Primary spinal extradural hydatid cyst associated with acute bleeding

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Aims: The purpose of this article is to report a case of unilocular primary spinal extradural hydatid cyst which manifested as acute bleeding. Methods: The clinical presentation, diagnosis, and surgical treatment of this rare case are discussed and published cases of primary extradural hydatid cysts are reviewed. Results: Complete recovery was achieved. Repeated clinical, radiological, and serological examinations did not show any evidence of local recurrence or systemic hydatidosis during the follow-up period of 50 months. Conclusions: Primary spinal extradural hydatid cyst may present as acute bleeding. (Wang Y, Geng D, Zhu G, Du G. North Am J Med Sci 2009; 1: 78-81).

Keywords: Spinal cord compression, parasitic infections, tape-worm, echinococcosis, hydatid cyst, complications, extradural hematoma, spasmodic torticollis, laminectomy.

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Introduction
The classical findings of hydatid disease are well documented. Infestation by hydatid disease in humans most commonly occurs in the liver (75%) and lungs (15%); these two organs can be affected simultaneously in about 5~13% of cases. In addition, it can also affect the brain, heart, kidneys, ureter, spleen, uterus, fallopian tubes, mesentery, pancreas, diaphragm, and muscles. The clinical presentation of hydatid disease depends on the size and site of the lesion and the accessibility of the organ involved for clinical examination.

Hydatid disease of the spine occurs in only 1% of all cases of hydatidosis and is mostly located in the intradural region [1]. Primary isolated spinal extradural hydatid cyst is a very rare condition, and only a few cases have previously been reported [1-6]. When the cysts do not show characteristic features on magnetic resonance imaging (MRI), the differential diagnosis is quite difficult and the lesion can imitate different cystic pathology. The surgical intervention is always for the management of spinal hydatid cyst, thus, an incorrect preoperative diagnosis may compromise management of the lesion or lead to complications, such as intraoperative cyst rupture.

In this article, we report a case with unilocular spinal extradural hydatid cyst, which manifested with acute bleeding in the cyst.

Case report
A 3-year-old boy with an unremarkable previous medical history was referred to our department in January 2004 with rapidly progressive weakness of the bilateral upper and lower limbs throughout the prior five days. There was a history of head rotation and leaning consistently to the right that had developed over a three-week period. A clinical examination revealed neck stiffness and head rotation similar to that of spasmodic torticollis: the face turning to the left side and the head pulling to the right shoulder. Muscle strength was decreased and muscle tone increased in the bilateral upper and lower limbs. All the deep tendon reflexes became brisk, and ankle clonus was observed. Hemogram and routine blood chemistry were normal. Plain radiographs of the cervicothoracic spine did not reveal any abnormality, and chest radiograph was also normal. MRI study revealed an oval, unilocular, well-defined extradural lesion posterior to the thecal sac opposite the C6-T2 vertebrae. The lesion was hyperintense on both the T1W and T2W images. The spinal canal at the same level was slightly enlarged with a smooth edge. The lesion was compressing the underlying spinal cord, which showed hyperintensities on T2W images consistent with myelomalacic changes (Fig. 1).

Post-contrast study did not reveal any enhancement of the lesion. Vertebral bodies, pedicles, and posterior elements appeared normal. No pre- or paravertebral soft tissue masses were seen. Based on these imaging findings, we made a diagnosis of a spontaneous extradural hematoma with evidence of underlying compressive myelomalacia (at the C4-T3 level). Surgery was performed with the patient in the sitting position. C6 to T2 laminectomy was performed, and a cystic lesion was seen extradurally on the left side. The cyst contained black blood within a wall which was more than 1mm thick. The hematoma was evacuated and the cyst wall was removed. Cord decompression was performed. The postoperative course was uneventful, and the child made a gradual neurological recovery following surgery. The diagnosis of hydatid...
disease was confirmed upon histopathology (Fig. 2).

Postoperative sonography of the abdomen revealed no evidence of hydatid disease, thus albendazole was given orally for one month for prophylactic purposes. During the follow-up period of 50 months, repeated clinical, radiological, and serological examinations (every 6 months) did not show any evidence of local recurrence or systemic hydatidosis. MRI scanning showed that the spinal cord had recovered completely by 50 months after surgery (Figs. 3, 4).

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Fig. 1 Preoperative T1- and T2-weighted images. Demonstration of scoliosis and an oval, unilocular, well-defined extradural lesion posterior to the thecal sac, opposite the C6-T2 vertebrae. The lesion was hyperintense both on T1W and T2W images. The lesion was compressing the underlying spinal cord which showed hyperintensities on T2W images, consistent with myelomalacic changes.

Fig. 2 Microphotograph of the lesion reveals laminated membrane. The inner layer (germinative membrane) was composed of a germinal epithelium (A, 10×). The outer layer (cuticular layer) contained white colored chitin (B, 40×).

Fig. 3 Postoperative T1- and T2-weighted images 3 months after surgery. Demonstration of recovery from scoliosis; the spinal cord showed slight hyperintensities on T2W images.

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CT has proven helpful in recognizing the occurrence of hydatid cysts, playing a complimentary role to MRI [9, 10]. It can reveal intradural or extradural multiloculated cystic masses, the extent of bone destruction, widening of the spinal canal, extension of the disease into adjacent soft tissues, and possible calcifications of the peripheral rim. However, MRI is superior to CT in showing the cyst itself. Previous MRI studies reported in the literature [9, 10] have revealed primary extradural hydatid cysts manifesting as multiple small extradural cysts or as a multiloculated cyst. In our case, MRI revealed an extradural hematoma. To our knowledge, this phenomenon has not been previously reported.

It is reported that there is no development of an adventitious layer or pseudocapsule, which occurs in soft tissues where the cyst usually grows by endogenous vesiculation and radial expansion to produce a spherical cyst with compression of host tissue. In our case, there was only a thick white wall >1mm, similar to the endocyst in the liver or lungs. Histologic examination of the cyst wall revealed that it was composed of laminated membrane (Figure 2). Local complications of hydatid cysts include cyst rupture and infection. Rupture may occur due to minor trauma or it may take place spontaneously. Although an endocyst rupture in the liver may sometimes cause intracystic or extracystic hematoma, we have not found any spinal hydatid cyst case reports in which the cyst ruptured and formed a hematoma. The cyst in our case remained intact during surgery, with the hematoma contained completely within the cyst. The cause of the bleeding is still unknown. The cyst extended from C6 to T2 in the spinal canal, so we assume that the small vessel was torn by the formation of an intracystic hematoma when compressed by head and neck movement.

Surgery remains the preferred treatment for extradural hydatid cysts, where spinal cord decompression is the main purpose of the surgery [11, 12]. Ideally, treatment would entail total excision of all affected tissue, but this is very difficult in most cases. Except for preoperatively diagnosed cases, rare isolated intradural or extradural cysts without bone involvement can be removed unruptured. For this reason, recurrence is common when cysts rupture during surgical removal, causing diffuse spreading within the bone and spinal canal. Therefore, the intra-operative use of hypertonic saline or 0.5% silver nitrate solutions before opening the cavities tends to kill the daughter cysts and prevents further spread or anaphylactic reaction. Many authors [8, 11, 12] believe that after surgery, antihelminthic treatment is indicated even when the cyst remains intact during surgery. This is prescribed because microinfiltration of the vertebrae cannot be shown by radiological exploration. Bone infiltration makes radical

Discussion

Hydatid is a condition caused by the cyst stage of an infestation by the tape-worm *Echinococcus granulosus*. The adult form of the parasite lives in the intestinal tract of canines, whereas sheep, cattle, and humans are intermediate hosts. In humans, hydatid disease affects both sexes equally but occurs mainly in the younger population. Hydatid cysts are mostly located in the liver but can affect any organ of the infected host except hair, teeth, and finger nails. Hydatid disease involves the liver in approximately 75% of cases, the lungs in 15%, and other anatomic locations in 10% [7]. It affects bone in 0.5-2% of cases, and the spine is involved in approximately 45% of the cases with bone involvement. Cysts are most commonly located in the thoracic spine. Since the primary site of the infestation and the precise extent of the disease are very difficult to verify, Braithwaite and Lees have classified this disease into 5 types [8]: 1) primary intramedullary hydatid cyst; 2) intradural extramedullary hydatid cyst; 3) extradural intraspinal hydatid cyst; 4) hydatid disease of the vertebrae; and 5) paraspinal hydatid disease. We classified our patient as type 3. This disease occurs either by direct extension from a nearby infestation or, less often, in the vertebral body. Consequently, true intramedullary cysts and intradural extramedullary cysts are very rare.

Only a few cases have been reported in the literature. Generally, spinal hydatid cyst disease presents with radicular symptoms or symptoms of cord compression. In our case, head rotating and leaning consistently to the left were the first symptoms, although others may have not been recognized by the parents or were not clearly expressed by the young boy.

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![Fig. 4 Postoperative T1- and T2-weighted images 50 months after surgery. Demonstration that the spinal cord recovered completely and the spinal canal narrowed slightly without compressing the cord.](image-url)
excision difficult, and subsequently there is a high incidence of recurrence. If a diagnosis is made before surgery, pre- and postoperative 1-month courses of albendazole and 2 weeks of praziquantel should be considered in order to sterilize the cyst, decrease the chance of anaphylaxis, decrease the tension in the cyst wall (thus reducing the risk of spillage during surgery) and to reduce the recurrence rate postoperatively. Albendazole decreases ATP production in worms, causing energy depletion, immobilization, and, finally, death. To avoid an inflammatory response in the CNS, the patient must also take anticonvulsants and high-dose glucocorticoids. The prognosis for neurological recovery in hydatid disease of the spine is thought to be poor, and the disease has a reported mortality rate of more than 50%. However, despite preoperative misdiagnosis and a cyst that was not removed in toto during the surgical procedure, complete recovery was achieved. In this case, repeated clinical, radiological, and serological examinations (every 6 months) did not show any evidence of local recurrence or systemic hydatidosis during the follow-up period of 50 months.

When the cysts do not show characteristic MRI features, a differential diagnosis is quite difficult and the lesion can imitate different cystic pathology. The management of spinal hydatid cyst is always surgery, but incorrect preoperative diagnosis of these lesions can compromise management of the lesion or lead to complications related to cyst rupture intraoperatively. The most common complication is a rupture of the cyst into the subarachnoid space which leads to widespread dissemination followed by severe inflammatory or anaphylactic response. However, vertebral lesions are usually invasive and cause neurological symptoms due to compression. Almost all patients complain of radicular pain and motor deficits, and up to one-half of patients present with paraparesis.

In summary, Hydatid cysts that lack the typical radiographic appearance may be mistaken for other lesions. In our case, the lesion mimicked acute bleeding. A mistaken diagnosis can lead to inappropriate management or complications related to intraoperative cyst rupture.

Acknowledgements
This study was supported by Department of Neurosurgery, 1st Teaching Hospital of Xinjiang Medical University. The authors declare that they have no competing interests.

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