Biliary cystadenoma: A rare occurrence

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SUMMARY

Biliary cystadenoma is a rare benign tumour with the potential to transform into malignant carcinoma of the biliary ductal system. There is difficulty in differentiating a benign one from a malignant biliary cystadenoma, and therefore these lesions should always be completely resected. We report a case of biliary cystadenoma which underwent a complete resection.

INTRODUCTION

Biliary cystadenomas are rare cystic lesions of the liver. They account for less than 5% of nonparasitic cysts of the liver and occur frequently in middle-aged women. The size varies from 1.5 to 35 cm. These cysts need to be differentiated from other cystic lesions such as simple cysts, hydatid cysts, abscesses, hematomas, and polycystic liver disease.[1]

They are often benign lesions with a malignant potential. They develop from either an aberrant bile duct or directly from a primitive hepatobiliary stem cell. Majority are intrahepatic (85%) and occasionally are seen to arise from the gall bladder.[2]

Due to its premalignant nature and high rate of recurrence after incomplete removal, all suspected lesions should be surgically removed. It is not always reliably distinguish the simple cyst from the hydatid cyst or the benign biliary cystadenoma. In such situations, deroofing, marsupialisation, or partial resection of the suspected cysts has resulted in a very high rate of recurrence (>90%).[3]

We report here a case of recurrent cystic lesion of the liver that was subsequently confirmed as biliary cystadenoma upon total resection.

CASE PRESENTATION

A 50-year-old woman known to have recurrent cystic lesion of the liver was admitted for elective liver operation. Her initial complaint was vague: epigastric pain and distension. Apart from hepatomegaly, no other signs or symptoms were discovered on clinical examination. Upon further investigation, a cystic lesion on the left lobe of the liver was noted. Laparoscopic fenestration was done. A year later, she had the same complaint. Recurrence of the cystic lesion was diagnosed and laparotomy and marsupialisation of the cyst wall was done.

Two years after the second visit, she presented again with a similar problem. Her clinical examination revealed a hepatomegaly measuring 9 cm x 7 cm . However, her liver function tests were normal.

Ultrasonography investigation of the liver revealed a well-defined large cystic lesion of maximum diameter 8 cm occupying liver segments IV with multiple internal septations. Her CT scan that revealed a large 12 cm x 12 cm encapsulated mutilocular cystic lesion at segment IV with internal septations. (Fig.1) suggesting of biliary cystadenoma. On the basis of the internal features of the cyst by CT scan, hydatid cyst, which is rare and non-endemic in this region, was deemed to be an unlikely aetiology at that stage of the investigation. Additionally, medical history of the patient did not suggest any previous exposure to the parasitic infection. Laparotomy was done and revealed a large cystic formation. Aspiration of the excised cyst content showed clear yellow fluid. Excision of the cyst was done. The histology report referred to a 12 cm x 7 cm x 3 cm cystic lesion with a smooth wall lining; the epithelial lining consisted of non-ciliated columnar cells with basal nuclei and mucinous vacuoles (Fig. 2). The final report was biliary cystadenoma.

DISCUSSION

Biliary cystadenomas constitute less than 5% of cystic lesions of the liver. Typically, the patient is middle-aged woman presenting with abdominal pain with distension and a palpable mass. Rare presentations include vomiting, dyspepsia, anorexia, and weight loss.[1] Acute presentation is often pain due to intracystic haemorrhage or rupture of the cyst and fever secondary to infection of the cyst[3] Cystadenomas are known to increase in size during pregnancy and following oral contraceptives suggesting hormonal dependency.[4]

Biliary cystadenomas are usually large multiloculated cystic tumours and are of two types: those with and those without mesenchymal (ovarian-like) stroma. The ovarian-like stroma is thick consisting of compact spindle-shaped cells and supports the epithelium and is often seen exclusively in women. Microscopically the loculi are limited by single layer of cuboidal or non-ciliated columnar epithelium resting on a basement membrane.

Cystadenomas with mesenchymal stroma are considered premalignant with a good prognosis while those without are known to transform to malignancy more often with a poor prognosis.[5]

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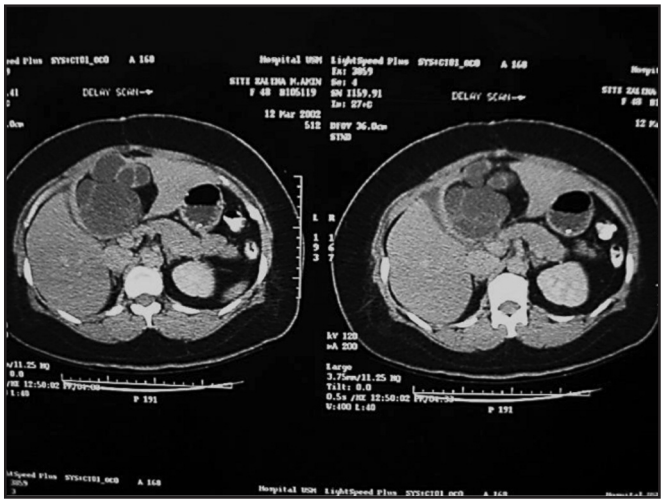

Fig. 1: CT abdomen showing a well-demarcated multi-loculated thick wall cystic mass with internal septations.

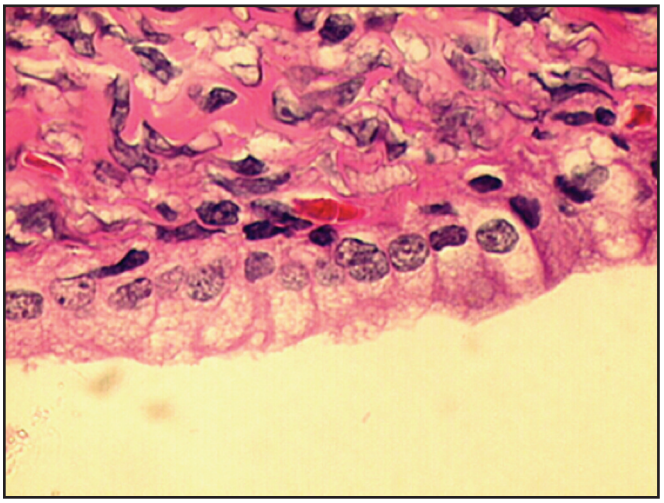

Fig. 2: Microscopic view of part of the cyst wall (x40 magnification) showing the epithelial layer of non-ciliated columnar cells with basal nuclei and mucinous vacuoles.

The cystic fluid may be clear and mucinous. Blood stained fluid within the cyst indicates a malignant component (cystadenocarcinoma). Rarely, the fluid may be bile stained, purulent, proteinaceous, or gelatinous.[6] The septa within the cyst may rarely show calcification.

Differential diagnosis of cystadenomas include simple liver cysts, parasitic cysts (particularly hydatid cysts), haematomas, post-traumatic cysts, polycystic diseases biliary and cystadenocarcinoma. Extra hepatic biliary cystadenomas can typically mimic choledochal cyst.[6]

Diagnosis of biliary cystadenomas is often possible on an ultrasonography and CT scan abdomen. On ultrasound, the cystic nature of the lesion with multiple loculi, septations, and internal echoes, with papillary projections, is typical. CT scan in addition demarcates the anatomic relation to surrounding structures, particularly major vessels. Coarse calcifications in the septae may be seen.

An irregular thickness of the cyst wall, the presence of mural nodules, or papillary projections indicates the possibility of a malignancy.[6]

A preoperative cyst fluid aspiration for diagnosis has been advocated in publications and the diagnostic value is controversial. Promising reports have suggested that cyst fluid CA 19-9 and CEA levels can be helpful in enhancing the accuracy of the diagnosis of biliary cystadenoma and cystadenocarcinoma from other cystic lesion.[7] However, it is not accurate in differentiating both the lesions as inadequate sampling may miss the microscopic foci of the carcinoma in a cystadenoma. Fine needle aspiration and biopsy may risk dissemination of tumour cells. A normal level does not exclude a biliary cystadenoma; some simple liver cysts may also show elevated serum or cystic fluid CEA or CA 19-9.

Two key concerns are of importance in making a diagnosis of biliary cystadenoma. One is the incomplete excision of the cyst, misdiagnosed as a simple cyst or a hydatid cyst, resulting in recurrence and the second is the difficulty in differentiating biliary cystadenomas from biliary cystadenocarcinomas either before or during surgery. Hence the recommendation for any suspected biliary cystadenoma is a complete excision.[8]

CONCLUSION

The diagnosis of biliary cystadenoma should be considered in any multilocular cystic lesion of the liver, particularly in a middle-aged woman. The recommended treatment of choice for any suspected biliary cystadenoma is complete resection as it is extremely difficult to differentiate preoperatively, a benign from a malignant neoplasm.

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