Case Report **Open Access**

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Uncommon primary hydatid cyst occupying the adrenal gland space, treated with laparoscopic surgical approach in an old patient

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Abstract: Hydatid disease (HD) is caused by Echinococcus Granulosus (EG), which is a larva endemic in many undeveloped areas. The most common target is the liver (59%–75%). The retroperitoneal space is considered as a rare localization. We report an uncommon case of HD located in the adrenal gland space.

Presentation of case. This is a 78-year-old Moroccan woman, with right flank pain for eight months previously. She denied contact with dogs or sheep. Her physical examination was normal. There was no pathological alteration of laboratory exams. CT scan measuring 5 cm without clear signs for a sure diagnosis found a round lesion in the right adrenal gland. An abdominal MRI showed a round mass of 34 x 27 mm with fluid component without a clear plane of dissection from kidney and liver. A laparoscopic procedure was performed to obtain a histological diagnosis. We reached a conclusive diagnosis of Hydatid cyst of right adrenal gland space.

Hydatid cysts often develop in the liver. The location in the adrenal bed is rare without clinical signs related to alteration of the gland's secretion. Hydatid cyst identification in the adrenal gland space is based on ultrasonography, CT or MRI scans. The differential diagnosis includes various benign and malignant lesions. Laparoscopic procedure is the best approach available to obtain a histological diagnosis and a curative treatment.

The best treatment for HD is the pericystectomy. Laparoscopic surgery can guarantee a radical resection of these lesions when it performed by an expert surgeon.

Keywords: Hydatid disease; Adrenal Gland; Laparoscopic Pericystectomy; MRI Cystic lesions; Laparoscopic Adrenalectomy

Highlights

- HD may occur in an uncommon organ or tissue
- HD diagnosis is sometimes difficult to gain
- Radical surgery is the best treatment in HD disease
- Laparoscopic surgery can also be possible in HD

1 Introduction

Hydatid disease (HD) is caused by Echinococcus Granulosus (EG), which is a larva endemic in many undeveloped areas such as the Middle East, Africa, Latin America, etc. It has maximum diffusion among people in poorer socioeconomic status so it poses a great problem to public health in countries where EG is endemic. Although HD is a relatively rare disease, it has a big impact on patient health because it may occur in any organ or tissue. The most common and preferential target organ is the liver (59%-to 75%), followed in frequency by lung (27%), bone (14%) and brain (12%) almost at the same level and finally the kidney (3%) [2- 6]. Other sites such as spleen, pancreas, heart, and adrenal gland are very rare, despite this, in the international literature; there are some reports about these localizations.

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The retroperitoneal space occupied by the adrenal gland is considered as a rare localization for a hydatid cyst, indeed the reported frequency for a hydatid cyst invading the adrenal gland is 3%[3], although the real incidence for primary localization in this site is even more rare; we report an uncommon case of HD localized in the retroperitoneal space normally occupied by the adrenal gland without any clinical and histological signs of adrenal gland encroachment.

2 Case presentation

A 78-years old Moroccan woman was admitted to our department for a right flank pain without any other relevant symptoms or signs; she referred to only a light pain radiating to the epigastric region. The pain had started eight or ten months previously, she had no nausea or vomiting, she was afebrile and her vital signs were normal. She denied any contact with dogs or sheep and she had moved from Morocco to Italy in 2009. She was unemployed. Her physical examination was normal, without any significant signs or symptoms, which could suggest retroperitoneal abdominal masses. When she was a child she had the common infective diseases and at the moment of physical examination she had only an iron deficiency anemia. She underwent two Caesarean Births in 2009 and 2012. We performed routine laboratory exams on her, such as hemocromocitometric panel, major biochemical values and principal enzymes; there was no pathological alteration. She underwent ultrasound abdominal exam so we found a strange ovoidal hypoechoic formation in the right adrenal gland space; it was four centimetres in maximum diameter, and there were no signs of peripheral or central vascularization at Doppler's exam; it was impossible to establish a sure diagnosis so we proposed a CT scan of the abdomen. The CT scan showed a solid mass of maximum diameter 5 cm, with a light peripheral vascularization and degenerative cystic areas; it had no clear linkage with either liver or with right adrenal gland. Therefore we decided to admit her to perform other imaging and laboratory exams; in fact we required an abdominal MRI which showed: "mass of 34 x 27 mm with a central cyst composed of a multilocular structure, there are signs of parietal vascularization and there aren't signs of a clear dissection plane from kidney and liver"(image 1,2,3,4). Despite this we continued with a screening exam for HD such as indirect hemaglutination test and also hormonal exams both for adrenal cortex hormones and medullar catecholamines; all these exams were negative. We decided to perform a surgical approach to this mass, because we had to have a sure histological diagnosis of the mass; we gained the informed consent for surgical procedure. We set up the patient on the operating table in lateral left decubitus, we obtained a pneumoperitoneum with open technique and after an accurate exploration of abdominal cavity placed another three trocars to complete the procedure. We gained access to the retroperitoneal space with the same surgical steps of a laparoscopic adrenalectomy. There were significant adhesions between the unknown mass and the liver, kidney and adrenal gland. The mass was removed after a dissection through an unclear plane between liver and lesion. We extracted it into a laparoscopic bag from the access in right flank (image 5). The postoperative course was regular, without relevant complications. Unfortunately the definitive histological report was unclear. There was a significant rise in IG indirect hemagglutination one month after the surgical procedure (Indirect Haemoagglutination Assay was 1:600). Therefore we called a specialist for expert advice and finally we reached a conclusive diagnosis: Hydatid cyst localized in the retroperitoneal right adrenal gland space. Now the patient is in good health; even so, we have prescribed her a prophylactic drug treatment with albendazole.

Consent. Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available from review by the Editor-in-Chief of this journal.

Ethical approval: The research related to human use has been complied with all the relevant national regulations, institutional policies and in accordance the tenets of the Helsinki Declaration, and has been approved by the authors' institutional review board or equivalent committee.

3 Discussion

Traditionally HD has two clinical categories: cystic hydatidosis-caused Echinococcus granulosus and alveolar hydatidosis, caused by Echinococcus multilocularis [1]. Infection begins with the ingestion of tapeworm eggs, which in the human intestine hatch into embryos that penetrate the small bowel mucosa and travel to the liver. Hydatid cysts most often develop in the liver. However, when embryos jump through this first filter,

they reach the second most frequent location, the lung, even though hydatid cysts occur anywhere in the body [2]. The symptoms and clinical signs depend on the structures and organs of the body affected by the pathogens. In the adrenal affection the major clinical signs could be hypertension, pain, hormone disorders and other manifestations because there are many substances produced by this gland. However there have been 20 cases of primary hydatid cysts of the adrenal glands reported up to 2007 [3] and it is impossible to learn much about so rare a disease; in addition to this our case is the most rare of the above-mentioned cases because it is an isolated cystic mass without significant involvement of the adrenal gland. Imaging diagnostic problems linked with so rare a localization of the mass as well as an unspecific alteration in the hemagglutination indirect exam, suggested treating this lesion directly with a radical surgery. The imaging about HD is current variable; indeed it depends on the stage of evolution of the disease. Early lesions appear purely cystic; after modification of the germinal layer and a reduction of intraluminal pressure the capsule becomes fibrotic and sometimes calcified, while the daughter cysts discharge from the wall and float in the lumen. Hydatid cyst identification in the adrenal gland space is based mainly on ultrasonography and CT scans [4-6,14-17]; they reach a sensitivity range from 93% to 98% for Ultrasonography while CT scans reach around 97%. On radiology hydatid cysts can be described according to the classification of Gharbi et al. [7] which is based on ultrasonography features and includes five types. In our case the Cyst was a Type IV sec. Gharbi classification described as nonspecific solid mass with unclear hypoechoic pattern. The undefined diagnosis forced us to require additional exams, indeed we underwent the patient further to MRI. Few reports of MRI of adrenal HD have been published. On MRI the complex cyst contents can be well displayed and the cyst membrane, whether collapsed or not, can be clearly seen as a low-intensity curvilinear structure on both short and long repetition time spin-echo images [11], unfortunately in our case these characteristic were not in the MRI exam [14]. The differential diagnosis of an adrenal cyst should include an endothelial cyst, a pseudocyst due to infarction or hemorrhage in the adrenal gland, cystic neoplasm, ectopic glandular tissue and a post traumatic cyst [8-10,12,18-20]. For the above reasons treatment of undefined cyst in the retroperitoneal adrenal gland space is mostly surgical [11]. Pericystectomy or resection of the entire adrenal gland is the two preferred choices. Resection of the cyst with the conservation of the gland remains the optimal procedure [13]. It is preferred to follow an appropriate dissection plane between the cyst and the adrenal gland in order for a complete cystectomy to be achieved. However, it is not always possible and may cause hemorrhage because of adherence of the cystic wall to the adrenal gland's parenchyma. In the case of hemorrhage or failure to perform the cystectomy, ablation of the entire adrenal gland including the cyst should be performed. Partial dissection should be avoided because of a higher risk of dissemination or recurrence and, in rare cases, anaphylactic shock. As shown by the definitive histological report, we reached a complete eradication of the lesion, without damage to the adrenal gland. Laparoscopic access to adrenal gland space could be performed either anterior trans-abdominal and trans-peritoneal access or posterior and retroperitoneal access. We decided for a laparoscopic trans-abdominal and trans-peritoneal access [13]; indeed we think this approach guarantees a good dominance of the surgical field and at the same time it permits ergonomic movements to the first operator; we think that in an expert surgeon's hands this is the best approach available for elder patients [21-24].

4 Conclusion

In every country, due to migrant flows and intercontinental touristic journey, HD has to be considered the most probable diagnosis when there is an uncommon cystic or semi-cystic lesion even if there are no laboratory markers which confirm this suspect.

The best treatment for these cystic lesions still remains pericystectomy.

Laparoscopic surgery can guarantee a radical surgery for these lesions, when an expert surgeon in this field performs it.

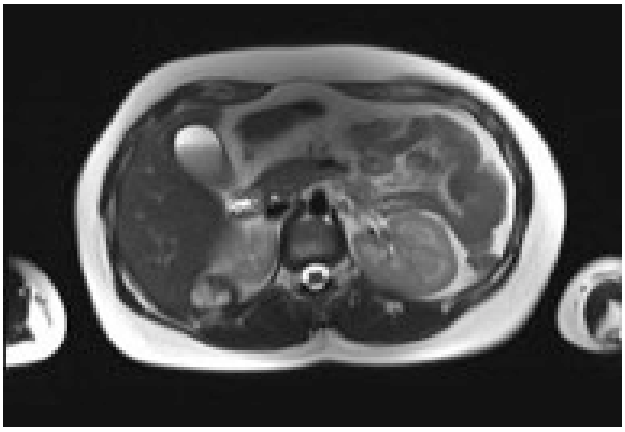

Figure 1: Rm scan T1weighted axial image showing the adrenal mass

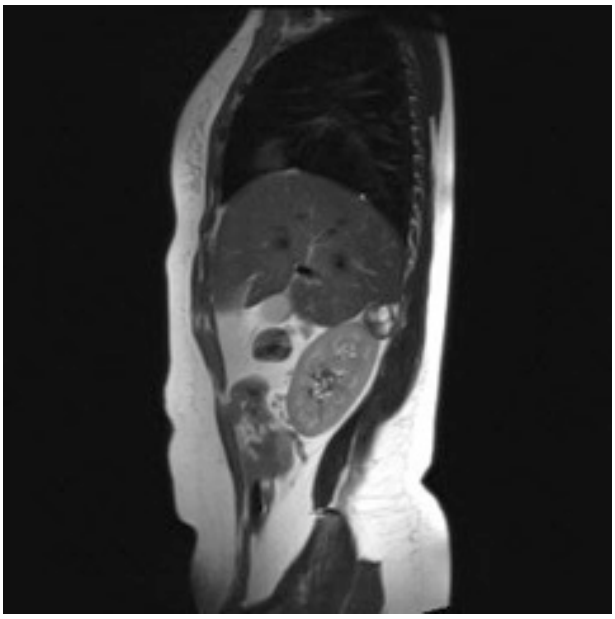

Figure 2: Rm scan T1weighted sagittal image showing the adrenal mass

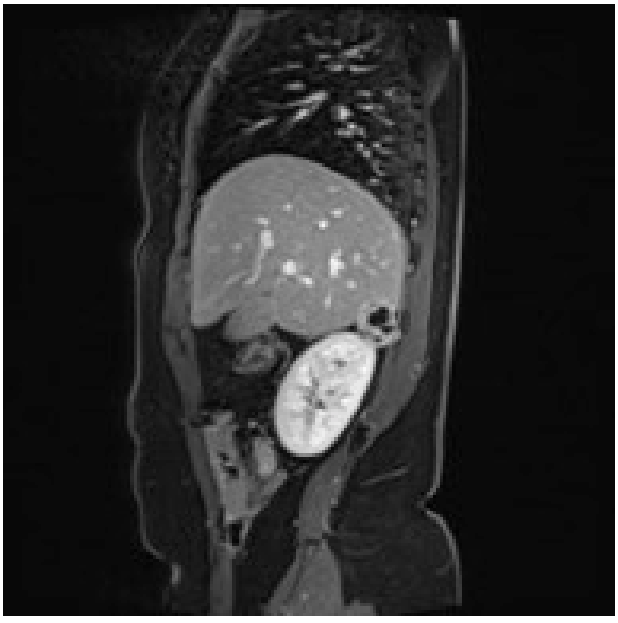

Figure 3: Rm scan T2weighted sagittal image showing the adrenal mass

Conflict of interest: The authors declare no conflict of interest.

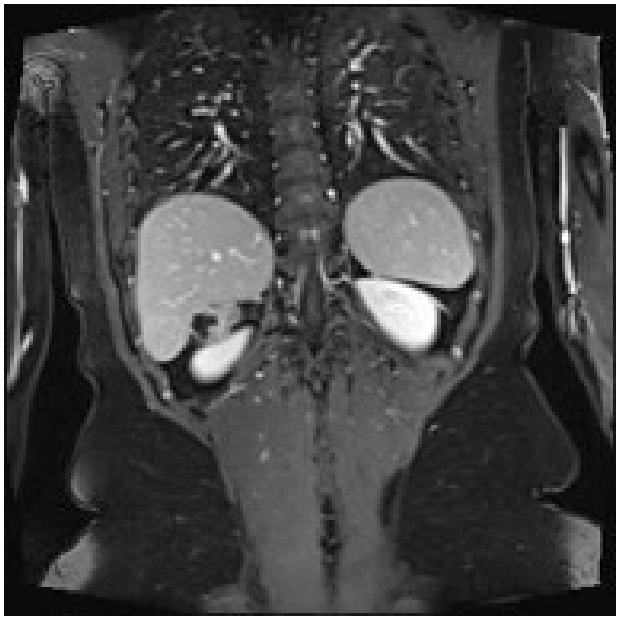

Figure 4: Rm scan T2 weighted coronal image showing the adrenal mass

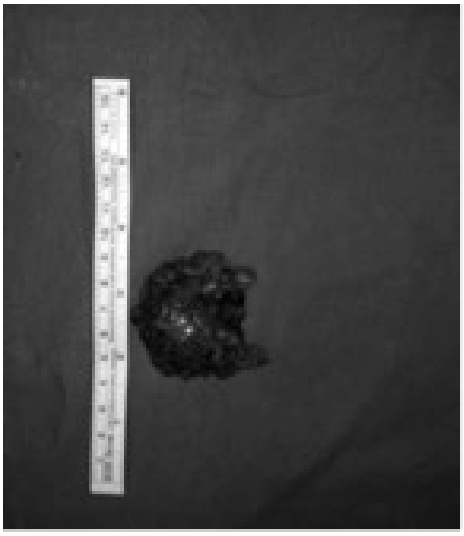

Figure 5: Specimen

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