IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 17, Issue 7 Ver. 16 (July. 2018), PP 63-66
www.iosrjournals.org

Large cystadenoma of the liver - diagnostic and therapeutic approach

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Abstract: *Hepatic cystadenoma is a rare tumor, preferentially observed in women after forty years. The lesion is usually voluminous and made of a cystic cavity. It is characterized by its tendency to recur after surgery and its risk of malignant transformation into cystadenocarcinoma. The great contribution of imaging and especially the careful analysis of radiological aspects is very helpful. Only the histopathological examination can decide on the benign or malignant nature of the lesion. Treatment consists of surgical excision of the tumor. We report the case of a hepatic cystadenoma in a woman in her fifties and we discuss the diagnostic and therapeutic aspects of this type of lesion that justifies radical treatment, even in the absence of an argument in favor of malignancy.*

Keywords*: liver, cystadenoma, recurrence, imaging approach, surgical resection*

Date of Submission: 20-07-2018 Date of acceptance: 04-08-2018

I. Background / Introduction

Hepatic cystadenoma are rare multilocular cystic tumors of the liver. They are resulting from the biliary epithelium and are mostly located in the right lobe of the liver. They develop from either an aberrant bile duct or directly from a primitive hepatobiliary stem cell [1, 2].They account for less than 5% of nonparasitic cysts of the liver and happen commonly in women at middle-age. The size varies from 1.5 to 35 cm [2, 3]. Majority are intrahepatic (85%) [3, 4, 5, 6], fewer are extra hepatic [4–7, 8] and occasionally are seen to arise from the gall bladder [5, 7].In 1892, Keen reported the first case of hepatic cystadenoma, which now accounts for 5% of all cystic lesions of the live, less than 200 cases have been reported till dater.[1]

Hepatic cyst adenomas are benign tumors, but they have a high rate of recurrence and a probable for neoplastic transformation in approximately 10% of cases. Resecting these types of tumors and making an appropriate diagnosis is imperative, comparatively than observing them as is suitable for other common benign hepatic tumors, such as hemangioma, adenoma , and focal nodular hyperplasia (FNH).If not well resected they will have a very high rate of recurrence (>90%) [12].

II. Case presentation

A 52-year-old woman, operated three years ago for polycystic liver disease associated with gallbladder stones. A puncture-fenestration of the cystic tumor and a cholecystectomy by medial laparotomy were performed. She passed a smooth post-operative period. Four years ago the patient worried about the appearance of abdominal swelling with pain localized at the right upper quadrant of the abdomen. She was followed and treated for asthmatic disease. She had has no history jaundice or fever. On abdominal examination it has found a mass in the right upper quadrant not well limited with elastic consistency mobile with respiratory movement. There is no hepatomegaly. Respiratory and cardiovascular examinations were within normal limits. Biological investigationshad revealed an elevated ASAT and ALAT. There is no hyper leukocytosis. Tumor markers were within normal values. Hydatid serology was negative. Tumors marker were within normal values. Ultrasound examination showed an anchogenic mass of 20cm of diameter with fluid content and posterior reinforcement. It was partitioned evoking a hydatid cyst of the liver (stage III of Gharbi's classification)CT scan and MRI showed a multiple right lobe cystic masses coalescent; whose morphological aspect favors a benign origin: cystadenoma, without signs of loco regional infiltration (Figures 1-2-3).Absence of obvious communication with the biliary tree (Figures 4-5) – There is an arterial anatomical variant with left hepatic artery type that arises from the left gastric artery. Patient underwent total surgical resection of the cysts (Figure 6) . She passed a smooth post-operative period. The histopathological result was in favor of cyst adenoma of the liver (Figure 7).

III. Discussion

Biliary cystadenomas are rare and constitute fewer than 5% of cystic lesions of the liver. Naturally, theyoccur in a middle-aged woman presenting with abdominal pain and/or discomfort, associated with abdominal distension and sometimes palpable mass [1, 3, 4, 15]. Acute appearance is often pain due to intracystic hemorrhage or rupture of the cyst and associated with fever in case of infection of the cyst [15]. Jaundice [17–18]can appears when there is an extrinsic compression of the bile duct [19], biliary obstruction by an intraluminal tumor, or deposit of mucus secretion from a connectingcystadenoma [20]. Ascitesis present in case ofcompression of the inferior vena cava or the hepatic veins [18, 21, 22]. Cystadenomas are well-known to increase height during pregnancy and after oral contraceptives suggesting hormonal dependency [1, 9, 16]. Reappearance of a cyst following partial resection should increase a suspicion of cystadenoma. Hepatic cystadenomas are well-thought-out in the differential diagnosis of other hepatic cystic lesions, as well as simple cysts, echinococcal (hydatid) cysts, and cystadenocarcinomas.[23, 24]. Intracystic hemorrhage, mural nodularity or septations, can be present both in cystadenomas and in other cystic lesions of the liver. Less usually, cystadenomas may be confused with necrotic neoplasms, abscesses, cystic metastases, cystic hamartomas, embryonal sarcomas, hematomas, or other congenital cysts. Diagnostic questions also may arise in patients with Caroli disease or other forms of polycystic liver disease. Several bile duct hamartomas (von Meyenburg complex) can also simulate biliary cystadenoma, both for appearance and for imaging [25]. Radiologic studies, such as ultrasound examination and CT scan, may make known cystadenoma. Occasionally imaging reveals internal septations or papillary infoldings of the cyst itself. A significant solid component in the cyst suggests malignancy. Biliary cystadenoma is considered a premalignant condition, and only microscopic examinations can dependably differentiate it from its malignant homologcystadenocarcinoma. The demonstration of benign epithelium in common of the cystadenocarcinomas makes it extremely credible that they arose in previously benign cystadenomas. The occurrence of benign epithelium in most cystadenocarcinomas supports their origin from cystadenoma [8]CT scan and MRI commonly fails to distinguish the narrow communication [9] which is simply demonstratable during an intraoperative cholangiogram [28].Imaging studies are the key element of the diagnosis. UltraSound (USE) is more sensitive in finding internal septations, whereas CT scan provides anatomical relation to the liver. Hepatic cystadenomas appear on USE as anechoic lesions with internal septations. Focal hyperechoic areas within the lesion are common and can represent focal wall fibrosis, intracystic hemorrhage, or papillary projections. On conventional USE, cystadenomas tend more often to be multilocular than do cystadenocarcinomas, while cystadenocarcinomas are more likely to have a septal nodule and a nodule diameter of over 1 cm. [29]Other tests are proposed to better identify the diagnosis it is about 1/Endoscopic retrograde cholangiopancreatography (ERCP)may demonstrate intraluminal filling defects or a cystic cavity communicating with the biliary tree. Apart from helping in the diagnosis of a cystadenocarcinoma, ERCP is also helpful in decompressing the biliary system in patients with biliary obstruction. 2/ Magnetic resonance cholangiopancreatography (MRCP) is an alternative to ERCP in the evaluation of pancreatic and biliary duct systems. Even though the resolution of MRCP is somewhat inferior to ERCP, the procedure is noninvasive and less expensive.[30]. A Preoperative core needle biopsy to identify malignancy is not recommended as this is not precise and brings the risk of needle seeding and dissemination [29, 30]. Elevated CEA and CA 19-9 in the serum or the cystic fluid helps in diagnosis and follow-up of patients. A normal level does not eliminate a biliary cystadenoma; some simple liver cysts may also show raised serum or cystic fluid CEA or CA 19-9 [31]. Cystadenomas may fast a progesterone receptor in the mesenchymal cell part. Other markers demonstrated on immunohistochemistry are CA19-9, CEA, vimentin (structural protein that in humans is encoded by the VIM gene.),and cytokeratin. In situ hybridization has confirmed selective positivity for albumin messenger RNA in cystadenocarcinomas. Available evidence shows that biliary cystadenomas tend to happenmainly in women because these tumors are hormonally responsive. This concept is more reinforced by immunohistochemical studies demonstrating positive estrogen/progesterone receptors associated with biliary cystadenomas.[28].

Previously, biliary cystadenomas have been treated with many techniques like marsupialization, internal Roux-en-Y drainage, aspiration, sclerosis, or partial resection. Though, all these procedures have been associated with great proportions of recurrence [32]. Therefore the treatment of choice for hepatic cystadenomas is surgical resection. Complete resection of the tumor is imperative to avoid local recurrence and malignant transformation. A total lobectomy is sometimes necessary for larger lesions or in the presence of adenocarcinoma. For minor lesions, enucleation alone can usually be achieve with conservation of the residual hepatic parenchyma except if the tumor is in a central location close to the hepatic hilum. Enucleation is conceivable because cystadenomas have a thick fibrous capsule that can be separated frankly without major bleeding or biliary leak.[28].

In a study of 51 patients, Gamblin et al investigated the efficacy of laparoscopic resection of symptomatic hepatic cysts. 90% of the lesions in this study were simple cysts, and 10% were cystadenomas. The results of this study support that a routine laparoscopic attitude can be indicated to treat benign

symptomatic cysts. However, traditionalsurgical approach should be reserved for cases of expected malignancy or for those in which laparoscopy is contraindicated or the cyst recurs after laparoscopic treatment.[33].Abu Hilal et al concluded that the laparoscopic approach represents a harmless choice for the management of benign and uncertain liver lesions, even when major hepatectomy is necessary. [34].

Liver transplantation may be required in the rare manifestation of wide bilobar extension of the tumor

IV. Conclusion

In conclusion, the diagnosis of biliary cystadenoma must be suspected whenever radiological imaging is indicative of a multilocular cystic liver lesion, especially in a middle-aged woman. It is an essential differential diagnosis for a hydatid cyst of the liver especially in endemic areas. As it is extremely difficult to distinguish in preoperative period, a malignant from a benign lesion, the recommended treatment of choice for any suspected biliary cystadenoma is a complete resection (Gold standard) to be safe and to prevent recurrences. Enucleation is alternative possibility and is indicated where resection is difficult due to the size and site of the cyst. Laparoscopic surgery represents a safe approach for the management of benign and uncertain livercystic lesions

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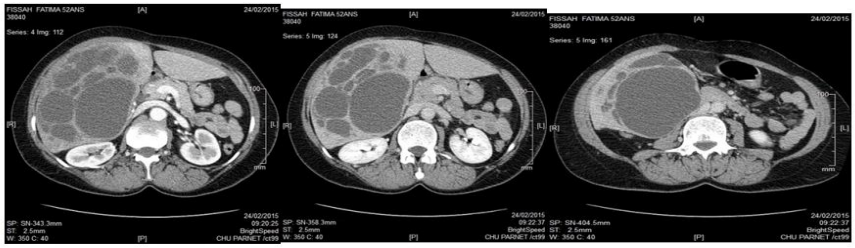

Figure 1-2-3 showed: cystadenoma, without signs of loco regional infiltration

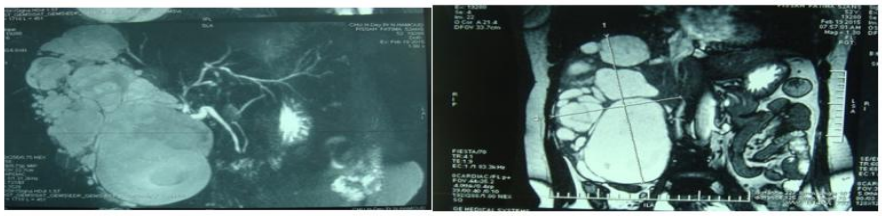

Figure 4-5 showed: Absenceof obvious communication with the biliary tree

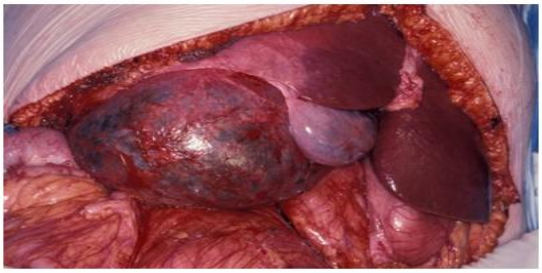

Figure 6:showing intraoperative a large hepatic cystadenomas

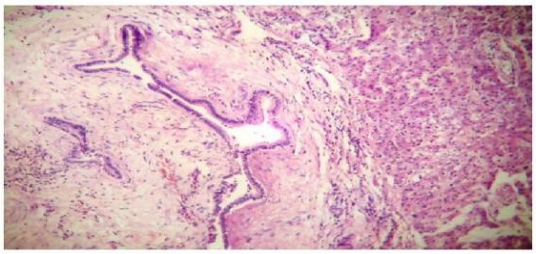

Figure 7: showing dilated bile duct with low columnar epithelium and spindle-shaped cells in stroma.

A.Bendjaballah. MD FACS « Large cystadenoma of the liver - diagnostic and therapeutic approach."IOSR Journal of Dental and Medical Sciences (IOSR-JDMS), vol. 17, no. 7, 2018, pp 63-66.