Case Report

Biliary Cystadenoma: A Case Report With Review Of Literature

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Abstract: A rare case of mucinous biliary cystadenoma presented as an upper abdominal mass and progressively increasing jaundice in a 45 years old female. A free lying component of the cystadenoma producing obstruction was present in the common bile duct. Marsupialization of outer cyst with electro-coagulation of its epithelium and excision of inner cysts was performed as a palliative procedure. Diagnosis was confirmed on histopathology with positive estrogen receptors. Postoperative treatment with 20 mg Tamoxifen produced good results without any evidence of recurrence or progression even 30 months after the surgery.

Keywords: Cystadenoma, Biliary tumors, Obstructive jaundice,

Introduction

Mucinous biliary cystadenomas are extremely rare multilocular benign neoplasms. These tumours usually involve hepatic parenchyma and occasionally extrahepatic biliary tract. Their true incidence in different areas of the world is unknown. Eighty to 85% of tumours occur in females with peak incidence between 30-50 years of age. The exact etiology of these tumours remains unknown. The resemblance of embryonic structures originating from foregut suggests their possible origin from ectopic remnants of Von Meyenberg complexe,[1,2] other possibilities includes origin from peribiliary endocrine cells.[3] Environmental factors may play a role. Marked female preponderance suggests a role for hormonal influence. Some of these tumours are positive for estrogen receptors.[4] Different therapeutic options including aspiration and sclerotherapy[5,6,7] fenestration, partial excision masupialization and epithelial electro-coagulation[8,9,10]

However complete surgical resection appears to be the only possible mean to cure the disease. Liver transplantation is also a valid option in cases with extensive or bi-lobar lesions. Presence of estrogen receptors in some of these tumours may warrant treatment with Tamoxifen after palliative resection with possibility of cure or long term effective disease control.

Case Report

A 45 years old female was referred by a local G.P with epigastric mass and progressively increasing jaundice. On presentation she had deep yellow sclera, passing dark urine, clay colored stools and complained of marked itching. She had lost some weight in the past few weeks with decreased appetite. She had eight issues, last child birth five years back. She had never been on any hormonal treatment including oral contraceptives at any stage of her life. On physical examination, she was a medium built female with deep jaundice. A large, firm, non tender smooth mass was palpable in the epigastrium that moved with respiration. There was no ascities. Laboratory investigations revealed her serum bilirubin 21.1mg/dl, Alkaline phosphatase 1033U/l, ALT 37 U/l, INR 1.2, urine was positive for bilirubin. Haemagglutination test for hydatid disease was negative and X-ray chest was unremarkable. Ultrasound reported a large mulitlocular cyst occupying the left lobe of liver and reaching portaheptis. Intrahepatic biliary ducts were dilated, while common bile duct could not be defined.

Computed tomography (CT) confirmed the ultrasound findings and also reported that cysts close to portahepatis having connection with the biliary channels, however commo

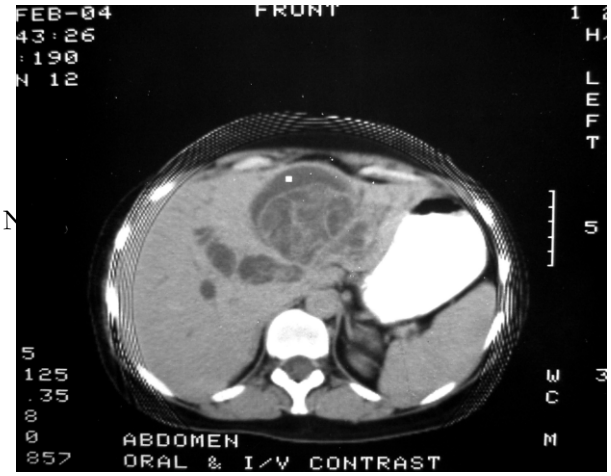

Fig-1: CT scan prior to surgery in Feb. 2004 showing large bilobar biliary cystadenoma

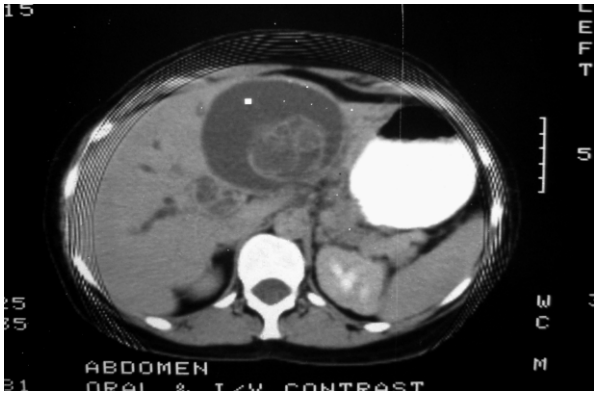

Fig-2: Larger outer cyst and smaller inner cysts, resembling hydatid cyst.

A provisional diagnosis of Biliary cystadenoma was made with a differential diagnosis of hydatid cyst. Surgical exploration was performed under general anesthesia in supine position through upper abdominal midline incision. A large cyst occupying almost whole of the left lobe of liver was encountered. Bile stained mucinous fluid was aspirated and cyst opened for palliative procedure as the lesion was involving the region of portahepatis, with extension into right lobe of liver as well, complete surgical resection was not possible. Unlike echinococcus cyst no germinal layer found instead smooth inner lining and multiple cysts, extending to portahepatis with intervening hepatic paryncyma at places. Common bile duct was enormously dilated. Cysts were excised in toto except close to hepatic confluence where small cysts removed piecemeal. Choledochotomy was performed and a separate cyst of 4cmx8cm **(Fig.3)** removed, bile duct t

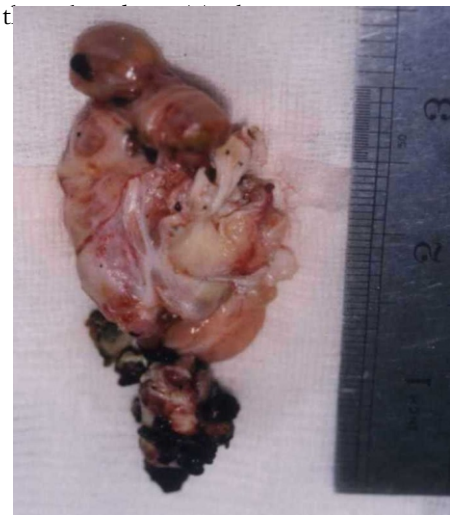

Fig-3: Large free component of biliary cystadenoma removed from common bile duct, (Note the black calcium billirubinate deposits on the distal tapering end of cystadema impacted in distal common bile duct)

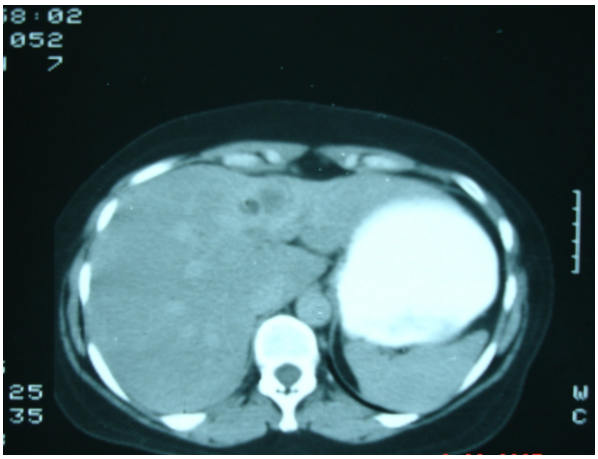

Fig-4: Mucinous cystadenoma showing a dilated cystic space lined by mucus secreting columnar epithelium and surrounded by vascularized fibrocollagenous tissue. (H&E x 40)

Wall of outer cyst de-roofed with electrocoagulation of its epithelial lining. On histopathological examination, multi-locular cystic lesions, lined by mucous secreting benign columnar epithelium. Intervening stroma composed of collagenous fibrous tissue, congested blood vessels and focal chronic inflammatory cells **(Fig.4&5).**

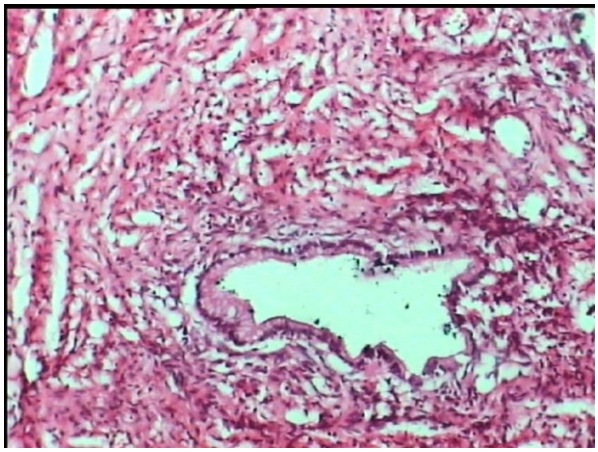

Fig-5: Multiloculated cysts lined by benign exfoliated epithelium and containing eosinophlic secretions(HE10X)

A diagnosis of mucinous biliary cystadenoma was made. Estrogen receptor status was then requested

that turned out to be positive. Post operative course was uneventful. Patient was discharged on the 5th post operative day with a T-tube in position. On 12th day T-tube was removed after a normal T-tube cholangiogram. Patient was prescribed Tamoxifen 20 mg daily.

Patient was last examined 18 months after surgery with no clinical signs of recurrence and patient symptom free. However ultrasound scans at six months intervals revealed small 2.5x3 cm area of residual cysts with areas of fibrosis. This area of residual disease/ recurrence is static in a recent follow up scan **(Fig.6).**

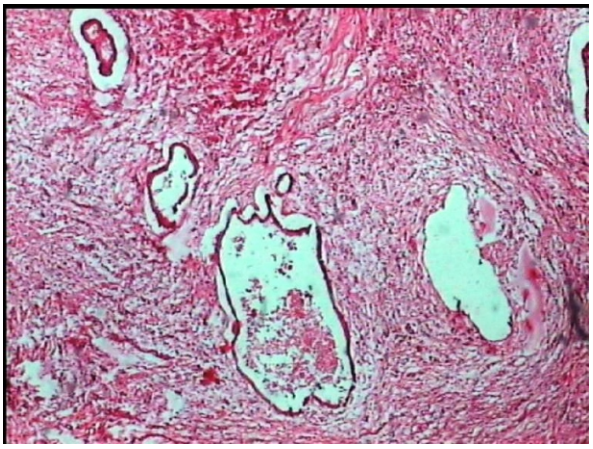

Fig-6: Recent CT scan showing small residual cyst with thick fibrous walls

Discussion

Biliary cystadenoma is a rare condition with an overall incidence of less than 0.05% for the cystic tumours of the liver (11). The condition predominantly affects females after their fourth decade of life (12). The exact etiology of the condition remains unknown though; its origin has been attributed to aberrant bile ducts (1). Presence of estrogen receptors in some reported cases and marked female preponderance may also suggest hormonal influence as a possible etiological factor (4,13). Immunohistochemistry and microscopic studies have ruled out the possible origin from an ectopic ovarian tissue (14). Biliary cystadenomas usually present as multilocular (rarely unilocular) cystic lesions lined by cuboidal or columnar epithelium surrounded by thick fibrous capsule and an outer layer of loose connective tissue. They have variable vascularity and true connections with bile ducts are rare (8,15). Most cysts contain serous, clear transudate with electrolyte composition similar to that of serum (5,16). Rarely bile stained fluid is encountered (8, 9, 10). Some of the biliary cystadenomas usually asymptomatic are discovered incidentally after ultrasound scan done for some other reason. Pain, nausea, upper abdominal fullness and palpable mass are the common symptoms at presentation. These symptoms appear usually when the cyst is more than 5cm in size (11). Obstructive jaundice, cyst rupture, portal vein compression and intracystic hemorrhage are rare and may lead to unusual presenting features (17). Ultrasonography, CT scan and MRI are all very sensitive investigations to diagnose the condition. ERCP can be helpful in establishing the cause and site of obstruction in biliary cystadenomas presenting with obstructive jaundice (external compression/direct involvement or communication). FNAC and cytology of aspirated fluid is reasonably accurate to differentiate from other similar lesions (18). CA 19-9 in serum or cyst fluid may be helpful in diagnosis of cystadenocarcinoma though this test is not 100% reliable.

Other cystic lesions of the liver that are to be differentiated from biliary cystadenoma include simple cysts, echinococcal cysts, necrotic neoplasms, abscesses, embryonal sarcoma, polycystic liver and cystic metastases.Conservative medical treatment is not effective. Different surgical treatments have been attempted including aspiration and sclerotherapy with ethanol (19,20,21), doxycycline (7) and minocycline (22), partial excision, epithelial electrocoagulation, marsupialization, fenestration and omentopexy (23-28). The recurrence remains high after these procedures (29-30) Complete surgical resection is required to avoid local recurrence. For smaller lesions enucleation and for larger lesions or in case of associated malignancy complete lobectomy is the procedure of choice (13,17,18,24,31). Hepatic transplantation may be required in some patients with extensive bilobar disease (32-35). Biliary cystadenomas have 2-4% risk of malignant transformation (36, 37). Benign to malignant ratio of biliary cystadenoma and biliary cystadenocarcinoma is 57-1, it is not clear whether cystadenocarcinoma starts de novo or as a result of malignant transformation in an already present biliary cystadenoma (36, 38-40). The case under discussion is unique in many ways. Communication with the biliary channels is rare and this may be the cause of bile stained mucinous fluid instead of serous fluid in the cyst cavity in this patient. Presence of large 8cmx4cm size cyst adenoma within common bile duct has not been reported previously. As this segment of cyst adenoma most probably would have come from the parent cystadenoma in left lobe of liver. The presence

of calcium billirubinate salts on the distal tapering end indicates its presence in common bile duct for more than few weeks. The size of the cystadenoma in common bile duct indicates that it might have been growing in the bile duct and later got detached from the parent cystadenoma in the left lobe the liver.Role of Tamoxifen in estrogen receptor positive biliary cystadenomas has not yet been investigated. Post operative treatment with Tamoxifen in our case brought good clinical results and its role in such cases needs to be further evaluated. As no medical treatment has been effective in the treatment of biliary cystadenoma, possible role of Tamoxifen in biliary cystadenoma merits further evaluation.

Summery

Biliary cystadenoma is a rare benign cystic disease of the liver which predominantly affects women. Patients usually present in the fourth decade of their life with symptoms due to its size or complications. Treatment of cystadenoma is complete surgical resection to prevent recurrence. Extensive disease especially involving both lobes of the liver may not be possible to eradicate with routine surgical measures. Liver transplantation may have a place for such patients. Some of the cystadenomas are estrogen receptor positive. The role of Tamoxifen need to be further explored for such patients. In our case palliative resection in an estrogen receptor positive cystadenoma followed by Tamoxifen treatment produced good clinical results.

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