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CASE REPORT

Intradural extramedullary primary hydatid cyst of the spine in a child: a very rare presentation

S. H. Arif · Sufian Zaheer

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Abstract Spinal hydatid cyst is a serious but fortunately uncommon manifestation of the parasite Echinococcus, involving less than 1% patients with hydatid disease. Intradural hydatid cysts are extremely rare compared to other types of spinal hydatid cysts. We report a rare case of intradural, extramedullary spinal hydatid cyst in a 9-year-old male boy, who presented with weakness of both lower limbs for the last 4 months that was confirmed histopathologically; a better understanding of this rare but clinically challenging disease is intended by reporting this case.

Keywords Hydatid cyst · Spine

Introduction

Hydatid is a Greek word meaning "watery cyst". Hydatid disease is caused by the parasitic tapeworm Echinococcus (*E. granulosus*, and less commonly *E. multilocularis*) [6]. The infection with this parasite is endemic to sheep-raising areas of the world. Humans are an accidental intermediary host, with lungs and liver being the most commonly affected sites Hydatidosis involving the spine comprises less than 1% of the total cases of hydatid disease [5]. On reviewing the English language medical literature to the best of our knowledge, we found this to be the second pediatric case of primary intradural extramedullary hydatid cyst, among the 26 cases of primary intradural extramedullary hydatid cyst reported so far. The first pediatric case of intradural extramedullary hydatid cyst was reported in a 8-year-old male boy in 2007 by Kalkan et al. from Turkey [5].

S. H. Arif · S. Zaheer (✉)
Department of Pathology,
Jawaharlal Nehru Medical college Hospital,
Aligarh Muslim University, Aligarh, India
e-mail: sufianzaheer@gmail.com

Case report

A 9-year-old boy presented to our institution, a tertiary care medical referral center, with a complaint of weakness in the both lower limb for the last 4 months. There were no other neurological abnormalities at the time of admission. Plain X-ray showed no bony abnormalities. Laboratory data including hemoglobin, total blood count, erythrocyte sedimentation rate, liver function test, renal function tests, and coagulation profile were normal.

On magnetic resonance imaging (MRI) examination, an intradural, extramedullary cystic lesion was seen extending from L1 to L4 spine measuring 9.8 × 1.8 cm with a uniform and regular contour. The lesion was hypointense on T1- and hyperintense on T2-weighted (Fig. 1) MRIs. The radiological diagnosis was made of arachnoid cyst.

The cystic lesion measuring 10 × 2 cm was removed totally with its capsule (Fig. 2) and the specimen was submitted for histopathological examination. The histopathological examination showed characteristic cuticular layer of the cyst wall in the form of amorphous densely staining laminated chitinous material (Fig. 3). Characteristic scolices were also visualized with its hooklets confirming the diagnosis (Fig. 4).

Postoperatively the patient was receiving a minimum 6-month course of albendazole. The patient's follow-up visits showed that the weakness of the lower limb gradually

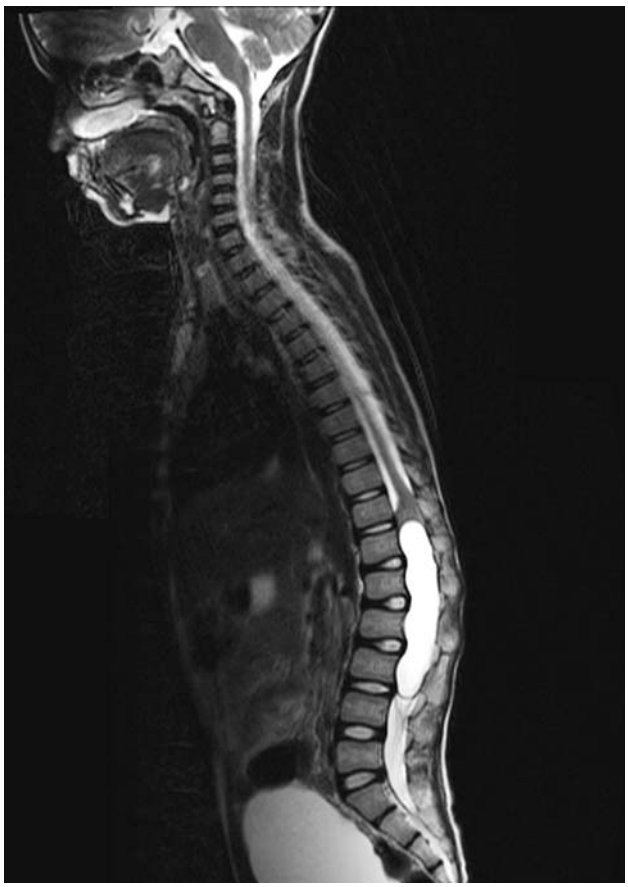

Fig. 1 MRI spine (T2W1)—showing hyperintense cystic lesion is seen extending from L1 to L4 spine

recovered. Postoperative MRI at 6 months follow-up did not show recurrence of spinal cystic lesion and neither it was present in any other parts of the body.

Discussion

Hydatid disease (HD), a rare parasitic infection caused mainly by the larval form of Taenia *Echinococcus granulosus*, and less commonly by *Echinococcus multilocularis*, the latter primarily causing alveolar Echinococcus [6].

Cases of HD are commonly seen in areas where canine dogs (definitive host) and livestock (intermediate host) including sheep coexist. The most common sites of infection are liver (75%), lung (15%), brain (2–4%), and genitourinary tract (2–3%) [7]. Bone involvement is very rare (1%). Spine is the most common site for the hydatid disease of the bone. According to İslekel et al. [4] bone disease was first described by Bidloo in 1708, the first description of spinal hydatid disease was made by Churrier in 1807 and the first surgical intervention was reported by Reydellet in 1819.

Fig. 2 The cystic lesion measuring 10 × 2 cm was removed totally with its capsule

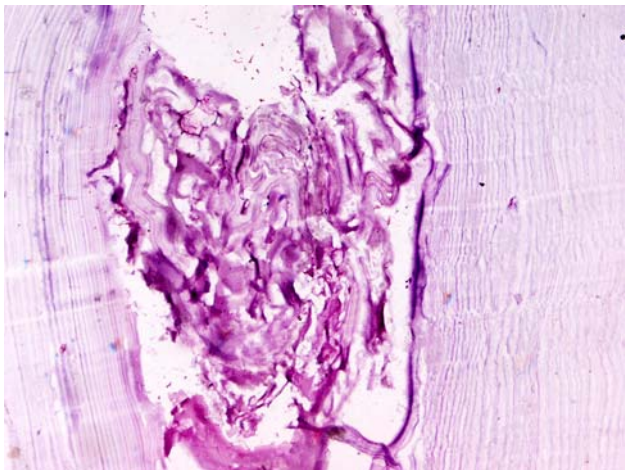

Fig. 3 Pictomicrograph shows characteristic cuticular layer of the cyst wall in the form of amorphous densely staining laminated chitinous material

Braithwaite and Lees [1] have classified the spinal involvement by hydatid disease in five groups: (1) primary intramedullary hydatid cyst; (2) intradural extramedullary hydatid cyst; (3) extradural intraspinal hydatid cyst; (4) hydatid disease of the vertebrae; and (5) paravertebral hydatid disease. The first three types in this group are considered rare.

According to Schnepper and Johnson [8] spinal involvement is believed to occur through vertebral–portal venous anastomosis. The intervertebral discs, however, are

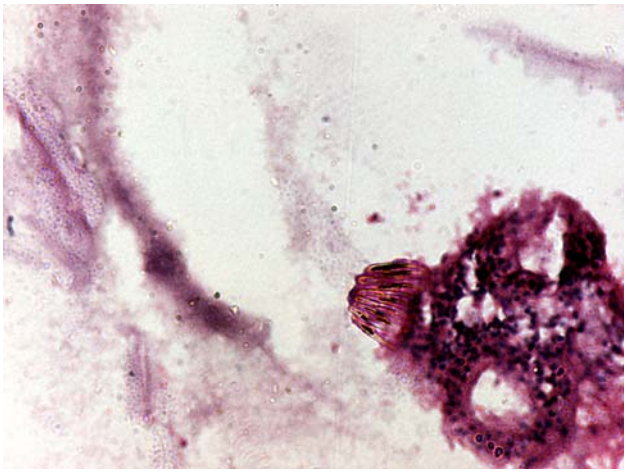

Fig. 4 Characteristic scolex with its hooklets

usually spared because the cyst growth is confined within the periosteum. Spinal hydatid cysts are located most commonly at the thoracic (52%), followed by the lumbar (37%) and then the cervical and sacral levels [8].

Histopathologically, three layers can be identified in the wall of the hydatid cyst: a peripheral adventitial layer which consists of fibrous tissue containing many eosinophils, an intermediate cuticular layer containing amorphous densely staining laminated chitinous material, and an inner germinal layer that contains nucleated epithelium. It is the germinal layer that gives rise to brood capsules, and scolices (the larval stage of the parasite) develop within these vacuolated structures. The brood capsules (daughter cysts) eventually detach and float freely in the fluid of the hydatid cyst. The number of scolices increases within the daughter cysts over time, causing enlargement of the cyst. The pathology of hydatid disease is caused by the mass effect of the growing cyst [7].

Radiological studies are usually inconclusive but may be helpful in the diagnosis of hydatid disease. Spherical cysts with peripheral calcification may be seen on a plain radiograph and are indicative of hydatid cyst. CT scan and MRI are the current diagnostic modalities of choice. Irregular bony erosions along with multilocular, non-enhancing flattened sausage-shaped lesions with very thin, non-septated walls are the hallmark of vertebral hydatid disease. Extraosseous lesions are usually unilocular with thicker cystic walls and acquire a spherical shape due to the lack of resistance from the hard bone tissue during their growth; calcifications may develop as the disease progresses [9]. Magnetic resonance imaging shows cyst which generally have two dome-shaped ends, which usually have no debris in the lumen, and usually look like flattened sausages, with thin, regular walls without septations. Intradural cysts may be single or multiple; extradural cysts are always multiple and involve the bone [3]. On MRI, the lesions appear isointense to the cerebrospinal fluid (CSF) (hypointense on T1-weighted images and hyperintense on T2-weighted images) [2, 9].

For the treatment of spinal hydatid disease, surgery is currently considered the treatment of choice [4, 10]. Fortunately as most of the cases are admitted with sign and symptoms of spinal cord compression syndrome, surgeons usually act urgently. It is of crucial importance that the diagnosis of spinal hydatid disease and its extent be properly made out before operating the patient [10].

Posterior approach is usually preferred for pure intradural or pure epidural lesions at all the spinal levels. In these cases it is important that complete excision without intraoperative rupture of the cyst be performed otherwise the risk of recurrence is increased substantially [10]. Involvement of the bone is the major challenge for the operating surgeon. As the involvement of the bone amounts to infiltrative lesion, the chances of spillage are unavoidable [4, 10].

Postoperative risk of recurrence is up to 40% even after extensive surgery. The major goal in recurrent lesion is restoration and preservation of neurological status and spinal stability [11]. Many surgeons also advocate reinstitution of anti-helminthic pharmacotherapy and its indefinite continuation. If there is recurrence of spinal disease it is considered a bad prognostic sign [11]. However if the lesion is multiply recurring the primary goal should be spinal stability.

We want to report this case due to its uniqueness, both because of the young age of the patient and its intradural extramedullary location. Only one similar case has been reported in a child in the English language medical literature to the best of our knowledge. In fact, our case was somewhat different from this single case reported as it was reported to be present in the intradural extramedullary region at the level of the thoracic spine and our case was found to be at the level of lumbar spine. Further our aim of reporting this case is to bring home the point that, although spinal hydatid disease is rare (especially intradural, extramedullary location), but it should always be considered in the differential diagnosis of spinal cord compression syndrome or space occupying lesion in endemic countries and should be confirmed by imaging, serology and histopathological examination.

Conflict of interest statement None of the authors has any potential conflict of interest.

References

1. Braithwaite PA, Lees RF (1981) Vertebral hydatid disease: radiological assessment. Radiology 140:763–766

2. Chang KH, Han MH (1998) MRI of CNS parasitic diseases. J Magn Reson Imaging 8:297–307. doi:10.1002/jmri.1880080209
3. Fahl M, Haddad FS, Huballah M et al (1994) Magnetic resonance imaging in intradural and extradural spinal echinococcosis. Clin Imaging 18:179–183. doi:10.1016/0899-7071(94)90078-7
4. İslekel S, Zileli M, Ersahin Y (1998) Spinal hydatid disease. Spinal Cord 36:162–164
5. Kalkan E, Cengiz SL, Çiçek O, Erdi F, Baysefer A (2007) Primary spinal intradural extramedullary hydatid cyst in a child. Spinal Cord Med 30(3):297–300
6. Pamir MN, Ozduman K, Elmaci I (2002) Spinal hydatid disease. Spinal Cord 40:153–160. doi:10.1038/sj.sc.3101214
7. Rosenblatt GS, Walsh CJ (2001) Recurrence of genitourinary hydatid cyst after nephrectomy. Infect Urol 14(3):75–79
8. Schnepper GD, Johnson WD (2004) Recurrent spinal hydatidosis in North America: case report and review of the literature. Neurosurg Focus 17(6):1–6. doi:10.3171/foc.2004.17.6.8
9. Tsitouridis I, Dimitriadis AS (1997) CT and MRI in vertebral hydatid disease. Eur Radiol 7:1207–1210. doi:10.1007/s003300050275
10. Pamir MN, Akalan N, Ozgen T, Erbengi A (1984) Spinal hydatid cysts. Surg Neurol 21:54–57. doi:10.1016/0090-3019(84)90402-6
11. Ozek MM (1994) Complications of central nervous system hydatid disease. Pediatr Neurosurg 20:84–91. doi:10.1159/000120770