Hydatid disease of the vertebrae with intraspinal involvement. A medical and surgical disease. A two-case report

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ABSTRACT

Hydatidosis is a disease caused by the larval stage of *Echinococcus*, the most prevalent species being *Echinococcus granulosus*. Hydatidosis is endemic to Eurasia, Africa, Australia and South America. In Chile, it has a reported incidence of 2.1 cases per 100,000 inhabitants and, although it is associated with the livestock areas of southern Chile, there have been reports throughout the territory due to the displacement of the population and its long incubation period. It is characterized by the presence of a cyst usually at the hepatic (75%) or pulmonary (15%) level. The involvement of other regions is less common and bone involvement is a rarity that does not exceed a 1-2.5% prevalence, either in the trabecular tissue or spinal canal. A combined medical and surgical approach is the most common treatment which involves disparate clinical outcomes due to the high rate of recurrences and sequelae. Experience in the management of patients with intraspinal hydatidosis is limited because of their low frequency. Therefore, we wanted to report 2 cases treated in our center and to analyze the progress in its management.

Key words: Hydatidosis; intraspinal; spine; *Echinococcus*.
Level of Evidence: IV

Hidatidosis vertebral intrarraquídea. Una patología médico-quirúrgica. Reporte de dos casos

RESUMEN

La hidatidosis es una enfermedad causada por el estado larval del platelminto *Echinococcus*, cuya especie más prevalente es *Echinococcus granulosus*. Se trata de un cuadro endémico en Eurasia, África, Australia y Sudamérica. En Chile, tiene una incidencia notificada de 2,1 casos por 100.000 habitantes y, pese a estar asociada a las zonas ganaderas del sur del país, se detecta en todo el territorio debido al desplazamiento de la población y a su largo período de incubación. Se caracteriza por la presencia de un quiste habitualmente en el hígado (75%) o los pulmones (15%). La afectación de otros sitios es menos frecuente y el compromiso óseo es raro, no supera el 1-2,5%, ya sea en el tejido trabecular o el canal medular. El tratamiento, en general, es médico-quirúrgico con resultados clínicos dispares debido a la alta tasa de recurrencias y las secuelas. La experiencia en el manejo de pacientes con hidatidosis intrarraquídea es limitada a causa de su baja frecuencia. Por este motivo, quisimos reportar dos casos atendidos en nuestro centro y analizar su manejo.

Palabras clave: Hidatidosis; intrarraquídea; espinal; *Echinococcus*.
Nivel de Evidencia: IV

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INTRODUCTION

Hydatidosis is a disease caused by the larval stage of *Echinococcus*, *Echinococcus granulosus* being the most prevalent species. It is endemic to Eurasia, Africa, Australia and South America. This zoonotic parasitic disease caused by ingestion of the *E. granulosus* ova which are excreted in dog feces; through ingestion of contaminated water or food or contact with infected canines carrying them on their fur. The transmission cycle is commonly associated with ovine and caprine livestock, intermediate hosts that develop visceral cysts, thus allowing the cycle to continue as dogs eat the parasite-infested animal viscera. Humans are accidental intermediary hosts in this cycle.[1,2] In Chile, it has a reported incidence of 2.1 cases per 100,000 inhabitants and, although it is associated with the livestock areas of southern Chile, there have been reports throughout the territory due to the displacement of the population and its long incubation period. It is characterized by the presence of a cyst usually at the hepatic (75%) or pulmonary (15%) level that may produce symptoms due to the pressure exerted on surrounding tissues or their complications. The involvement of other regions is less common and bone involvement is a rarity that does not exceed a 1-2.5% prevalence, either in the trabecular tissue or spinal canal.[3]

In the Región de Los Ríos region, the average incidence is 6.1 cases per 100,000 inhabitants, the mortality rate is 0.1 cases per 100,000 inhabitants, and the case fatality rate is 1.3%.[4]

Experience in the management of patients with intraspinal hydatidosis is limited because of their low frequency. Therefore, we report 2 cases treated in our center and analyze its challenging management.

CLINICAL CASE 1

Eighty-year-old patient with history of high-blood pressure and type 2 diabetes mellitus, and native to the endemic region of southern Chile. He has been diagnosed with liver hydatidosis, and is under a long-standing follow-up following a digestive surgery. In 1990, he underwent surgery due to an abdominal retroperitoneal mass that was removed in block. The operative record shows a cyst at L4 level that was completely removed and multiple cysts related to the iliopsoas muscle. There is no record of antiparasitic therapy. After the surgery, the patient evolved with motor weakness involving the left thigh anterior aspect, and sensory involvement. This symptom was considered to be an intraoperative injury of the left femoral nerve.

In 1995, the patient attended controls due to urinary retention. Control CT showed multiple retroperitoneal cysts. The patient fails to comply with the controls and follow-up until 2002, when he is controlled for his digestive surgery.

He underwent a new surgery in August 2003 due to an abdominal hydatid cyst. The discomfort and neuropathic pain in the left thigh persisted. During surgery, a cyst with a major diameter of at least 20 cm was drained and removed in block. A cyst sample or anatomical pathology testing was taken, which report revealed a hydatid cyst report. After surgery, the patient was treated with albendazole 400mg every 12 hours, for 8 twenty-eight-day cycles with a fifteen-day rest period between cycles.

In 2008, multiple retroperitoneal cysts and a hydatid cyst in the psoas muscle are detected. The neuropathic pain persisted with an incapacitating effect, and the patient had to use a cane to walk. In addition, he developed bowel and bladder incontinence and erectile dysfunction. MRI shows spine involvement at L2/L3, with cystic images in the left paravertebral region suggestive of hydatidosis associated with spinal stenosis due to spinal osteoarthritis at L3/S1, and also intraspinal hydatid involvement, accounting for the neurologic symptoms (Figure 1).

The patient remained hospitalized for evaluation, with a diagnosis of cauda equina syndrome secondary to intraspinal hydatidosis. In October 2008, neurosurgeons from the Neurosurgery Department removed the hydatid cyst. In addition, the patient underwent an L3 laminectomy and a partial facetectomy of L3 medial portion and L4 left facet, with significant root compression. The cyst was punctured and then completely dissected out. Twenty-eight days after surgery, the patient was treated with albendazole 400mg every 12 hours, fifteen-day rest period between cycles, for 6 months.

In 2010, due to lumbar chronic pain, the patient was observed by members of the Orthopaedic Department. Radicular pain at the left superior lumbar region was detected. Regular control imaging showed osteolytic lesions in L2-L4 vertebral bodies (Figure 2).

In April 2010, the patient underwent a new decompressive laminectomy which revealed a large main hydatid cyst with a major diameter of at least 12 cm. Although the cyst was punctured and then completely dissected out, multiple intraspinal cysts were detected. The patient was treated with albendazole 400mg every 12 hours, in twenty-eight-day cycles with a fifteen-day rest period between cycles.

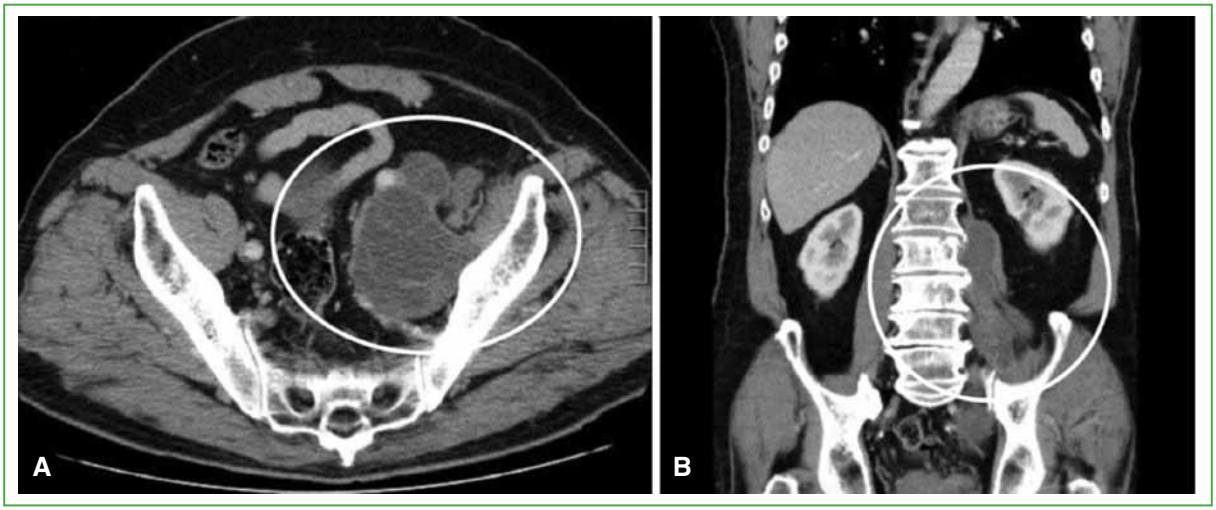

Figure 1. Abdominal and pelvic CT. **A.** Transverse section. Intrapelvic lobulated mass of approximately 14cm long, related to the left iliopsoas muscle. **B.** Frontal section. Lobulated mass related to L1-L2 and left aspect of L3. (taken in 2008).

Serial controls showed L2-L4 vertebral collapse and hydatid cysts in the iliopsoas muscle, findings which led to deciding in favor of a long posterior fixation.

In November 2012, the patient underwent T12-L5 posterior fixation using pedicular screws, and a wide L3-L4 laminectomy (Figure 2).

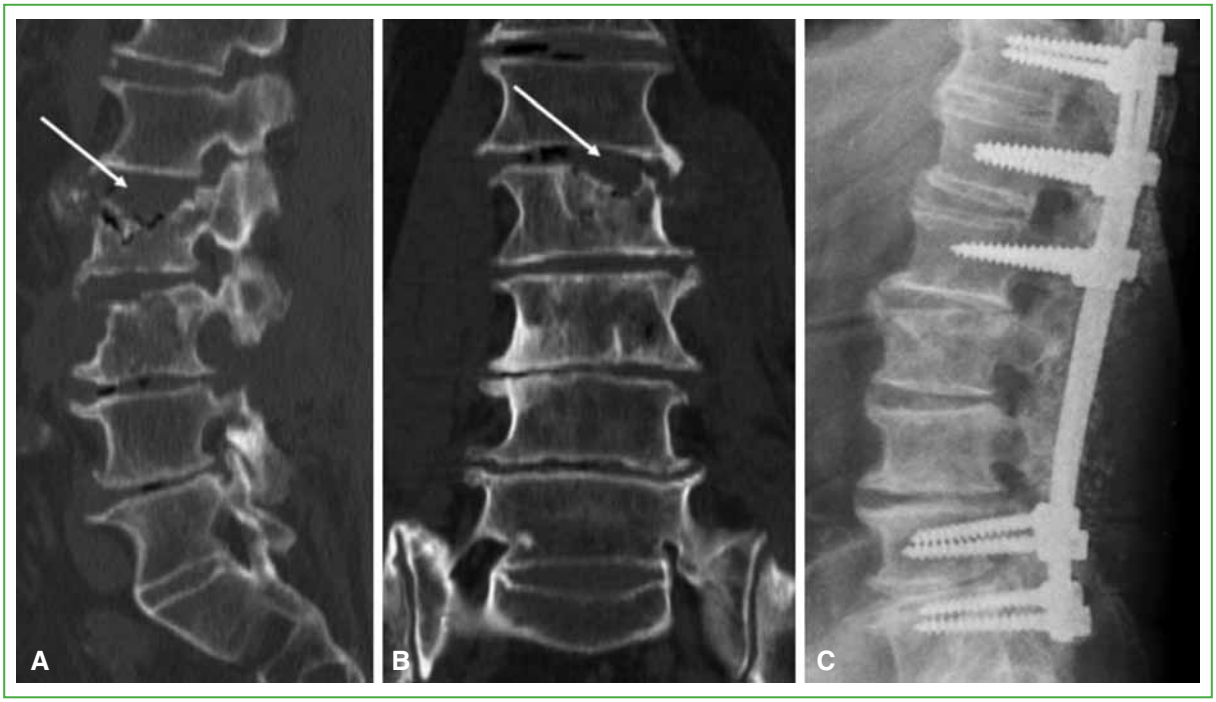

Figure 2. Lumbar spine CT. **A.** Sagittal section. Osteolytic lesions in L3 and L4 upper plates and bodies. **B.** Frontal section. Osteolytic lesions in L3 and L4 upper plates and bodies. **C.** Lumbar spine X-ray. Sagittal section showing the result of the T12-L1-L2-L5 and S1 long lumbar fixation (November 2012).

The pathology report revealed osteochondral tissue, adipose tissue and striated muscle tissue, and no evidence of recurrence. The patient was treated with albendazole 400mg every 12 hours, for 6 twenty-eight-day cycles with a fifteen-day rest period between cycles.

The patient had a favorable course; he still experienced hypesthesia at L1-L3 level and strength reduction (M2-M3) at L4-S1 level. There is no sphincter involvement. The patient currently attends to the orthopedic follow-up, although not on a regular basis, and his treatment adherence is low (Figure 3). In his last control, in late 2016, a new retroperitoneal mass with displacement of abdominal structures was detected. The patient was referred to digestive surgery services to continue the retroperitoneal mass treatment.

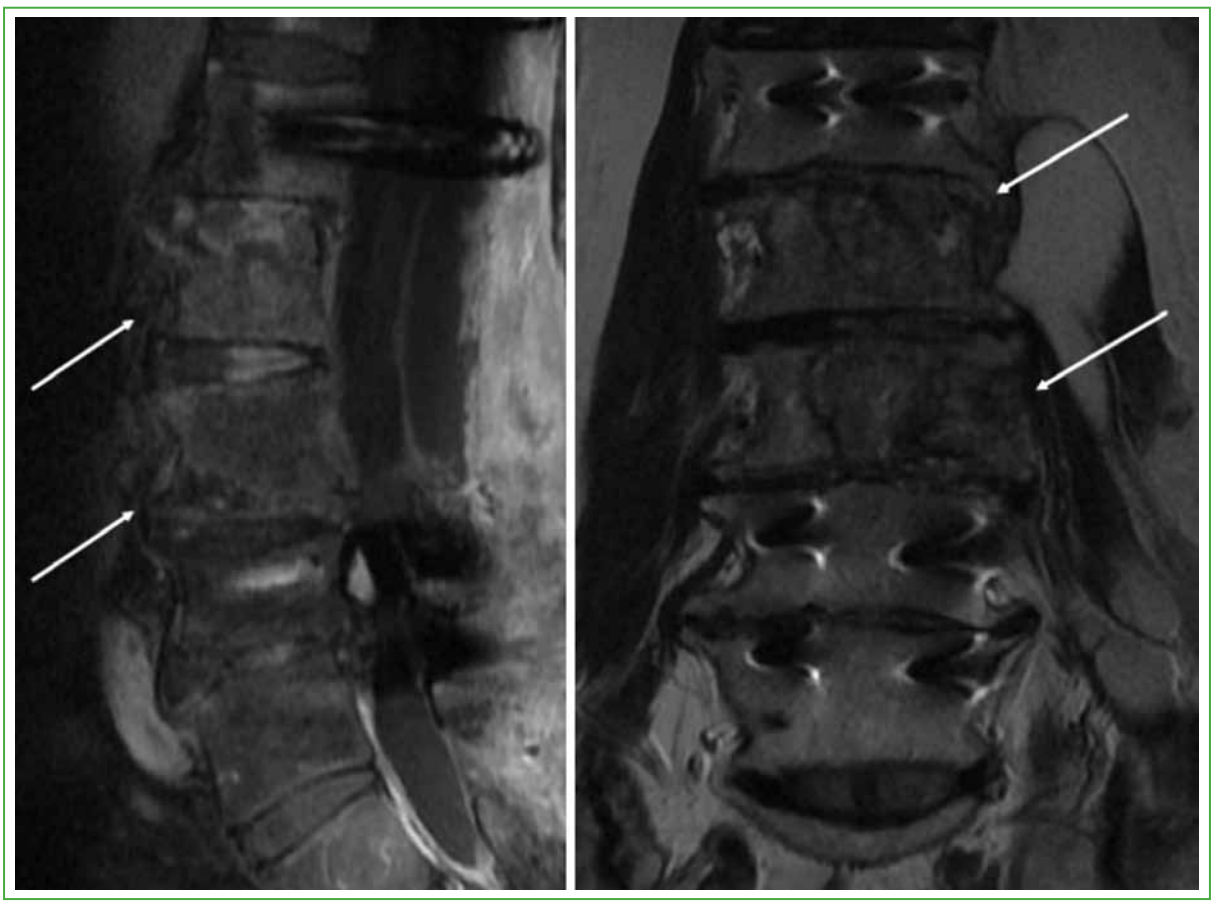

Figure 3. Lumbar spine MRI. Sagittal and frontal sections showing osteolytic involvement of the bodies of L2, L3 and L4 (October 2015).

CLINICAL CASE 2

Fifty-four-year-old patient with no pathological history at disease onset. Patient is native to the endemic region of southern Chile. In March 2008, the patient consulted for a 10-year low-back pain in an Orthopaedic Department. The pain was persistent and progressive, and associated with a palpable lumbar paravertebral mass, with a rubbery consistency, and was approximately 10 x 4 x 3 cm. A CT scan showed a soft tissue expansion process in the right paravertebral posterior region, with erosive involvement on the L4-L5 interapophyseal joint. MRI showed lumbar extradural expansion process and paravertebral extension with segmental spinal stenosis from L4 to S1, of 14 x 6 x 5 cm, with lobulations and well-defined margins. The examination of a puncture sample concluded: sample

consisting of normal muscle fibers accompanied by mucoid material and chronic inflammatory elements, including foreign-body giant cells. The patient was referred to the Hospital Base Valdivia to continue the examination of the lumbar paravertebral mass (Figure 4).

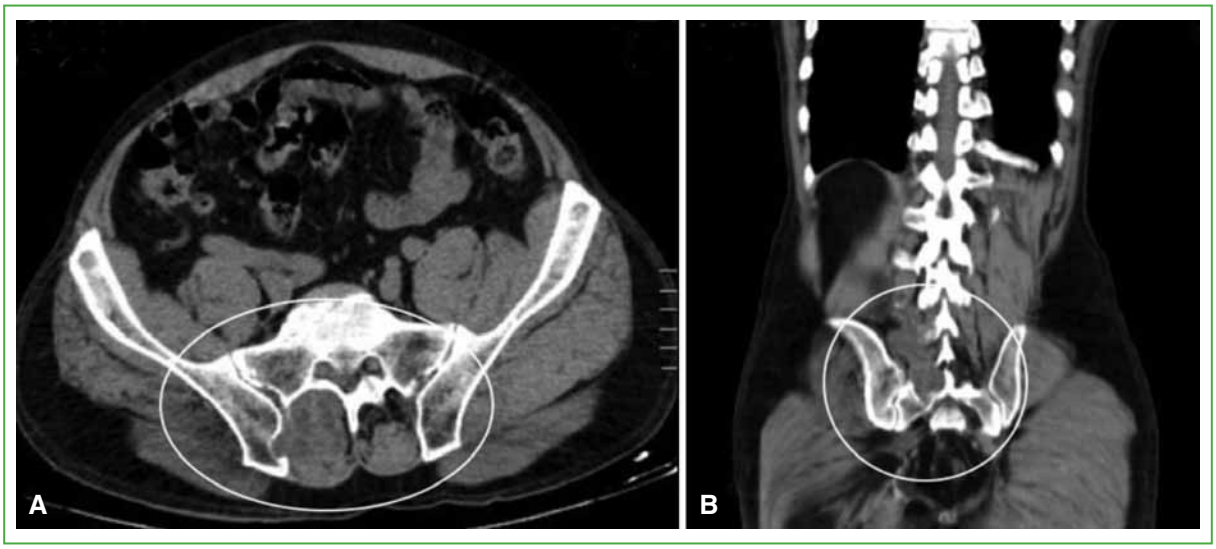

Figure 4. Abdominal and pelvic CT. **A.** Transverse section. Right paravertebral lesion related to the sacroiliac joint. **B.** Frontal section. Right paravertebral mass from L4 to S1 (August 2009).

Physical examination at admission revealed the aforementioned palpable mass. The patient had a positive right straight leg raise test, L5 region paresis (M4) associated with L5 hypesthesia. A new MRI showed images compatible with extraspinal cystic lesions in the paravertebral region, with no intraspinal lesions, compatible with hydatidosis. Thorax, abdomen and pelvis CTs showed no hepatic or pulmonary lesions. The *Echinococcus* IgG ELISA Test was inconclusive (two consecutive analyses were indeterminate).

In January 2010, lesions were removed in block. During sample collection, the capsule of the tumoral lesion ruptured and drained abundant cloudy fluid and friable, cerebellar-like material. Cystic samples for anatomical pathology testing were taken (Figure 5).

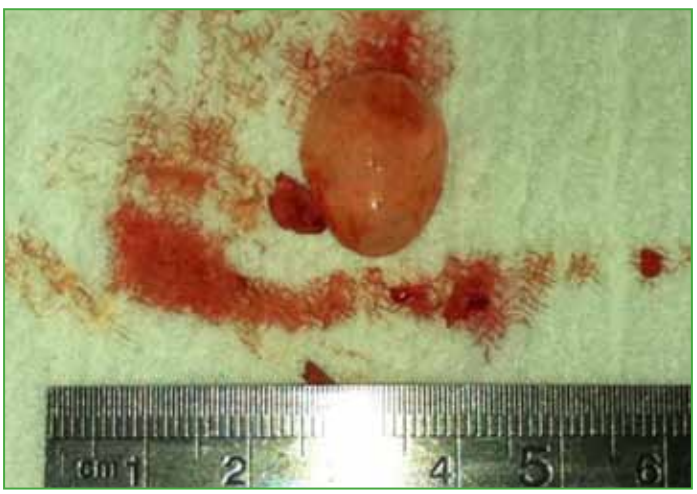

Figure 5. A 2 x 1,5 cm cystic lesion from a hydatid cyst removed in January 2010.

The quick biopsy report reveals hydatidosis. After surgery, the patient was treated with albendazole 400mg every 12 hours, for 2 twenty-eight-day cycles with a fourteen-day rest period between cycles. Postoperative neurological examination showed no changes.

In late 2010, patient developed pain and severe radiculitis at L5 and right S1 with muscle strength loss. Control MRI showed multiple intraspinal cystic lesions at L4-S1 (which were not detected in previous MRI scans) and joint damage at L4-S1.

In April 2011, the patient underwent L4-S1 posterior fixation using pedicular screws, decompression by L4 hemilaminectomy, and an L5 laminectomy. Upon opening the spinal canal, the surgical team found multiple cysts compressing the dural sac and the left roots L5 and S1. Cysts were removed and the area was properly cleaned up. The patient was treated with albendazole 400mg every 12 hours, for 2 twenty-eight-day cycles. After surgery, the patient still experienced radicular pain without any improvement in motor and sensory tests. The surgical biopsy confirms the presence of structures compatible with hydatidosis (Figure 6).

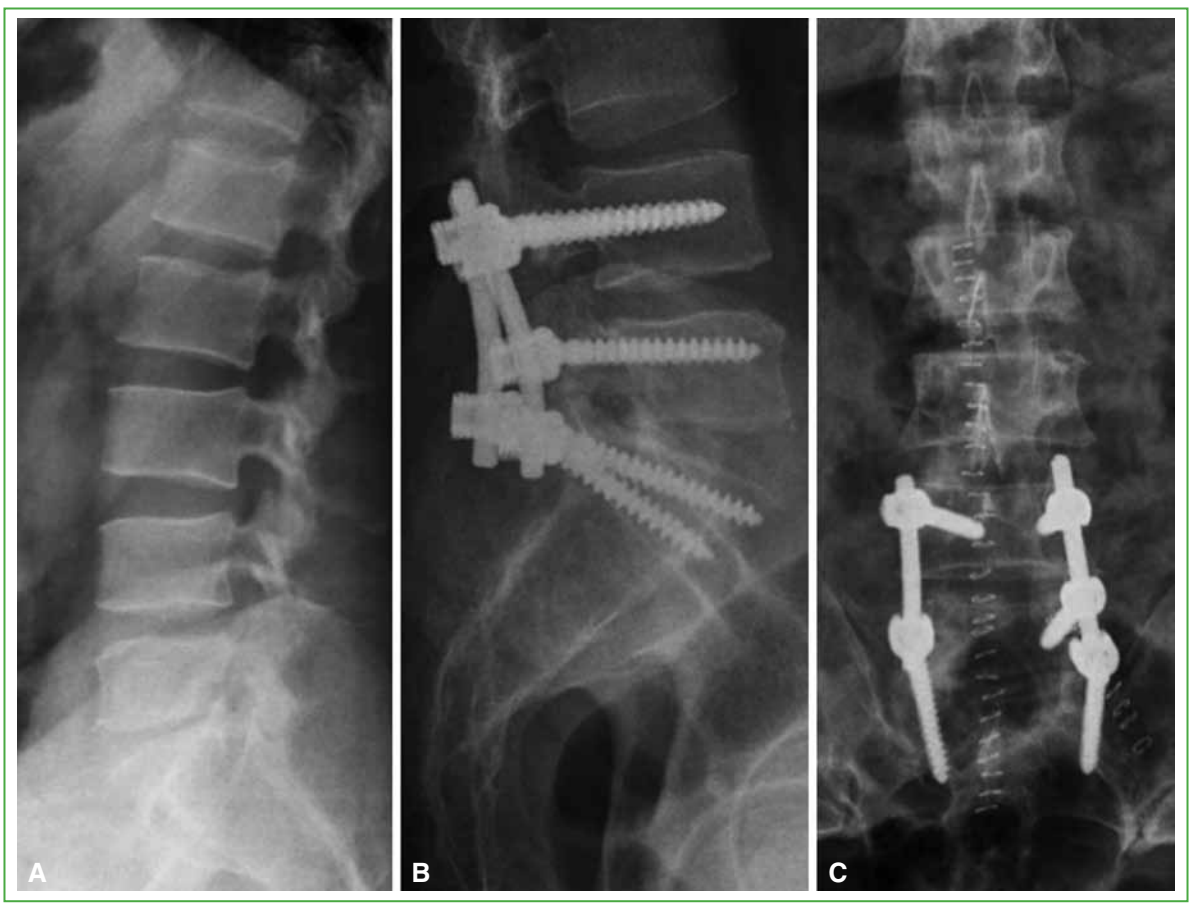

Figure 6. A. Lumbar spine X-ray. Sagittal section showing no bone lesions. **B and C.** Lumbar spine X-ray. Short posterior fixation of L4-L5-S1 (April 2011).

The patient had a favorable course. The 3-week follow-up showed M5 at L3-L5 and M4 at S1. The use of non-steroidal anti-inflammatory drug significantly subsided pain.

In the outpatient controls, the patient remained with muscle strength loss at the right L5-S1 level, which was associated with hypesthesia of the same segments. In 2014, the patient developed a condition with symptoms compatible with cauda equina syndrome, with loss of control of the urethral sphincter. This condition prompted a new lumbar spine MRI that revealed a recurrence of large lesions at the intraspinal lumbar level. A watchful waiting

strategy is employed, and albendazole therapy is re-instituted (400mg every 12 hours, in 2-month cycles with a 2-month rest period and liver testing between cycles); symptoms subdue (Figure 7). The patient currently has no cauda equina involvement, but suffers from residual radiculopathy at the right L5 and S1 level, with good sphincter control. The albendazole therapy continues in 2-month cycles with a 2-month rest period between cycles.

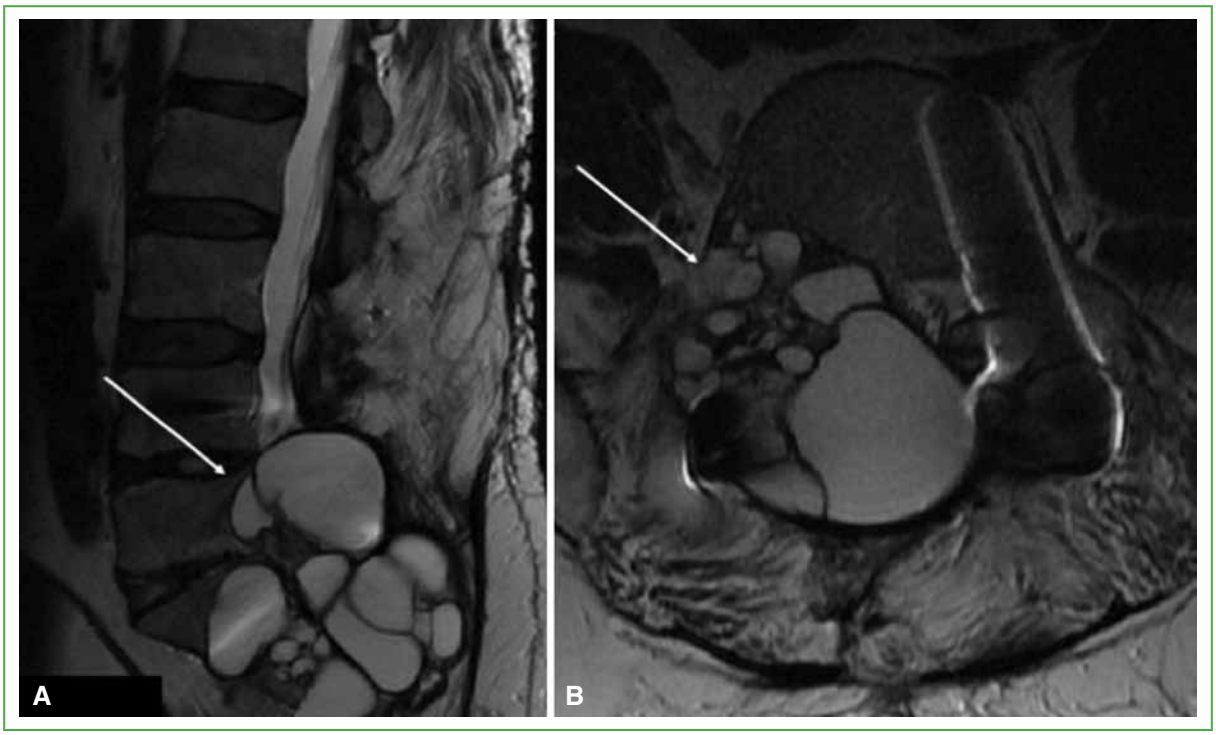

Figure 7. Lumbar spine MRI. **A and B.** Sagittal and transverse sections. Multiple cystic lesions related to L4-L5 and S1, showing intraspinal involvement (May 2014).

DISCUSSION

Bone hydatidosis is rare, less than 2.5% of hydatidosis patients present bone involvement. Bone hydatidosis patients average 51 years at diagnosis and are twice as likely to be women. The most common bone sites involved include the vertebrae (50%) and the hips (30%).[1] Any vertebral segment may be involved: thoracic (50%), lumbar (20%), sacral (20%), and cervical (10%). Cyst presentation may be single or multiple.[5]

The clinical presentation of spinal hydatidosis is variable, including paraparesis (62%), paraplegia (26%), back pain or radicular pain (55%), numbness or sensitivity loss (36%) and sphincter disturbance (30%).[6,7] Its asymptomatic and slow evolution disease, so it is usually diagnosed at an advanced stage. Clinical signs are unspecific, they typically include pain, volume increase, and low-energy fractures. Patients may also develop symptoms related to compression of the surrounding structures. Bone cysts may spontaneously rupture and its leakage may result in allergic reactions, fever and rashes. The differential diagnosis should consider Tuberculosis, Chondrosarcoma, Multiple Myeloma, Cancer of unknown primary site, and Giant cell tumor.[8]

Imaging and serological tests may be useful in diagnosing the disease. X-ray imaging, although unspecific, may show multilocular osteolysis with expansion of the bone, thinning of the cortex, radiolucent cystic areas, and reactive sclerosis. CT has high specificity and sensitivity, although its main use limits to preoperative strategy.[3,9] In turn, MRI is useful to assess soft tissue involvement, which, if confirmed, indicates a poor prognosis.[3] The indirect hemagglutination test via ELISA serological test has high specificity (IgG); however, its sensitivity is suboptimal in patients with extrahepatic involvement. Fine-needle aspiration should be avoided because of the risk of spreading the disease.[1]

Braithwaite and Lees[10] have classified the spinal canal involvement into five types, according to its morphological characteristics: 1) primary hydatid intramedullary cysts; 2) intradural extramedullary cysts; 3) extradural intraspinal cysts; 4) hydatid disease of the vertebrae; and 5) paravertebral hydatid disease. The intradural extramedullary involvement is extremely rare, although it may be found mainly in endemic countries and regions. Bone destruction occurs through three mechanisms: compression, ischemia, and osteoclast proliferation around the compressed bone tissue.[3] Articular cartilage and intervertebral discs offer little resistance to the parasite growth.[3]

Our two reported cases belong to the type 3 Braithwaite and Lee classification.[10]

The spreading mechanism of the disease into the spine has not yet been completely outlined. The hydatid disease of the vertebrae usually begins in the vertebral bodies and may progress into any spine level, reaching the spinal canal or the intervertebral discs. The spinal cord compromise is believed to occur through vertebral-portal venous anastomosis. In addition, extradural disease may spread through the neural foramen into the surrounding soft tissues. The mechanisms for the intradural extramedullary involvement are not yet clear. Primary hematic dissemination is regarded as the most plausible mechanism. Another considered mechanism is by contiguity; this mechanism is well known for bacterial dissemination, but not for parasite spread.[11]

Spinal involvement has a poor prognosis, with a high mortality rate (close to 50%), especially when associated with paraplegia. The average length of survival is 5 years.[12] The treatment requires a combination of radical surgery and chemotherapy.[1,13] Surgical procedures are the treatment of choice for spinal cord compression caused by a hydatid cyst. Laminectomy with simple decompression is the most frequent procedure.[13,14] Wide surgical excision is particularly difficult to achieve in the spine and the pelvis as it may compromise critical structures.[1] In addition, in extensive surgical excisions, the long duration of surgical exposure increases the risk of developing an infection. Recurrence rates after the surgical treatment vary from 70% to 80%.[1,2] Cysts must be removed without rupture. Otherwise, the infection spreads to contiguous tissues.

Pharmacological treatment of hydatid disease when combined with surgical treatment seems to achieve better outcomes, with success rates of 25-30% and improvement rates of 40-50%; however, the treatments have been found ineffective in approximately 25-30% of the cases.[14] Albendazole and mebendazole are the main anthelmintic drugs used, the former is regarded by the World Health Organization as the agent of first choice.[14] Nevertheless, bone involvement has demonstrated a less favorable response than other sites. The results of chemotherapy with other agents, such as praziquantel and cimetidine, have been controversial.[15]

In spite of early diagnosis and early institution of combined medical and surgical treatment, the prognosis for bone hydatid disease is still very poor, even comparable to that of a malignant neoplasm, with a latent recurrence possibility.

In conclusion, our two cases of hydatid disease of the vertebrae with intraspinal involvement had an asymptomatic course with a progressive neurological involvement. Both patients required a combined medical and surgical treatment consisting of several surgical interventions and albendazole cycles with favorable clinical outcomes. However, these favorable outcomes were transient due to recurrences and neurological sequelae. In addition, lack of therapy adherence hindered the treatment. Imaging follow-up, particularly with MRI, proved useful in evaluation recurrences and planning new surgical interventions.

These patients treatment had as interfering factors: living in a rural area, a poor adherence to anthelmintic therapy, and failure to comply with their follow-up and control regimen. Therefore, a longstanding, uninterrupted albendazole therapy was not possible to achieve in these patients, as reported by the Infectology Department.

We report these two challenging cases with the aim of sharing our experience and of emphasizing the need to consider Hydatidosis as a tentative diagnosis, especially in rural endemic areas.

Conflict of interest: Authors claim they do not have any conflict of interest.

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