Eur Spine J
DOI 10.1007/s00586-016-4896-3

CASE REPORT

Primary intramedullary hydatid cyst: a case report and literature review

Zhenhai Zhang[1] · Juan Fan[1] · Yuanyuan Dang[1] · Ruxiang Xu[1] · Chunsen Shen[1]

Received: 15 October 2016 / Accepted: 22 November 2016

Abstract Intramedullary hydatid cyst is extremely rare. We present a case of pathologically confirmed primary intramedullary hydatid cyst in an otherwise healthy patient. A 17-year-old boy presented with lumbar pain, weakness, and numbness in both lower limbs, and urinate difficulty interrupted for 2 years. The patient had no other signs of systemic hydatid cyst disease. An intramedullary cystic lesion was identified with magnetic resonance imaging and was shown to be a hydatid cyst by histopathologic examination after the surgical removal. Although extremely rare, primary intramedullary hydatid cyst pathology might be the cause of lumbar pain, weakness, and numbness in both lower limbs for those living in endemic areas. Surgical removement is feasible and effective for intramedullary hydatid cyst.

Keywords Spine · Hydatid disease · MRI · Intramedullary · Surgery

Introduction

Hydatid disease is caused by the parasitic tapeworm *Echinococcus*, including the larval stage of *Echinococcus granulosus*, and less commonly *Echinococcus multilocularis* [1]. The *Echinococcus* can infect many hosts, including sheep, cattle, goats, and humans. Humans could be contaminated through direct contact with the definitive host or by ingesting food infected with parasite eggs [2]. It is endemic in North Africa, Spain, Greece, Turkey, Portugal, Middle East, Australia, New Zealand, South America, Baltic areas, and the Philippines [3]. The most common sites of infection are liver (75%), lung (15%), brain (2–4%), and genitourinary tract (2–3%) [1]. Spinal involvement is rare, with an incidence of 1% in all cases of hydatid disease [4]. Spinal hydatid cyst disease presents with cauda equina symptoms or symptoms of cord compression [5]. It is typed into five: (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 [6]. Intramedullary involvement is only reported in two cases [7, 8]. To the best of our knowledge, the present study is the first case of primary intramedullary hydatid cyst in the thoracic region reported in the literature.

Case report

Presentation and examination

A 17-year boy admitted to our hospital in February in 2016. He presented with lumbar pain, weakness, and numbness in both lower limbs, and urinate difficulty interrupted for 2 years. 10 days before arriving at the hospital, he had experienced difficulty with walking, and his feet had no sense of cold and hot. Neurological examination revealed bilateral lower extremity weakness, spastic paraparesis, and hypoesthesia below T10. In addition, he had obvious chest bondage feeling and abdominal tension. He grew up in a rural area and was in frequent contact with farm animals, such as horse, cow, and sheep.

✉ Chunsen Shen
gfzzh@126.com

[1] Affiliated Bayi Brain Hospital, PLA Army General Hospital, Nanmencang, 5, Dongcheng, Beijing 100700, China

Published online: 28 November 2016

Neuroimage

The cyst had thin walls and cerebrospinal fluid-like signal intensity on MRI. The lesion was hypointense on T1-weighted images and hyperintense on T2-weighted images. The body of T7–T9 vertebrae and the paraspinal muscles appeared to be intact. Cranial, cervical, lumbosacral,lung, and abdominal computed tomography were carried out without any other signs of systemic hydatid cyst disease.

Operation and histopathological examination

According to MRI, enterogenous cyst was diagnosed (Fig. 1). Due to the patient's rapid progression and serious presentation, he underwent an emergency surgery, using a posterior approach. T8 was first marked with Methylene Blue. A T7–T9 laminectomy was performed. After opening the dura, the spine was still intact. The operation went on into the intramedullary by median myelotomy, and a whitish, pearl-like, semitranslucent, cystic material was found. The cyst was excised in a whole, followed by dexamethasone irrigated the operative site to avoid an anaphylactic reaction. The spinal dura was watertight sutured. The wound was closed in layers. In the histopathological examination, the cyst wall was noted to stain with Hematoxylin and Eosin (H&E). Granuloma and inflammatory granulation tissue constituted with hyperplastic fiber and vessel, epithelioid cell, and multinucleated giant cells. The hydatid cyst and *Echinococcus* could be seen. The chronic inflammatory cells existed in interstitium, which supported the presence of a heavy inflammatory reaction (Fig. 2).

Postoperative course

The patient's showed paraplegia in 2-day post-operation and improved gradually later. He could ambulate independently at 9-month follow-up, and radiological evaluation showed no evidence of disease recurrence (Fig. 3).

Discussion

If the worm is not lodged in liver or lungs, it may be trapped virtually anywhere in the body, such as peritoneum, spleen, kidney, heart, brain, spine, bony skeleton, and muscles [9]. Up to now, only two intramedullary hydatid cases have been reported. Ley A Jr reported a 28-year-old man admitted for urinary incontinence, severe leg pain, and paraparesis of a few months' duration. He had an operation for hydatid cyst in the lung. A syringostomy was performed, and drainage was established. 4 months later, secondary operation was performed for a complete paraplegia. An intramedullary cystic mass was found and completely removed. However, the patient remained paraplegic after 8-month follow-up [7].

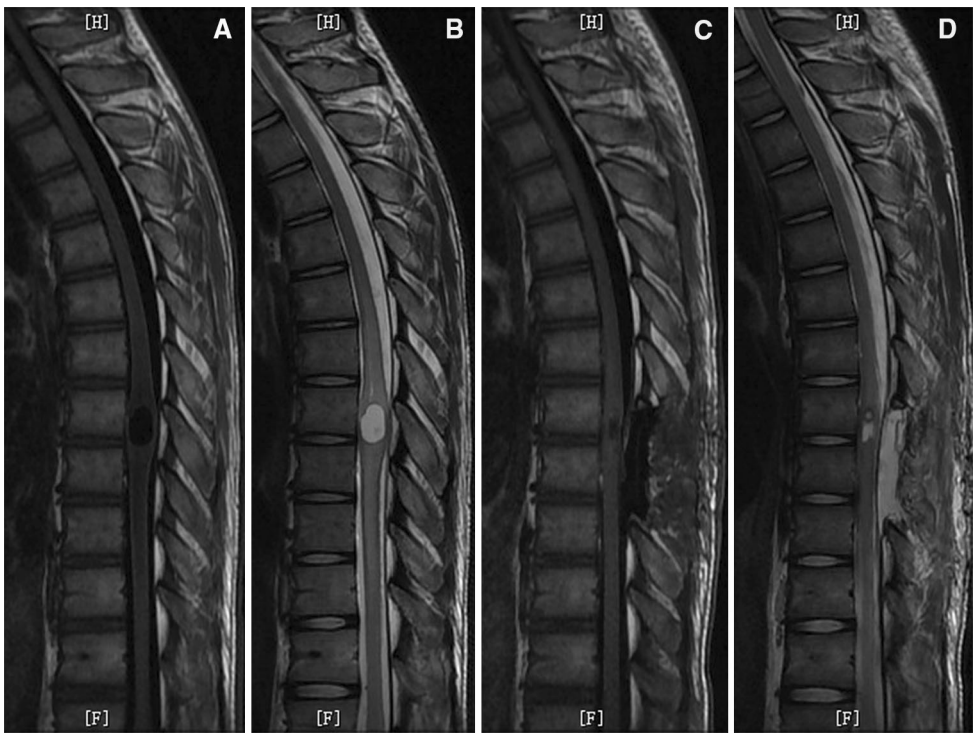

Fig. 1 **a**, **b** Magnetic resonance imaging of the thoracic spine showed an intramedullary cysticlesion; **c**, **d** MRI post-operation showed that the cysticlesion was removed

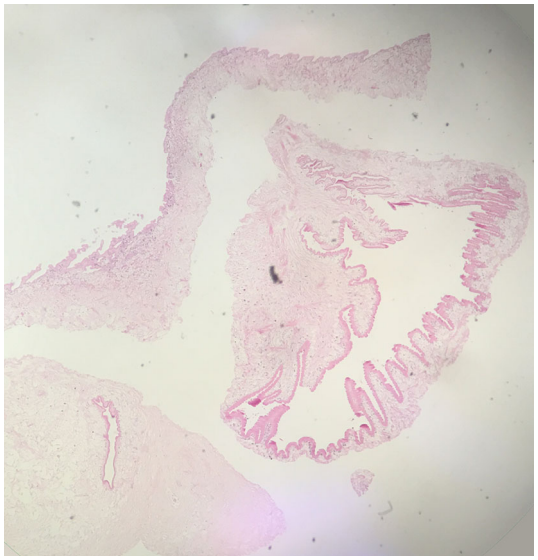

Fig. 2 In the histopathological examination, the cyst wall was noted to stain with Hematoxylin and Eosin (H&E). The *Echinococcus* could be seen

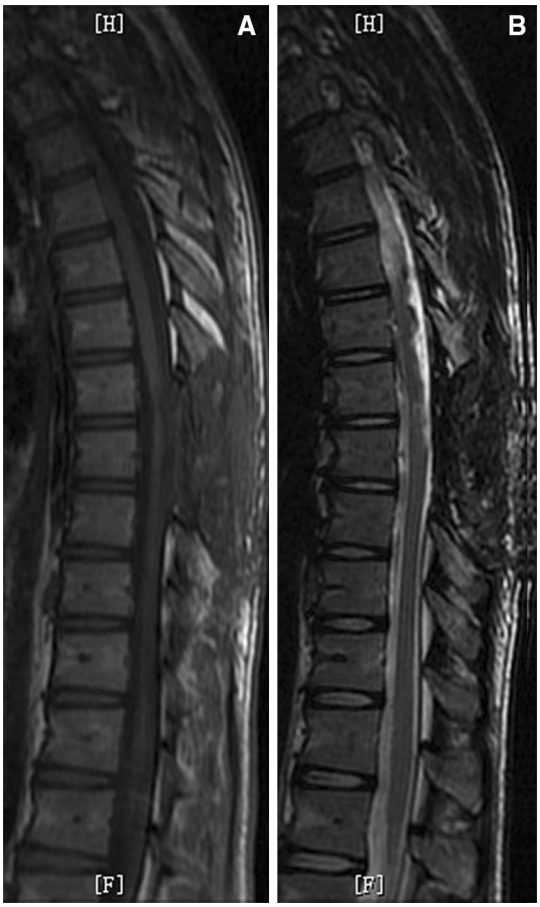

Fig. 3 9 months after operation, MRI, showed no evidence of recurrence

Senol MG reported a 55-year-old female patient of intramedullary hydatid cyst in cervical spine, multiple cysts in her liver, spleen, and lungs, and thoracic vertebrae were also detected by thoracic and abdominal computed tomography. Serological tests confirmed the diagnosis of hydatid disease. Cysts in her lung, spleen, and liver were successfully removed by consecutive operations. Surgery for intramedullary cyst was avoided because of high risk. Clinical symptoms improved after albendazole treatment during 6 years of follow-up [8].

We had misdiagnosis before surgery because of the rarity of the case. MR is of great use for diagnosis before surgery, as well as finding recurrence soon. The cysts consist of an outer fibrous layer and a cestode-derived inner germinal membrane containing scolices [10]. Histopathologically, three layers of the hydatid cyst can be identified, including 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. Cysts occur in two types: unilocular and multilocular. Unilocular cyst is more common (57.5%) [11]. Differential diagnosis of intramedullary cystic masses includes syrinx cavities, myelomalacia, hematoma, arachnoid cyst, neurenteric cyst, epidermoid cyst, cystic hemangioblastoma, and astrocytoma-like cysticneoplasms [12]. The early and correct diagnosis is needed. When we encounter this well-defined, smooth thin-walled, homogeneous cysts, where the fluid appears to be similar to the cerebrospinal fluid, we should highly suspect hydatid disease, especially for those living in endemic areas.

With the development of surgery technique, surgical removal of the intramedullary cyst is feasible. Total removal without cyst rupture should be aimed. Cyst rupture can lead to recurrence [13]. Spillage of the content may provoke a variety of hypersensitivity reactions, such as pruritus, urticaria, edema, dyspnea, asthma, vomiting, diarrhea, colicky abdominal pain, and even anaphylactic shock [9]. The recurrence rate is reported 30–40% [5]. The cyst becomes fertile in 6–9 months [14], which is a key time window for estimating recurrence.

Conclusion

Although extremely rare, primary intramedullary hydatid cyst pathology might be the cause of lumbar pain, weakness, and numbness in both lower limbs for those living in endemic areas. Surgical removement is feasible and effective for intramedullary hydatid cyst.

Compliance with ethical standards

Informed consent Informed consent was obtained from all individual participants included in the study.

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

References

1. Arif S, Zaheer S (2009) Intradural extramedullary primary hydatid cyst of the spine in a child: a very rare presentation. Eur Spine J 18(Suppl 2):179–182
2. Baykaner M, Dogulu F, Ozturk G, Edali N, Tali T (2000) A viable residual spinal hydatid cyst cured with albendazole. J Neurosurg 93(1 Suppl):142–144
3. Thaler M, Gabl M, Lechner R, Gstottner M, Bach C (2010) Severe kyphoscoliosis after primary *Echinococcus granulosus* infection of the spine. Eur Spine J 19(9):1415–1422
4. Lakhdar F, Arkha Y, Rifi L, Derraz S, Ouahabi A, Khamlichi A (2009) Spinal intradural extramedullary hydatidosis: report of three cases. Neurosurgery 65:372–377
5. Pluchino F, Lodrini S (1981) Multiple primitive epiduralspinal-hydatidcysts: case report. Acta Neurochir (Wien) 59:257–262
6. Braithwaite PA, Lees RF (1981) Vertebral hydatid disease: radiological assessment. Radiology 140:763–766
7. Ley A Jr, Marti A (1970) Intramedullary hydatid cyst. Case report. J Neurosurg 33(4):457–459
8. Senol MG, Tekeli H, Kendirli MT, Kaya S, Turhan V, Sonmez G, Saracoglu M (2012) Intramedullary hydatid cyst of the cervical spine. Indian J Med Microbiol 30:480–481
9. Pamir M, Zduman K, Elmaci I (2002) Spinal hydatid disease. Spinal Cord 40(4):153–160
10. Kahilogullari G, Tuna H, Aydin Z, Colpan E, Egemen N (2005) Primary intradural extramedullary hydatid cyst. Am J Med Sci 329(4):202–204
11. Lotfinia I, Sayyahmelli S, Mahdkhah A, Shoja MM (2013) Intradural extramedullary primary hydatid cyst of the spine: a case report and review of literature. Eur Spine J 22(Suppl 3):S329–S336
12. Guzel A, Tatli M, Yilmaz F, Bavbek M (2007) Unusual presentation of cervical spinal intramedullary arachnoid cyst in childhood: case report and review of the literature. Pediatr Neurosurg 43:50–53
13. Prabhakar M, Acharya A, Modi D, Jadav B (2005) Spinal hydatid disease: a case series. J Spinal Cord Med 28:426–431
14. Araj GF, Matossian RM, Malakian AH (1977) The host response in secondary hydatidosis of mice. II. Cell mediated immunity. Z Parasitenkd 52(1):31–38