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Retroperitoneal Mesothelial Cyst: An Incidental Finding

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1. Abstract

1.1. Introduction: Intrabdominal masses can be solid or cystic. Depending on their location, they can be classified as intrabdominal, retroperitoneal, pelvic or dependent of the abdominal wall. Imaging studies are crucial to characterize the lesions and their locations. Abdominal ultrasound allows the description of the lesion, making it possible to differentiate solid masses from cysts, however a CAT scan or an MRI (with contrast) is necessary to better describe them and obtain more information on the location, content and anatomical relationships. Many patients are scheduled for surgery with an erroneous diagnosis. The definitive diagnosis is given by direct anatomopathological study.

1.2. Case Description: We present the case of a young female patient, in whom a large intraabdominal cyst apparently depending of the liver was discovered incidentally as part of workup for an unrelated medical problem. We discuss the diagnostic challenges and differential diagnosis, as well as the therapeutic management. Our patient was initially scheduled for hepatic resection. During the operation, it was established that the lesion was not hepatic but retroperitoneal in origin. Only after the anatomopathological study, it was defined as a benign mesothelial cyst.

1.3. Conclusion: Retroperitoneal cysts are rare lesions, with non-specific associated symptoms, making the diagnostic process difficult. The anatomopathological analysis is necessary to define the nature of the cyst. Imaging tests provide approximate location information and anatomical relationships with the rest of the organs. Many patients are scheduled for resection with erroneous diagnosis, as in our case, and it is during the operation when the type of tissue and location is defined. The anatomopathological diagnosis is important to determine the evolution and prognosis. Up

to date literature about these lesions is scarce and dissemination of case descriptions like ours may help other clinicians facing this challenging diagnosis in individual patients.

2. Introduction

Intraabdominal masses can be solid or cystic. Depending on their location, they can be classified as intraabdominal, retroperitoneal, pelvic or dependent on the abdominal wall [1]. Imaging studies are crucial to characterize the lesions and their location. Abdominal ultrasound allows the description of the lesion, making it possible to differentiate solid masses from cysts, however a CAT scan or an MRI (with contrast) is necessary to better describe them and obtain more information on the location, content and anatomical relationships. The definitive diagnosis is given by direct anatomopathological study [2]. These lesions have low incidence. They may be diagnosed in childhood or at an older age and their presentation is usually late, when they have reached considerable size before becoming symptomatic (abdominal pain, palpable mass, etc.). In many cases their diagnosis is incidental [3].

We present the case of a young female patient, in whom a large intraabdominal cyst apparently depending of the liver was discovered incidentally as part of workup for an unrelated medical problem. We discuss the diagnostic challenges and differential diagnosis, as well as the therapeutic management.

3. Case Description

A nulliparous 29 year old, otherwise healthy female, presented with Raynaud's phenomenon in both hands. The rheumatologist requested a complete blood test and a chest X-ray as part of the workup. The results came back as normal, except for mild iron deficiency anemia (Hb 11,2 g/dL, Hct 33% and Fe 21ug/dL) and a right upper quadrant abdominal mass with calcified borders on

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the chest X-ray (Figure 1). The patient was reevaluated, referring no relevant symptoms. She had no apparent blood loss, other than heavy menses and the physical examination of the abdomen was unremarkable. An abdominal ultrasound was indicated and revealed an unilocular, well defined cystic lesion 6x5cm with echogenic images inside which changed depending on the patient's body position. The cyst was described as located in the liver, however a relationship to the right kidney could not be excluded (Figure 2). Due to the patient's childbearing age, an MRI with contrast was chosen to further investigate the lesion, rather than a CAT scan which would subject her to greater radiation. The result was a single chamber cystic lesion which appeared dependent of

segment VI of the liver, with calcified wall, no septa, solid areas nor papillary structures. The lesion was slightly hyperintense on the T1 weighted sequence. There was no contrast diffusion restriction (Figure 3).

Although the images were not suggestive of a hydatid cyst, it was considered as a differential diagnosis because of its high prevalence in the local population. A new blood test was performed including Echinococcus granulosus, Hep B, HIV, serology and tumor markers (CEA, Ca 125, Ca 19,9, Ca 15,3, AFP). The only positive result was a slight elevation of Ca 125 (38U/ml, cut off value 35). A transvaginal ultrasound was also performed and was normal.



Figure 1: Chest X-ray

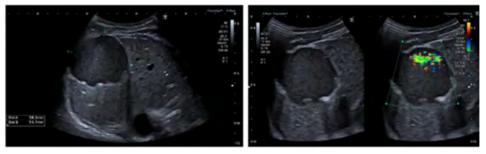


Figure 2: Abdominal ultrasound

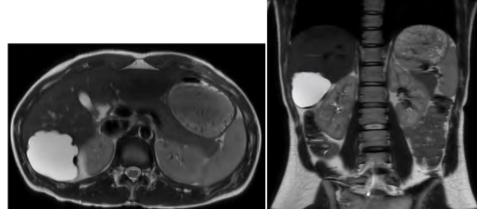


Figure 3: Contrast MRI image of the lesion

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Because of the diagnostic difficulty, the case was presented to radiologists, oncologists, GIT physicians and general surgeons in a joint clinical round. After considering the different management options, complete laparoscopic resection and subsequent anatomopathological analysis was decided as the treatment of choice. Preoperative lesion biopsy was discarded because of poor diagnostic utility in cystic lesions and the possibility of content spillage inside the abdominal cavity.

The patient was scheduled for laparoscopic hepatic resection. Since such operations have a high risk of bleeding, erythrocyte mass optimization was necessary. The patient was included in the Patient Blood Management program and received 2 doses of 500mg of Iron Carboxymaltose (Ferinject ®) intravenously at weekly intervals as outpatient. Two weeks later, the preoperative blood sample revealed Hb 12,7g/dL.

After anesthetic induction, two large caliber intravenous lines were secured to permit rapid fluid infusion if it became necessary, and the left radial artery was cannulated for invasive hemodynamic monitoring and frequent Hb monitoring. As soon as the laparoscopic ports (four) were established, the surgeons gently explored the lesion and determined that it did not depend of the liver but was retroperitoneal in origin. It was tightly adherent to the lateral aspect of the diaphragm. A radical cystectomy was performed and the whole lesion (7,5x6,3x5,7cm) was removed via a Pfannenstiel incision. During the course of resection, the right parietal pleura was incidentally opened without respiratory or hemodynamic repercussions. It was closed with V-Loc 2/0 and respirator assisted Valsalva maneuver pneumothorax resorption. Otherwise the procedure was uneventful and the patient was extubated in the operating theatre and transferred to the post anesthetic recovery unit.

24 hours later, a control chest X-ray was performed, revealing a small right apical pneumothorax, a minimal flap of right subdiaphragmatic pneumoperitoneum and an air-fluid level in the right upper quadrant. Pain was well controlled with IV NSAIDs and Acetaminophen, without the need for opiates. At 72 hours postop, blood tests were performed (Hb 10.7 g/dL, Hct 31.8%). A new dose of intravenous Iron Carboxymaltose 500 mg was administered prior to discharge. The same dose was repeated a week later as outpatient. The postoperative course was uneventful. The anatomopathology analysis of the lesion revealed a benign mesothelial cystic formation partially covered by muscle and fibroadipose tissue measuring 7.5x6.3x5.7 cm. When disected, it presented seronecrotic content and a smooth whitish internal surface with yellowish-orange and calcified areas which are included in B1 after decalcification in nitric oxide.

The patient had sudden onset of meralgia paresthetica in the right thigh, two weeks after the surgical procedure. It was attributed to body posture at rest, inflammation and edema of the abdominal wall. She was taking oral NSAIDs with good pain relief. At six weeks review, she referred clinical improvement. Blood test at eight weeks reflected Hb 12,3 g/dL and Hct 35 %. She was discharged with appointment for repeat MRI with contrast at one year postop.

4. Discussion

Retroperitoneal cysts are rare lesions (1 in 250.000), mostly benign, although malignant masses with an unfavorable prognosis or turning malignant have also been described [3]. In 2000, Perrot classified these lesions based on histopathology (epithelial or mesenchymal), and their origin (pleural, pericardial, peritoneal, genitourinary, enteric, mesothelial, lymphatic, mature cystic teratoma), or nonpancreatic pseudocysts [4]. Due to their retroperitoneal location, the cysts reach large sizes before becoming symptomatic, so diagnosis is usually late, when the lesions reach up to 20 cm in diameter in 50% of patients [5]. The symptoms are non-specific: palpation of an abdominal mass, pain, abdominal distension, weight loss, or symptoms of complications of the cyst such as bleeding, infection, rupture or degeneration [6]. In many cases they are diagnosed as an incidental finding. Imaging tests are essential for the correct characterization of the lesion and describe the anatomical relationship, however, they are not specific. The first test to perform is usually an abdominal ultrasound. An anechoic lesion with posterior reinforcement is usually observed. It can be unilocular or multilocular, can present echogenic images, septa and calcifications. It is necessary to perform a CAT scan or an MRI with contrast, to provide more precise information about the size, content, anatomical relationship with the surrounding organs and to help plan the surgical approach [2]. The treatment of choice is surgical: complete resection of the lesion to reduce the recurrence rate, which is usually high (around 50%), although it does not usually turn malignant. Laparoscopic approach is preferred, but it must be performed by highly trained surgeons with advanced laparoscopic techniques [6,7]. Cases of aspiration and marsupialization of the lesions as first option, have been described, although they have high rates of recurrence and infection. Sometimes, intestinal or other organ resection is needed, to achieve complete cyst removal. In cases in which total resection is not possible, partial resection and marsupialization of the cyst have to be done [8]. There are some cases where instead of surgical resection, the lesion was punctured and subsequently sclerosing substances were injected. Even though malignant mesothelioma is associated with asbestos exposure, the benign type is not [9]. Its pathogenesis is not well known. It may be hormone dependent because of its higher prevalence in women. It also appears more frequently in women with a history of abdominal surgery, endometriosis or pelvic inflammatory disease [10]. It is usually diagnosed at 40-50 years of age. The prognosis of these lesions is favorable, although with high recurrence rates.

5. Conclusion

Retroperitoneal cysts are rare lesions, with non-specific associated symptoms, making the diagnostic process difficult. The anatomoajsccr.org 4

pathological analysis is necessary to define the nature of the cyst. Imaging tests provide approximate location information and anatomical relationship with the rest of the organs. Many patients are scheduled for resection with erroneous diagnosis, as in our case, and it is during the operation when the type of tissue and location is defined. The precise anatomopathological diagnosis is important to determine the evolution and prognosis. Up to date literature about these lesions is scarce and dissemination of case descriptions like ours may help other clinicians facing this challenging diagnosis in individual patients.

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