Case Report

DOI: 10.7860/JCDR/2016/14841.7813

Pathology Section

An Unusual Combination of Biliary Cystadenoma and Renal Angiomyolipoma- A Case Report

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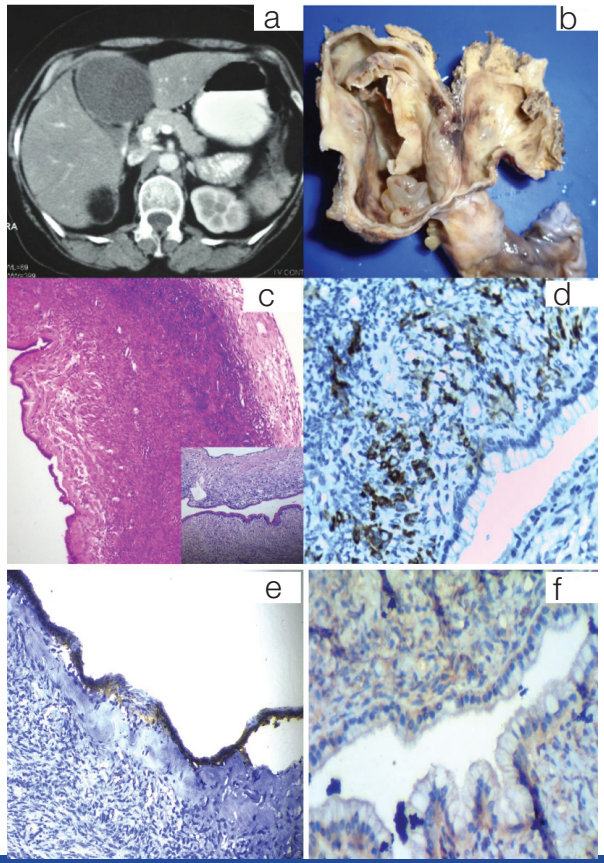
ABSTRACT

Hepatobiliary cystadenomas are mucinous cystic neoplasms arising in the liver, extrahepatic bile ducts or gall bladder. Extrahepatic cystadenomas are rare neoplasms requiring complete excision as there is a chance for recurrence and malignant transformation. Angiomyolipoma (AML) belongs to the perivascular epithelioid cell group of neoplasms, and kidney is the commonest site involved. We report a case of an unusual combination of extrahepatic biliary cystadenoma with angiomyolipoma in an adult female without evidence of tuberous sclerosis. The patient presented with abdominal discomfort and on ultrasonological examination showed a cyst close to liver suggestive of hydatid disease and an incidental mass in kidney which was diagnosed as angiomyolipoma. Histopathological examination revealed biliary cystadenoma of liver and renal angiomyolipoma. Clinical and radiological evaluation did not show any manifestations of tuberous sclerosis. Such an unusual incidence of two separate neoplasms in a patient without syndromic association was not obtained even after extensive literature search.

Keywords: Benign, Laparotomy, Neoplasm

CASE REPORT

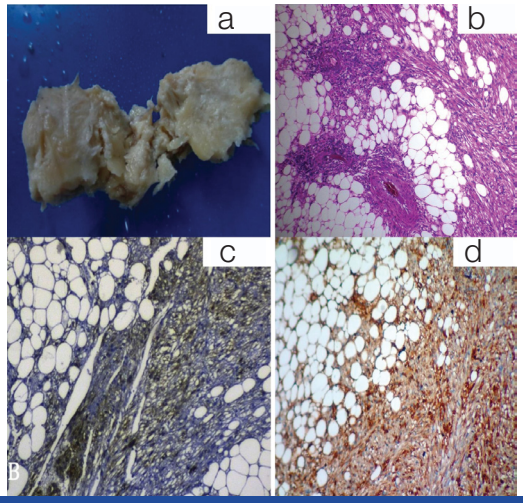
A 50-year-old female presented with vague upper abdominal discomfort and blotting sensation for the period of one month. There was no evidence of jaundice, vomiting, fever or any abdominal mass on examination. All investigations were within normal except raised Ig anti-echinoccus antibody. USG Abdomen showed a single multiloculated cyst in the left lobe of liver suggesting a possibility of hydatid cyst and a hyperechoic lesion in the right suprarenal region with fat density devoid of calcification and haemorrhage, suggestive of renal angiomyolipoma. CECT showed a 6.8x5.7 cm well defined cystic lesion with internal septations of varying density at the porta hepatis. No enhancing mural nodule/ calcification was seen [Table/Fig-1a]. With the provisional diagnosis of hydatid cyst liver and renal angiomyolipoma, laparotomy was performed. A 6x6 cm cystic lesion in the antero-inferior surface of liver involving inferior border along with segment 4 of liver was excised. It was close to gall bladder and right branch of portal vein. The aspirated cyst fluid was serous in nature and hydatid sand was not demonstrated. Right kidney was exposed and a lipomatous circumscribed mass in the upper pole was enucleated. Specimens sent for histopathology examination. Of the two gross specimens received, one was a collapsed multiloculated cyst measuring 6x6x3cm with smooth inner wall and attached liver tissue on periphery [Table/Fig-1b]. Microscopy revealed liver tissue with a cystic neoplasm lined by columnar to mucinous epithelium. Cyst wall was composed of thick fibro collagenous tissue and ovarian like stroma. There was no evidence of intestinal metaplasia, nuclear atypia or malignancy [Table/Fig-1c]. IHC studies of the liver cyst showed CK 7 positivity of the lining epithelium. Ovarian stroma was positive for inhibin and Melan A negative [Table/Fig-1d-f]. The second specimen was an enucleated renal mass as two yellowish white tissues with faint lobulations and was composed of mature adipocytes, whorls of smooth muscle cells and few proliferated blood vessels [Table/Fig-2a&b]. HMB 45 & Melan A positive and a negative desmin confirmed the perivascular origin of renal neoplasm [Table/Fig-2c&d]. Final histopathological diagnosis was extrahepatic biliary cystadenoma and renal angiomyolipoma. The patient is doing well on regular follow up.



[Table/Fig-1a]: CT scan showing liver with a well-defined cystic lesion **[Table/Fig-1b]:** Gross specimens showing multiloculated collapsed cyst with smooth inner wall **[Table/Fig-1c]:** Microscopy of cyst wall with lining epithelium and fibro collage nous ovarian like stoma. (H &E stain, 100x), Lining epithelium was mucicarmine positive (inset) **[Table/Fig-1d]:** Ovarian stroma showed inhibin positivity (IHC stain, 400x) **[Table/Fig-1e]:** CK 7 positive lining epithelium (IHC stain, 200x) **[Table/Fig-1f]:** Melan A negative stroma (IHC stain,400x)

DISCUSSION

Liver is a common site for non-neoplastic and neoplastic cystic lesions; hence a correct diagnosis is important for treatment. Hepatobiliary cystadenomas are rare cystic mucinous neoplasms of liver, extrahepatic duct or gall bladder. This belongs to mucinous cystic neoplasms of WHO classification. It is similar to ovarian and pancreatic mucinous neoplasms. Cystadenoma of liver can be intrahepatic (90%) or extra hepatic (10%) in origin [1,2]. There is a strong female predominance with few cases reported in males and children [1,3]. The presenting symptoms are obstructive jaundice, abdominal pain, fever and haemobilia [1,2,4]. The characteristic



[Table/Fig-2a]: Gross specimen of enucleated renal mass with whitish and yellowish areas. **[Table/Fig-2b]:** Microscopic appearance of AML (H &E stain, 100x). **[Table/Fig-2c&d]:** Neoplastic cells positive for HMB 45 and Melan A (IHC stain, 400x).

radiological appearance is a single multiloculated cyst with or without calcification. Distinction from benign lesions and malignant cysts are not always possible by radiology [4].

Gross examination shows a single multiloculated cyst containing clear fluid and smooth glistening inner wall. The solid inner protrusion should raise a suspicion of malignant transformation. Microscopy shows cyst lined by mucinous lining epithelium, middle layer of cellular mesenchymal stroma, an outer layer of dense collagenous ovarian like stroma which border the lesion separating it from hepatic parenchyma [4,5]. Rarely, lining epithelium can show intestinal metaplasia. If epithelium shows dysplastic changes, malignant transformation into cystadenocarcinoma should be considered. Cyst wall can show haemorrhage, micro calcification, epithelioid granuloma and cholesterol clefts. Cases of biliary cyst adenoma without mesenchymal stroma are described in males and such cases are associated with increased risk of recurrences and malignancy [2,4]. Special stains like Alcian blue and Mucicarmine for lining epithelium and IHC markers-CK7, CK19, CA 19-9 can be used. The differential diagnosis are simple/multiple cysts and echinococcal cyst [2,3]. Unilocular nature and absence of ovarian stroma distinguish it from simple cyst of liver. Multiple locules and calcification mimic a hydatid cyst on radiological studies, but lamellate hyalinized capsule with germinal membrane and daughter cysts containing protoscolices are absent. In our case per operative examination excluded the diagnosis of hydatid cyst. Biliary cystadenomas require cyst enucleation and hepatic resection for complete excision as there is a chance for recurrence and malignant transformation into cystadenocarcinoma [2].

Angiomyolipoma of kidney is a mesenchymal neoplasm included in the PEComa group composed of perivascular epithelioid cells. Neoplastic cells are epithelioid to spindly arranged radially around the vascular lumen. Normal counterpart of PEC cell is not identified in any organ [6]. AML can present as isolated cases or associated with tuberous sclerosis. Isolated angiomyolipoma is common (80%), usually unilateral, solitary and more on right side [7]. The mean age group affected is fourth decade with female predominance. Our case falls into this category. Second group of AML is associated with tuberous sclerosis (20%). These tumours are larger, bilateral and multiple [7]. Mean age of incidence is first decade with equal incidence in males and females. One third of cases of AML show manifestations of tuberous sclerosis and 80% of patients with tuberous sclerosis develop AML [7]. AML can be asymptomatic detected incidentally or symptomatic presenting as acute flank/abdominal pain, haematuria and palpable mass. AML can involve liver, which is the most common site of extra renal AML [8]. Any association between AML and biliary cystadenoma is not reported in literature.

USG and CT findings are characteristic; a well circumscribed, cortical-based, heterogeneous tumour predominantly of fat density. Gross appearance is yellow to grey un-encapsulated mass demarcated from normal renal parenchyma. Rarely can it involve the IVC or peri renal lymph nodes. Microscopically tumour is composed of mature adipose tissue, smooth muscle cells and convoluted thick walled blood vessels. Neoplastic cells are epithelioid to spindly with clear to granular and lightly eosinophilic cytoplasm, small centrally placed, round to oval nuclei with small nucleoli [6,7]. Nuclear atypia, mitoses and necrosis can be seen. IHC shows both smooth muscle and melanocytic differentiation. HMB-45, Melan A, MiTF and tyrosinase are positive. Few cases show CD117 and Progestrogen receptor positivity. Desmin is less often positive. CK & S100 are negative. TSC1 & TSC2 genes play an important role in the pathogenesis of PEComa in patients with tuberous sclerosis. AML is a benign tumour and wide excision is the treatment. The exact criteria for malignancy are not established [7]. Few cases with large size, mitosis and necrosis have behaved in a malignant fashion and such cases need aggressive management [7].

CONCLUSION

The unusual occurrence of two benign neoplasms, extrahepatic biliary cystadenoma and renal angiomyolipoma is not reported in literature, especially in a patient without evidence of Tuberous sclerosis. The significance, whether it was an accidental case or whether it carries any genetic implication need to be found in future years.

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FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **May 10, 2015**
Date of Peer Review: **Aug 11, 2015**
Date of Acceptance: **Oct 15, 2015**
Date of Publishing: **May 01, 2016**