Primary spinal epidural hydatid cyst with intrathoracic extension

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ABSTRACT

يعد وجود الكيس اليرقاني الشوكي خارج الجافية مرضاً نادر الوجود. نستعرض في هذا التقرير حالة مريض يلغ من العمر 33 عاماً، أصيب بإنفلاج نصف الجسم السفلي خلال عامين. أظهر الكشف بالأشعة المقطعية والرنين المغناطيسي وجود كيس متعدد الفصوص داخل القفص الصدري، والذي يحتمل أن يكون ذو أصل داخل النخاع الشوكي. مكنت الجراحة من إثبات الطابع اليرقاني للكيس، كما أُجريت إزالة كلية للضغط الواقع على النخاع الشوكي. لم تسجل أية إصابة عظمية. من خلال هذه الملاحظة، نتحدث عن أول حالة كيس يرقاني شوكي أصلي خارج الجافية تمت إدارته بنجاح عبر مقاربة جراحية أمامية، كما نناقش المظاهر الوبائية، طرق التشخيص، أساليب التدبير، وعلاج هذه الحالة النادرة.

Spinal epidural localization of hydatid cyst is quite rare. We report a case of a 33-year-old patient who experienced paraparesis over 2 years, with an umbilical sensitive level. A CT scan and MRI showed an intrathoracic multilobar lesion, probably of intra-spinal origin. An anterolateral transpleural surgical approach confirmed the hydatic character of the observed lesion and enabled total spinal cord decompression. No osseous involvement was noted. We report a case of spinal epidural hydatid cyst successfully managed by an anterior approach, and we discuss epidemiological, diagnosis, and therapeutical features of this rare localization of hydatid cyst.

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Hydatid disease of the spine occurs in 1% of hydatidosis; it is mostly located in the thoracic spine.[1-5] The disease either occurs by direct extension from a pulmonary infestation,[6] or begins less often primarily in the vertebral body.[2] Rarely, the disease starts at the extradural area.[6] Isolated epidural hydatid cysts are extremely rare. So far, only 20 or so cases have been previously reported in the literature.[6-10] Hence, the objective of this paper is to report a particular case of primary spinal epidural hydatid cyst with intrathoracic extension that was successfully managed using a thoracotomy approach.

Case Report. A 33-year-old woman with unremarkable medical history was hospitalized at the Neurosurgery Department of the University Hospital Hassan II, Fez, Morocco in January 2002. She reported a sensation of inferior limbs weakness and numbness, which occurred 2 months earlier, without any sphincteral disorders. The initial neurologic examination showed a severe spastic paraparesis prominent in the left lower limb, with umbilical sensitive level. The knee jerks were brisk bilaterally, and the ankle jerks were clonic; Babinski sign was present bilaterally. The chest x-ray showed regular paravertebral opacity close to the seventh thoracic vertebrae without any pulmonary damage (**Figure 1**). The CT scan showed a multilobar lesion within the left conjugate foramina of the seventh dorsal vertebrae. The MRI showed an extensive process with CSF-like signal intensity on T1- and T2-weighted images, emerging from the seventh and eighth thoracic vertebrae; the process had both extra and intra-spinal components compressing the spinal cord (**Figures 2a & 2b**). These radiological features suggest either a neurinoma or a meningocele. Biological assessment did not show any abnormalities and the abdominal ultrasound was normal. A left anterolateral surgical approach was performed and enabled the extraction of 2 cysts of various dimensions. During surgery, a left latero-pulmonary bilobed mass was noticed; this strongly adhered to the prevertebral aponeurosis and was associated with an enlargement of the conjugate foramina of the seventh thoracic vertebrae (**Figure 3**). The cyst wall was ruptured during

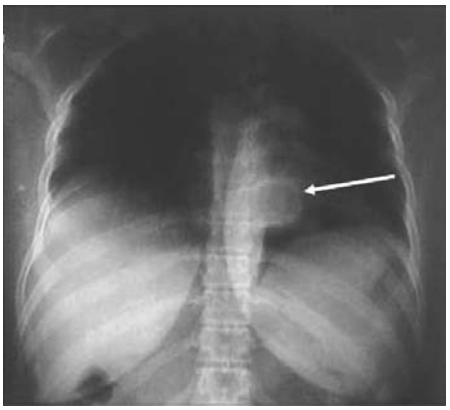

Figure 1 - A chest x-ray showing a left paravertebral homogeneous and sharp opacity (arrow).

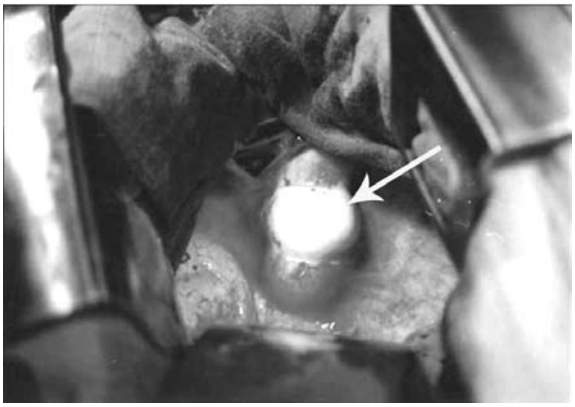

Figure 3 - Per-surgery picture showing a left latero-pulmonary multilobar mass adhering to the prevertebral aponeurosis.

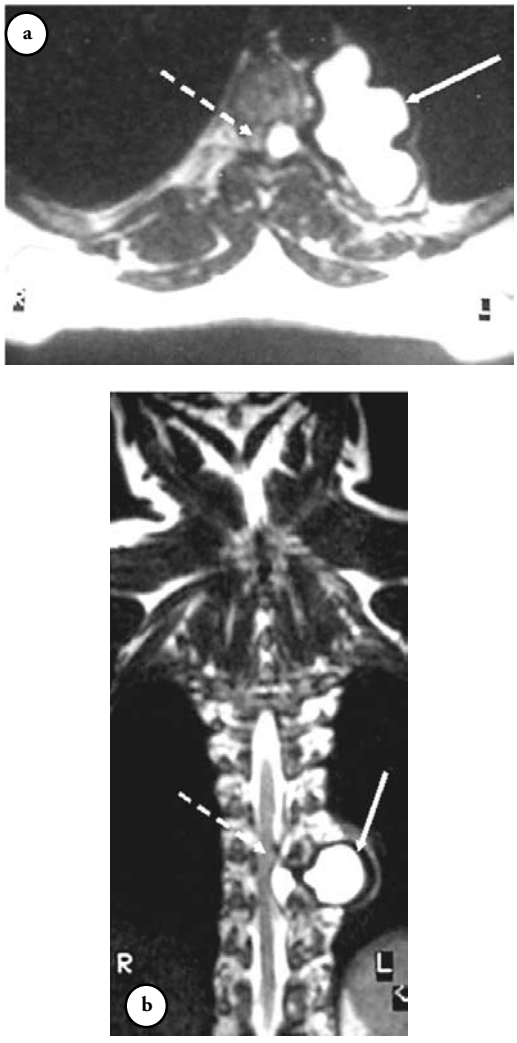

Figure 2 - T2-weighted MRI in axial **a)** and coronal **b)** slices showing a multilobar cystic mass (plain arrow) in the epidural space compressing the spinal cord (dashed arrow).

excision, and copious irrigation of the surgical field was performed using 10% hypertonic saline solution. The histopathology examination of the cyst wall confirmed the diagnosis of hydatid cyst, revealing an outer laminated non-nucleated layer and an inner germinative layer. The postoperative course was uneventful, and she underwent 400 mg albendazole therapy twice a day for one month alternated with 4 weeks treatment free interval for one year. At follow-up, MRI confirmed the complete excision of the hydatid cysts and demonstrated sufficient spinal cord decompression. At present, she remains very well without any evidence of recurrence of the parasite elsewhere in the spine.

Discussion. Hydatid disease or hydatidosis is the most widespread human cestode infection in the world. It is a serious parasitic infection usually due to infestation by the larval stage of the dog tapeworm, *Echinococcus granulosis*. The infestation occurs in humans as a result of feco-oral contamination, either by direct ingestion of parasitic eggs from contact with dogs or indirectly from contaminated water or food.[2] Primary vertebral hydatid disease without any other systemic involvement can be explained through the direct porto-vertebral venous shunt theory.[3] This theory supports the fact that the disease begins rarely from the extradural area.[3,6] The primary epidural location for the hydatid cyst is very rare.[9,11,12] In this location, the cyst may be single,[6,8,11] or multiple.[13] It is defined as the presence of an extradural hydatid vesicle without surrounding osseous lesion. Consequently, hydatid cysts originating from the vertebral bone and reaching the extradural space after destroying the bone are excluded. The extradural cyst may protrude laterally into the paraspinal gutter and then penetrate the fascia of the psoas muscle.[6] It may also protrude into the thoracic cavity, as occurred in our patient. There is a male preponderance of the disease,[9] and most patients affected are younger than 30 years.[10] It presents generally with symptoms of increasing

severity, progressing to a cauda equina syndrome or a slow spinal cord compression according to the location of the disease.[10] Correct preoperative diagnosis is very difficult. Serological tests for hydatid disease are often negative, and the disease has no pathognomonic signs; hence, the diagnosis is considered if the patient visited or lives in an endemic geographic region.[6] Mostly, the final diagnosis is established during the surgical procedure. Increasingly, MRI is becoming the gold standard for diagnosing spinal hydatid disease like many other types of spinal pathology. In fact, hydatid cysts have an almost unique appearance on MRI;[11] they are spherical and their walls are very thin and regular with no septa. On T1-weighted images, the cyst wall may be isointense or give a slightly lower signal than its contents; on T2-weighted images, it demonstrates a low intensity rim surrounding the homogenous high-signal cyst content.[11] In addition, MRI provides valuable information on the extent of the disease and delineates also the viability of the hydatid cysts.[6,14] According to the location and the extension of the hydatid cysts, Braithwaite and Lees,[15] have classified this disease into 5 radiological types: 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. Therefore, our patient belongs to the type 3 of this classification. Surgery is the treatment of choice for spinal cord compression by epidural hydatid cysts. Ideally, the treatment would be a radical excision of entire cysts without per-surgery rupture. All cases previously reported in the literature were managed via a posterior approach. In this paper, we report a case successfully managed using transpleural thoracotomy. The choice of this approach was based on the imaging data; this allowed a full exeresis of the hydatid cysts and satisfying spinal cord decompression. Many surgeons prefer to use scolecoidal agents as an efficient means of preventing dissemination of the parasite if accidental rupture of the cyst occurs during surgery.[5,10,16] The very weak adhesion of the cysts to both the dura and the bone is an important factor for benign surgical prognosis of the epidural hydatid disease. This allows successful and complete removal of cysts by simple maneuvers, such as irrigation and aspiration.[13] This feature contrasts the prognosis of spinal hydatidosis, which is commonly considered to be as hopeless as that of malignant disease of the spine. Furthermore, adjuvant medical treatment is mandatory in the management of the spinal hydatid disease. Albendazole is the preferred antihelminthic;[17] it is used typically in cycles of 3 months to one year and indefinitely if recurrence occurs.[17] There are recent articles reporting the effectiveness of solitary albendazole therapy in prolonging patient survival for inoperable cases.[18,19]

In conclusion, even if the serological tests are negative, hydatid cysts should be considered in the differential diagnosis of cystic lesions in the epidural space, especially in endemic countries. Radical surgery coupled with antihelminthic therapy seems to provide an efficient long-lasting cure. The surgical approach is dictated by the localization and extensions of the lesion. Compared to vertebral hydatidosis, primary spinal epidural hydatid cyst offers an exceptional possibility for a good prognosis.[8] Nevertheless, close follow-up should be observed according to the risk of recurrence.

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