Primary Cystic Echinococcosis of the Spine: A Rare Case Misdiagnosed as Chordoma

Dear Editor,

Cystic echinococcosis is a zoonotic disease caused by infestation of echinococcus tapeworms. It is endemic, especially in Mediterranean countries, and is most commonly localized in the liver and lung in humans. Spinal involvement is rare and usually occurs secondarily in cases with known visceral disease. This study aims to show an uncommon primary spinal cystic echinococcosis with imaging characteristics that resemble chordoma. A 47-year-old female patient with no known pre-existing disease was admitted to Trakya University Hospital with a persistent complaint of right sciatalgia for three months. Neurogenic claudication was positive at 50 meters and otherwise, the neurologic examination was unremarkable. Lumbar magnetic resonance imaging (MRI) and computed tomography (CT) were performed following the patient's complaints [Figure 1]. The patient was diagnosed with chordoma of S1 causing paravertebral extension, spinal stenosis, and infiltrating neural foramina of right L5-S1 and S1-S2 with microfractures. Consequently, the patient was taken to surgery and S1 lamina was revealed in the operation. After right S1 hemilaminectomy, a large number of echinococcosis cysts were detected. It could not be removed with its capsule due to its intramedullary and paravertebral extension. After excision, the cavity was irrigated with 20% hypertonic saline [Figure 2]. Histopathological examination showed multiple daughter cysts with a single membrane, suggesting

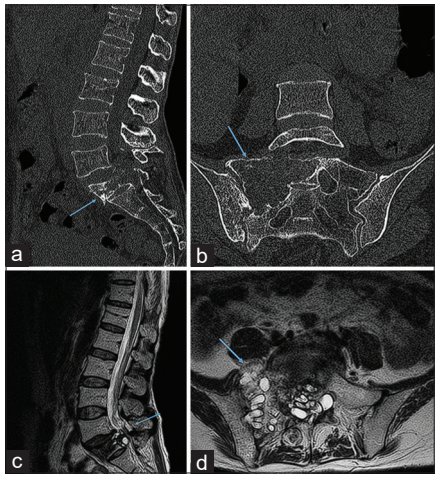

Figure 1: (a) Sagittal CT image shows pathological fracture of S1 vertebral body (blue arrow). (b) Coronal reformat view, lytic lesion with sacral involvement (blue arrow). (c) MRI T2 weighted image, sagittal view shows spinal canal stenosis (blue arrow). (d) MRI T2 weighted image, axial view shows multiloculated cysts with paravertebral extension (blue arrow)

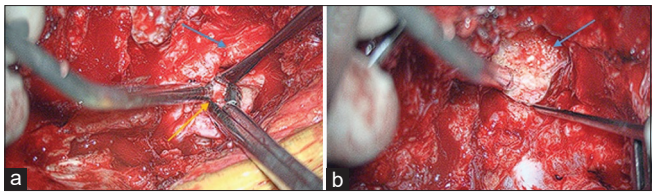

Figure 2: (a) Intraoperative images show the excision of the cyst (yellow arrow) next to the right S1 root (blue arrow) and (b) postoperative large cavity (blue arrow) after excision

the cystic echinococcosis [Figure 3]. Albendazole treatment at a dosage of 400 mg twice a day was started in the postoperative period. The postoperative stage was uneventful. After the second month, the follow-up neurologic examinations were normal. Albendazole treatment continued for a total of five months by giving drug-free intervals for 15 days each month. During the treatment period, liver function tests were under control. The subsequent abdominal ultrasounds and follow-up MRIs revealed no hydatid disease. Long-term monitoring was provided for the patient.

Cystic echinococcosis usually presents with lung or liver involvement in humans. It has been reported that only 0.5%–2% of spinal cystic echinococcosis cases have isolated bone involvement.[1] Although the ways of the spread of the disease are often hematogenous, it can occur in the form of direct invasion or seeding of the cerebral cystic echinococcosis through the cerebrospinal fluid.[2] There was no evidence of visceral cystic echinococcosis disease in our case.

Spinal cystic echinococcosis has no typical symptoms, and depending on the site of involvement, back or low back pain, radiculopathy and neurological deficits due to cord and root compression may be seen. In our case, only sciatalgia and neurogenic claudication were present. Spinal involvement of cystic echinococcosis is often limited to the bone and extradural space. Vertebral body, pedicle, lamina, and paravertebral area involvement may accompany. It has been reported that the intervertebral disc is relatively spared.[3] Further features confirming the diagnosis include the absence of reactive sclerosis and osteoporosis for bone lesions in addition to the usual spherical or tube-shaped multiloculated cysts.[2] Spinal tuberculosis, other vertebral infections, fibrous dysplasia, enchondroma, multiple myeloma, giant cell tumors, and malignancies are included in the differential diagnosis.[4] In the present case, sacrum involvement in addition to the lumbar vertebra, no known hydatid disease, and similar imaging features brought to mind chordoma, which is frequently observed in the sacrum. The specific imaging characteristics of spinal cystic echinococcosis include the presence of the typical multiloculated structure, the t2 low-signal rim, the absence of enhancement until complicated, and the absence of calcification. T2 hypointense rim can be considered in the differential diagnosis of extramedullary cystic lesions of the spinal cord and other cystic lesions of the spinal column. Findings indicative of chordoma that aid in differentiation include the presence of a lytic destructive soft tissue mass,

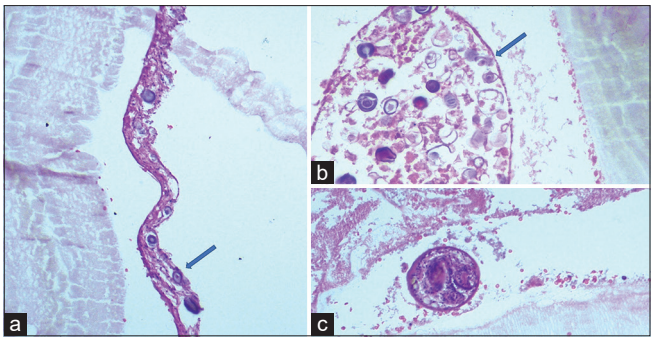

Figure 3: (a) Degenerated, laminar membranous structure on the left side and germinal epithelium with calcified scolex (arrow) on the right side (H and E ×10), (b) Daughter cyst with multiple calcified scolexes (Brood capsule) (H and E ×10), (c) Higher magnification of a scolex (H and E ×20)

heterogeneity caused by necrosis or hemorrhage, intratumoral irregular calcifications, and moderate-to-marked enhancement. The primary and most effective method for treating spinal cystic echinococcosis is surgery. Surgical treatment is usually necessary and urgent due to spinal cord compression or bone fracture as in our case. Anthelminthic therapy is used to reduce the risk of recurrence in the postoperative period. However, by its very nature, cystic echinococcosis has high recurrence rates.

To our knowledge, preoperative diagnosis is crucial for spinal cystic echinococcosis to act accordingly during the surgical approach since the disease recurs frequently.[3] Particularly in endemic areas, cystic echinococcosis should be considered in the differential diagnosis if there are lesions with prominent cystic components in regions like the sacrum where the chordoma is frequently located.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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Submitted: 01-Jun-2023 **Revised:** 21-Jul-2023 **Accepted:** 21-Jul-2023
Published: 27-Sep-2023

DOI: 10.4103/aian.aian_488_23