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Case Report

Paraplegia Revealing Primary Extradural Spinal Hydatidosis

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Abstract

Spinal hydatidosis is exceptional: prevalence estimated at less than 1% of all localizations of the hydatid disease. Extradural forms of spinal hydatid cyst are rare and only a few sporadic cases have been reported in the medical literature. It is characterized by its high clinical latency, potential severity, and frequent recurrence. It can present itself as a real neurological emergency: spinal cord compression and cauda equina syndrome. It is thus recommended to evoke this diagnosis in front of any symptomatology of non-traumatic spinal cord compression in endemic areas for this parasitosis.

We report an original observation of acute paraplegia revealing a primary extradural hydatid cyst of the dorsal cord in a young Tunisian man.

Introduction

Echinococcosis is an anthropozoonosis that is still epidemic in several countries of the world, particularly in rural areas where there is an intimate contact between humans and animals (mainly sheep and dogs) [1].

The main localizations of this parasitosis are the liver and the lungs: respectively 65-70% and 25% of the cases [1,2].

More rarely, other unusual locations [3] described as "ectopic" and "aberrant" [4] can be noted and represent real diagnostic challenges for clinicians [5-10].

Hydatid disease of the central nervous system (CNS) remains rare compared to other unusual locations; it accounts for only about 2% of all human echinococcosis [11]. It affects the brain in 2/3 of the cases and the marrow in 1/3 of cases [12]. The prevalence of spinal hydatidosis is thus estimated at less than 1% of all localizations of hydatid disease [13]. Extradural forms of spinal hydatid cyst (HC) are rare and only a few sporadic cases have been reported in the medical literature.

We report an original observation of acute paraplegia revealing a primary extradural HC of the dorsal cord in a young Tunisian man.

Case Report

A 23-year-old man, with no particular pathological history, presented to the emergency department for lower limb weakness evolving for 3 months with progressive installation and worsening. The walk became impossible the morning of his consultation at the emergency room. The examination showed paraplegia with hyperreflexia of the lower limbs, and a sensory level in T4. The patient was apyretic, eupnetic, conscious, oriented, and with stable hemodynamic state and vital constants.

Basic biological tests (total blood count, postprandial glucose, C-reactive protein, erythrocyte sedimentation rate, ionogram, calcium, magnesium, muscle enzymes, lipid parameters, transaminases, thyroid hormones, and electrophoresis of serum proteins), the electrocardiogram, and the chest x-ray were without significant abnormalities.

Medullary magnetic resonance imaging (MRI) revealed a cystic, extradural, multi-vesicular lesion, extended from T5 to T8, and compressing the medullary cord. The lesion was hypointense on T1-weighted images, hyperintense on T2-weighted images, and with no enhancement after gadolinium injection (Figs 1, 2 and 3). Thoraco-abdominopelvic CT did not note other associated visceral lesions.

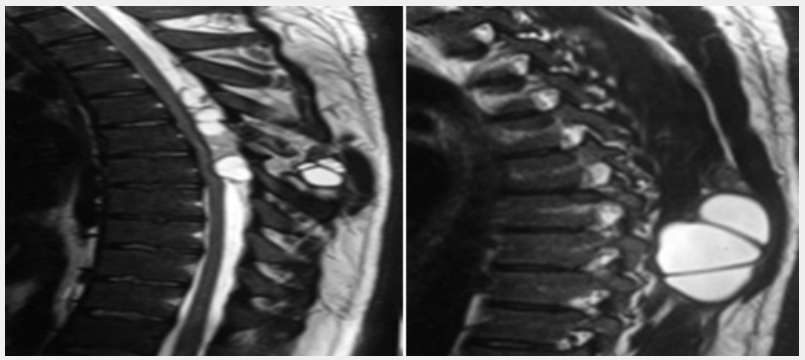

Figure 1: *T2-weighted sagittal spine MRI: extradural, multi-vesicular cystic mass, extending from T5 to T8, and with spontaneous hypersignal*

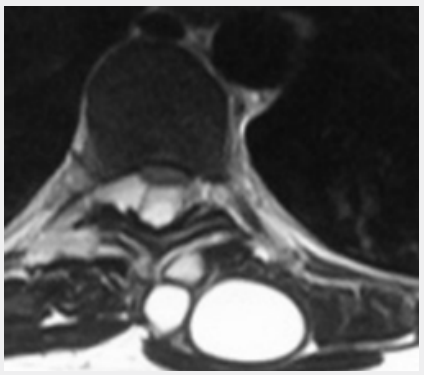

Figure 2: *T2-weighted axial spinal MRI: extradural, multi-vesicular cystic lesion, and with spontaneous hypersignal*

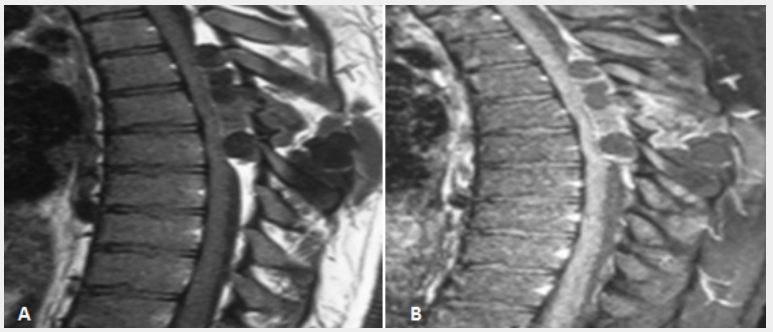

Figure 3: *T1-weighted sagittal spine MRI without (a) and with (b) gadolinium injection: extradural, multi-vesicular mass, extending from T5 to T8, hypointense, and without secondary enhancement*

A T5-T8 laminectomy was performed showing an extradural cyst with a colorless content suggestive of a HC. The cyst was removed completely (total cystectomy).

Histopathological examination confirmed the diagnosis of extradural spinal HC. The hydatid serology was negative and subsequent investigations did not reveal other hydatid locations confirming the primary character of the spinal HC. Postoperatively, the patient received oral albendazole at a dose of 800mg/day for 4 months.

Progressive improvement was noted with resumption of walking and muscle strength. The neurological examination was strictly normal after six months of surgery. No recurrence was noted during a two-year postoperative follow-up.

Discussion

The thoracic spine represents the preferred seat of spinal hydatidosis (52%), followed by the lumbar spinal (37%), then cervical and sacral levels [14,15].

Clinically, the majority of patients presented with back pain and/or lower limb weakness. More rarely sphincter disorders or even a cauda equina syndrome [11-15]. Apart from this symptomatology due to spinal cord compression, spinal hydatid disease has no specific manifestations [11-14].

In endemic countries, spinal HC is among the medical pathologies frequently responsible for medullary compression: 14% of spinal cord compressions were of hydatic origin in Bettaieb A *et al* series [16]. It is therefore recommended to discuss this diagnosis in front of symptoms of non-traumatic spinal cord compression in endemic areas for echinococcosis [16].

Biologically, eosinophilia is found only in less than 15% of cases and is particularly important in cases of cyst rupture [11-14]. The hydatid serology is also not very contributive to the diagnosis of the spinal localization of HC; it is only positive in about 25% of cases [17]. It is particularly useful for post-operative surveillance to detect recurrences [12,18,19].

MRI is the exam of choice for the positive diagnosis of spinal hydatidosis [20,21], and the radiological classification proposed by Braithwaite and Lees in 1981 distinguishes 5 types of spinal HC: Primary cyst in the cord, Intradural cyst, Extradural cyst, Hydatid disease of the vertebrae, and Paravertebral hydatid disease [22].

The first three types are considered rare [19]; the primary intramedullary type remains exceptional with only a few sporadic cases reported in the literature [13,22-24].

The treatment of choice for spinal HC is surgical [25]. Adjuvant chemotherapy with Albendazol perioperatively is classically recommended to inactivate the cyst preoperatively, and reduce postoperative recurrence [26]. Recurrences are frequent for this localization: 30 to 100% of the cases according to the series [14], and this could be explained by the difficulty of complete excision of the cyst and its sometimes unavoidable rupture, given the narrowness of the operative field and the close relationships with adjacent structures [14,25,26].

Conclusion

The primitive spinal localization of human echinococcosis remains exceptional and unusual. It represents a real diagnostic challenge for clinicians given its serious neurological complications and often urgent presentations.

A better knowledge of this location is the only guarantee of appropriate and urgent care to improve the prognosis often reserved.

It is thus recommended to evoke the diagnosis of spinal HC in front of any spinal symptomatology or spinal cord compression that remains unexplained in endemic areas for this parasitosis.

Conflict of Interest: None

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