In fact, this disease has few specific imaging features: a non-enhancing cystic lesion with no proportionate oedema, the water lily sign and presence of multiple daughter cysts [13]. CT scan and MRI are techniques of choice for extra-abdominal cysts and as a form of pre-surgical assessment. ELISA, indirect haemagglutination and latex agglutination can be used for detection of serum antibodies [14]. The result have to be confirmed by a high specificity serological test such as Western Blot if the result is positive or if false negative is suspected. In this case, the patient presented medical history of hepatic and pulmonary hidatidosis, therefore the serological tests were not performed because it did not contribute anything to the diagnosis. Confirmation of cases is given by identification of parasite structures in the microscopic examination of the hydatid cyst fluid or in the histological samples, as well as by identification of its genome in invasive samples [15].

Treatment of CE is based on three important points: use of antiparasitics, surgery or percutaneous drainage (PAIR), although the latter is contraindicated in cases of pulmonary cysts. In spinal hydatidosis, surgery is the treatment of choice with decompression of a compromised spinal cord and stabilization of a compromised spinal column [16]. Although it often does not prevent disease progression, it has been shown to prolong survival in some cases. Intraoperative use of benzimidazoles derivatives in the pre-, peri-, and postoperative is currently considered the treatment standard of spinal CE [7]. Monotherapy with albendazole have showed good results in patients with inoperable disease, in fact, el-Mufti M *et al.* reported a cure rate of 53% at a minimal follow-up time of two years with albendazole [17]. In addition, treatment with PAIR or radiotherapy could be useful in patients with extensive diseases, but up to now, its role remains undefined [7, 16]. Recurrence rates of spinal CE are high, especially in cases with vertebral bone involvement. Follow-up of the cases should be carried out every six months during the first two years with imaging and serological tests. Despite significant advances, spinal echinococcosis remains associated with a high degree of morbidity, disability and mortality.

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None to declare

CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

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