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Case report

Recurrent spinal hydatidosis causing Gibbus deformity: Report of a rare case

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ABSTRACT

Hydatid disease is caused by the larvae of the parasite Echinococcus granulosus and, less commonly, Echinococcus multilocularis in the endemic regions. A hydatid cyst of Echinococcus granulosus commonly develops in the liver and lungs of the affected individual and rarely occurs in the brain, bones, and heart. The spinal hydatidosis comprises 50% of the cases affecting the bones, which usually leads to severe morbidities. We present a case of a 52-year-old Afghan woman presented to the hospital with Gibbus deformity, complaining of lower back pain and flaccid paraplegia for one year. Findings of Magnetic Resonance Imaging of the lumbosacral spine were suggestive of a hydatid cyst involving the vertebrae with extension to the paravertebral soft tissue, leading to acute kyphosis, severe canal stenosis, and cord compression. Treatment included complete cyst removal. The diagnosis was confirmed by gross and microscopic evaluation of the cyst contents. The patient was discharged after significant pain relief and deformity correction. Vertebral hydatidosis is notorious for its recurrence despite vigorous surgical and medical treatment. The hydatid cyst grows rapidly and can cause irreversible damage to the spinal cord in neglected cases. Currently, the best treatment approach is complete surgical removal of the cyst, irrigation with a scolicidal agent, and postoperative follow-up with a long-term course of albendazole. Spinal hydatidosis is a rare, severe disease with significant morbidity. Despite significant scientific advances in understanding of disease pathogenesis, the parasite's predilection for the spine and the influence of host-specific factors on anatomical manifestations remain a mystery.

Introduction

Hydatid cysts are commonly caused by the larval stage of a tapeworm called Echinococcosis granulosus (EG) [1]. Humans are the accidental intermediate hosts; this parasite mainly infests the liver (70%) and the lungs (20%) [1]. The bones are involved in 0.5–4% of the cases and approximately half of those is in the vertebral column, which is usually associated with a high risk of morbidity and mortality [1]. Hydatid cyst (HC) can reach a growth rate of 7 mm per month, and it can disseminate to paravertebral soft tissue, epidural space, and even the spinal canal by perforating the vertebral cortex [2,3].

The Middle East, East Africa, the Mediterranean region, South America, and several regions of central Europe all have greater rates of hydatid disease (HD) incidence than other regions of the world [4]. Cystic echinococcosis (CE) poses a risk to at least 270 million people in Central Asia, including parts of Mongolia, Kazakhstan, Kyrgyzstan, Tajikistan, Turkmenistan, Uzbekistan, Afghanistan, Iran, Pakistan, and western China, accounting for 58% of the region's total population [5].

Although radiology helps to assess the nature of HC, a definitive diagnosis is usually based on the histopathological examination [6]. A

Abbreviations: EG, Echinococcosis granulosus; HC, Hydatid cyst; HD, Hydatid disease; CE, Cystic echinococcosis; SH, Spinal hydatidosis; GD, Gibbus deformity; MRI, Magnetic resonance imaging.

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combination of surgical and medical therapy is necessary for the treatment of bone hydatidosis [7,8]. Regardless of aggressive treatment, the disease recurrence is high, with vertebral column destruction and neurological impairment [9]. For an early diagnosis and the successful creation of an appropriate treatment strategy, it is helpful to have an accurate and thorough awareness of the clinical and imaging aspects of spinal CE [6]. Here we present a case of recurrent primary spinal hydatidosis (SH) with extension to spinal canal and paravertebral soft tissue, causing severe kyphosis and paraplegia.

Case presentation

A 52-year-old wheelchair-bound female from a pastoral society in Afghanistan was presented to the hospital with Gibbus deformity (GD). She had been complaining of disabling low back pain and flaccid paraplegia for one year. She had been operated on for backache with a diagnosis of primary SH 10 years ago, which resulted in significant pain relief. The patient had been taking albendazole for a few months, but it was discontinued due to the unavailability of health care services in the region. After a year without symptoms, the patient started experiencing mild backache that increased in severity throughout the years with the slow development of spinal deformity and paraplegia. Due to the scarcity of healthcare facilities and socioeconomic problems, the patient couldn't get any diagnostic or therapeutic health services in the region during that period of time.

Upon presentation, the patient was hemodynamically stable, had a fever, and there was a protruding bony deformity localized at the L2-L3 spinal area with associated spinal and paraspinal tenderness. She had an old surgical scar, and her lower extremities were atrophic. Neurological examination revealed bilateral lower extremity flaccid paralysis with paresthesia below the L2 dermatome. No sphincter disturbance was demonstrated. Spinal x-rays demonstrated L2 and L3 vertebral destruction with acute kyphosis formation. Magnetic Resonance Imaging (MRI) T1 and T2 weighted scan of the lumbosacral spine findings were suggestive of a hydatid cyst with bony involvement and extension to the spinal canal, which showed a 10 × 8 × 10 cm enhancing thick-walled cystic lesion with multiple daughter cysts within the left paravertebral region extending from L2 to L5 vertebral level, causing destruction of L2 and L3 vertebrae with acute kyphosis formation. Extension of the lesion in the spinal canal posterior to the bodies of L2 to L4 vertebrae has caused severe canal stenosis and cord compression (Fig. 1).

The indirect hemagglutination test was positive for Echinococcus antibodies. Abdominal ultrasonography and chest X-rays were used to rule out liver and lung hydatidosis. Albendazole 400 mg was administered twice daily to the patient for a week before to the operation.

Under general anesthesia, a posterior midline exposure was carried out from T10 to L5 on the previous surgical scar. Upon exploring, the L2 and L3 vertebra were completely lysed and dural involvement with twisted cauda equina was noted. The cyst was evacuated completely along with its daughter cysts by gently applying pressure to the abdomen without any rupture. Dural excision at the site of attachment to the cyst was done, followed by duroplasty. After rinsing the area with 95% ethanol, the spinal column was stabilized by transpedicular fixation at the level of T12 and L5, and the wound was closed in layers. Patient satisfaction was achieved with the operation's great pain relief and deformity correction, little neurologic development as she was able to sit straight, but regrettably, the paraplegia could not be reversed.

Upon histopathological examination, multiple round and oval daughter cysts of EG were seen grossly that were filled with a clear watery fluid. Microscopically, the Hematoxylin and Eosin (H&E)-stained sections of the cyst-wall fragments showed an acellular laminated layer with a nucleated germinal membrane and brood capsules arising from the germinal membrane. Protoscolices of the parasite surrounded by brood capsules with visible hooklets were noted that confirmed the diagnosis of HC (Fig. 2).

The patient was discharged on the fourth day of surgery with the direction to continue taking 400 mg of albendazole twice daily for the next three months. We kept in touch with the patient over the phone, and she appeared satisfied with the pain alleviation.

Discussion

HD is commonly caused by a zoonotic parasite called EG [10]. Humans are accidental intermediate hosts; the parasite is acquired by humans through direct contact with infected animals or by the feco-oral

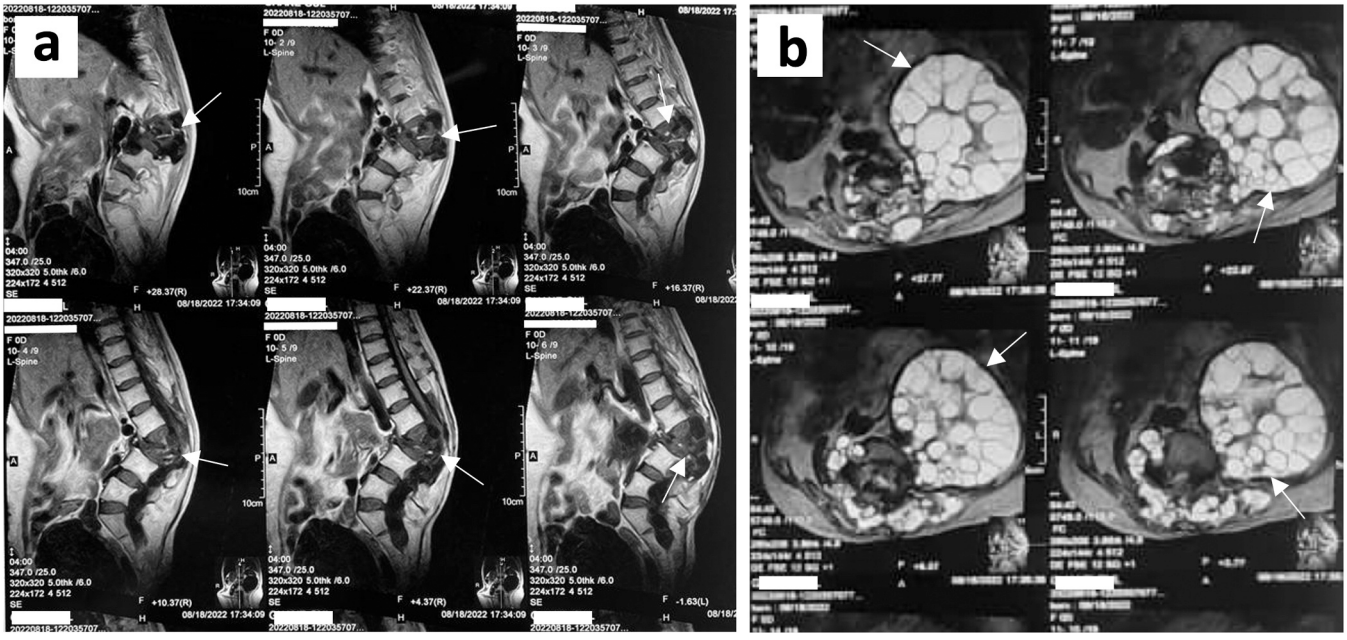

Fig. 1. MRI sagittal and axial scans. On the sagittal plane, destruction of L2 and L3 vertebrae with acute kyphosis formation can be seen with the extension of the lesion in the spinal canal posterior to the bodies of L2 and L4 vertebrae, causing severe canal stenosis and cord compression (a, arrows). The axial plane shows a 10 × 8× 10 cm thick-walled cystic lesion with multiple daughter cysts within, as seen in the left paravertebral region (b, arrows).

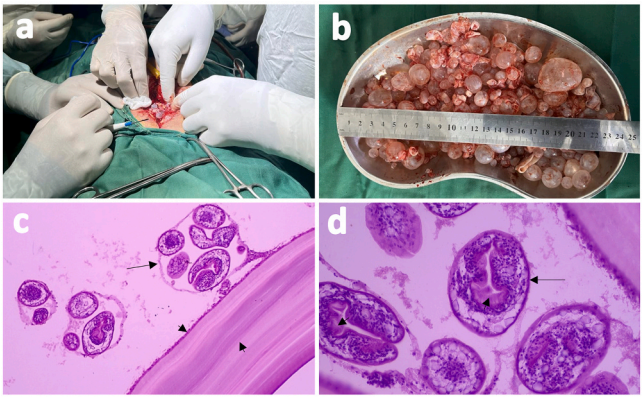

Fig. 2. Intraoperative, gross, and microscopic images of Echinococcus granulosus daughter cysts. Grossly, multiple well-defined round and oval daughter cysts of the EG are seen (a and b). The low- and high-power microscopic Hematoxylin and Eosin (H&E)-stained sections of the cysts exhibit wall fragments composed of an acellular laminated layer with a nucleated germinal membrane (c, arrow heads). Brood capsules (c, arrow), which arise from the germinal membrane, and protoscolices surrounded by brood capsules (d, arrow) with visible hooklets are visible (d, arrow heads).

route [11]. Primary extrahepatic and extrapulmonary hydatidosis occur due to the arterial dissemination of larvae after passing through the liver and lung capillary beds, which are acting as filters [11]. The parasite mainly infests the liver, lungs and bones [1].

Although SH, which is a rare form of the disease, usually presents with nonspecific symptoms, even large cysts may remain asymptomatic [11]. The symptoms of SH are usually due to complications associated with the disease [3]. The most prevalent symptoms and signs of vertebral hydatidosis are paraparesis (62%), increasing back pain (55%), sensory loss (36%), sphincter disturbance (30%), and paraplegia (26%) [3]. In neglected cases, HD can cause catastrophic damage to the vertebrae [11]. HCs can reach a growth rate of 7 mm per month and can disseminate to paravertebral soft tissue, epidural space, and even the spinal canal by perforating the vertebral cortex [1,2].

SH has a poor prognosis, which is often compared to the malignancies in disease severity [1]. It has a recurrence rate of above 50%, that requires a long-term follow-up, and anthelmintic therapy [3].

Due to the low sensitivity of laboratory tests such as Enzyme-linked immunosorbent assay (ELISA), indirect hemagglutination, and complement fixation for extrahepatic hydatidosis, the best diagnostic option is MRI followed by histopathological examination for confirmation [10].

A combination of surgical and medical therapy is considered the best treatment approach for bone hydatidosis [7,8]. The surgical approach can be posterior or anterior depending on where the cysts are located. If the cyst is isolated to the epidural space and there is no involvement of the bone or dura, it is removed by itself. It is necessary to do a dural excision and duroplasty if the dura is attached to the cyst [12]. The cysts should be meticulously removed with copious irrigation with scolicidal agents and standard anti-infectious therapy. In the event of cyst material spillage, recurrence is inevitable, and hypersensitivity reactions, including anaphylactic shock, are expected [3,10]. Other options include minimally invasive techniques like scolicidal agent injection into the cyst via percutaneous puncture-guided sonography with liquid aspiration [4].

The goal of the surgery is to remove the cyst and scolex. However, this objective is rarely achieved. A craniocaudal approach from healthy bone to healthy bone should be used when resecting the affected vertebrae. Following resection, a suitable vertebral stabilization technique should be used. The surgeon should utilize the least amount of stabilization material possible and anticipate repeated procedures in case of recurrence [12].

To reduce recurrence, post-operative albendazole or mebendazole is administered. A careful serologic, radiologic, and clinical follow-up is necessary to identify any potential disease recurrence as soon as possible [4].

In the present case, appropriate surgical intervention included careful removal of the cyst along with daughter cysts, dural excision at the site of attachment to the cyst and duroplasty, irrigation of the area with a scolicidal agent, deformity correction via a posterior approach, and perpendicular screw fixation at the level of T12 and L5.

Conclusion

SH is a rare, severe disease with significant morbidity. Despite significant scientific advances in understanding of disease pathogenesis, the parasite's predilection for the spine and the influence of host-specific factors on anatomical manifestations remain a mystery. As HD is well-known for its high recurrence rate, a careful follow-up is necessary to identify any potential disease recurrence as soon as possible.

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Consent

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CRediT authorship contribution statement

Abdullah Ahmad: Conceptualization, Data curation. **Torgot Ghani:** Writing – review & editing, Data curation. **Ahmed Nasir Hanifi:** Supervision. **Sayed Amrullah Faez:** Supervision. **Zekrullah Baset:** Data curation. **Haider Ali Malakzai:** Writing – review & editing, Data curation, Supervision. All authors have seen and approved the final manuscript.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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