Case Report

Volume - 1 | Issue - 3 | July - 2017

BENIGN MUCINOUS CYSTIC NEOPLASM OF LIVER-A RARE CASE REPORT AND REVIEW OF LITERATURE

Dr. Zubaida Rasool Additional Professor, Department of Pathology, SKIMS,Soura.

Dr. Huzaifa Tak Senior Resident, Department of Pathology, SKIMS, Soura

Dr Nazia Bhat Senior Resident, Department of Pathology, SKIMS, Soura

Dr Kanika Senior Resident, Department of Pathology, SKIMS, Soura

Dr Nuzhat Samoon Post Graduate, Department of Pathology,SKIMS,Soura

ABSTRACT Cystic diseases of the liver occur in about 5% to 10% of the population . Hepatobiliary cystadenomas (HBCs) and cystadenocarcinomas are rare lesions which comprise one percent of liver cystic neoplasms and 5% of symptomatic hepatic cysts. Here, we report the case of a female patient who presented with abdominal pain and on imaging revealed smooth walled cystic lesion in liver which was reported as biliary mucinous cystic neoplasm on histopathology.

KEYWORDS

Liver, Hepatobiliary Cystadenomas, Mcn

ARTICLE HISTORY

Received : 28 January 2017 | **Accepted : 16 March 2017** | **Published : 05 July 2017**

***Corresponding Author** Dr. Huzaifa Tak

Senior Resident, Department of Pathology,SKIMS,Soura huzy88@gmail.com

INTRODUCTION

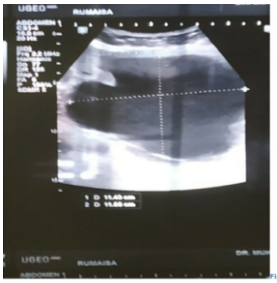
Cystic diseases of the liver occur in about 5% to 10% of the population [1]. Hepatobiliary cystadenomas (HBCs) and cystadenocarcinomas are rare lesions which comprise one percent of liver cystic neoplasms and 5% of symptomatic hepatic cysts [2,3]. In approximately 85% of cases the lesions usually develop in liver and these lesions are very rare in extrahepatic bile ducts and gallbladder [4].

Previously, mucinous cystic neoplasm of the liver (MCN-L) had been classified as biliary cystadenoma or biliary cystadenocarcinoma. However, the World Health Organization (WHO) classification of 2010 defined MCN-L as a counterpart of MCN of the pancreas (MCN-P). MCN-L is now defined as 'a cyst forming epithelial neoplasm, usually with no communication with the bile ducts, composed of cuboidal to columnar, variably mucin-producing epithelium, associated with ovarian-type subepithelial stroma' and is subdivided into non-invasive and invasive types. [5].

In this report, we present a female patient with a biliary mucinous cystic neoplasm of the liver.

Presentation of case:

A 38-year-old woman was admitted to the department of Surgical Gastroenterology of our Hospital which is a tertiary level super speciality hospital after an episode of severe abdominal colic. Ultrasound (USG) examination of the abdomen done in our hospital revealed a multicystic lesion seen in right lobe of the liver measuring 12.1x10.8x11.9 cm with a cyst volume of 814.2ml (Fig 1).



usg image showing a cystic lesion in the liver

The woman was incidentally diagnosed with this liver Space occupying lesion on ultrasound three years back at the time of routine antenatal checkups but no intervention was done then .The patient remained nearly asymptomatic throughout this time period except vague abdominal pain since a year from the time of admission to our hospital. There was no history of jaundice, loss of appetite or loss of weight.

On examination general condition of the patient was good, no pallor, icterus or any lymphadenopathy was noted.The patient had all viatls within normal limits.On local examination, a hard lump was felt in right hypochondriac area. No ascites was seen.

The patient was investigated for Liver function and full blood counts (Table 1) which were within normal limits. Anti-echinococcal IgM and IgG antibodies and viral markers for hepatitis B and C were negative. Tumor markers for CEA and CA19-9 were negative.

Table 1:

Hemoglobin (g/dL)	10.5
Leukocytes ($\times 10^3$ U/L)	5.5
Platelets ($\times 10^3$ U/L)	200
Total bilirubin (mg/dL)	3.5
Direct bilirubin (mg/dL)	1.2
AST (IU/L)	25
ALT (IU/L)	29
Alkaline phosphatase (IU/L)	401
ESR	43
Total protein (mg/dL)	7.2
Albumin (mg/dL)	2.9
Prothrombin time (s)	10

Triple phase Computerized tomography (CT) scan of abdomen which followed ultrasound revealed a large 11x11x10cm (multiloculated) cystic lesion with septation,predominantly exophytic seen arising from segment IV B of left lobe of liver with associated minimal dilatation of segmental IHBR [Fig-2].

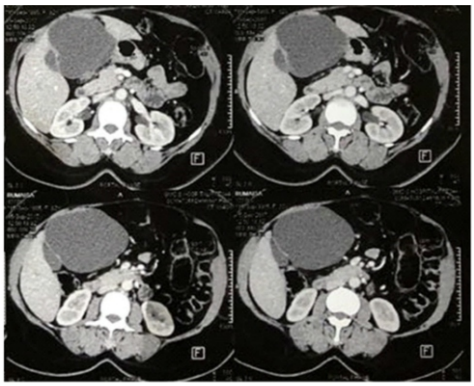


FIGURE 2 :TRIPLE PHASE CT IMAGE REVEALING CYST IN SEGMENT 4 OF THE LIVER

Rest of the liver parenchyma was unremarkable. 99mTc Mebrofenin hepatobilary study done showed normal findings. Magnetic resonance cholangiopancreatography (MRCP) done revealed an exophytic cystic lesion 14.7x10.9x10.7cm which appears to communicate with segment IV duct .On post contrast enhancement images, no evidence of contrast enhancement was seen and lesion appeared to be separate from gall bladder.[Fig-3].

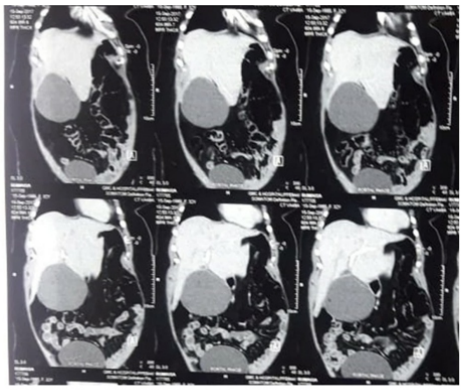
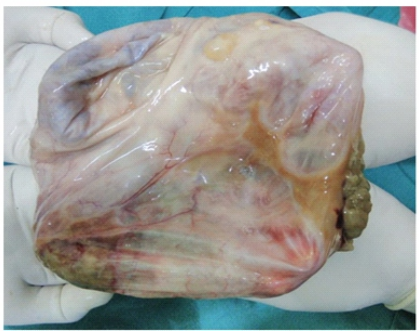


FIGURE 3 MRI image revealing a cystic image in the liver

The patient underwent a laparotomy that revealed a large cystic lesion within liver parenchyma which was followed by a cystectomy and the fully resected cyst along with portion of liver was sent for histopathological examination. Gross examination showed a multilocular cystic lesion measuring 16x15 cm with a pale grey smooth cyst wall [Fig-4]

of thickness that varied from 0.1 cm to 0.5 cm. On histology of the lesion showed a cyst wall lined by low columnar epithelium with basally oriented nuclei and apical mucin. Cuboidal epithelium with focal pseudostratification is also noted. No nuclear or cellular atypia was noted.An underlying mesenchymal stroma resembling ovarian stroma with hemosiderin laden macrophages and edema were seen. Hence a diagnosis of mucinous cystic neoplasm of liver (biliarycystadenoma) was made. [figures 5,6 n 7]. The postoperative course of the patient was uneventful and she was discharged one week later. The patient is on follow-up and is doing well.

FIGURE 4 cut surface of the cyst revealing smooth surface with no solid



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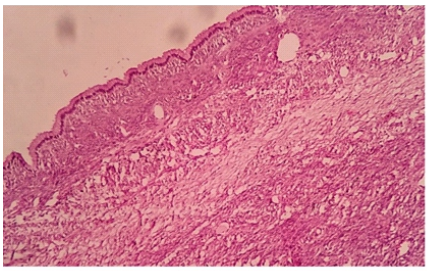

Figure 5.low power view revealing the cyst wall lined by mucinous epithelium and having ovarian type stroma in the underlying wall.(H&E X10)

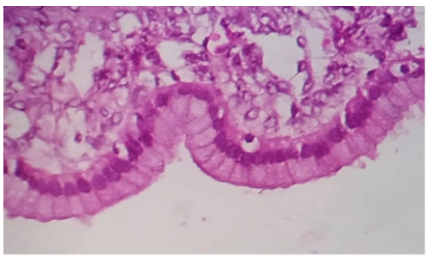

Figure 6 high power view of the same showing mucinous lining with basally placed nuclei and apical mucin(H&E X 40)

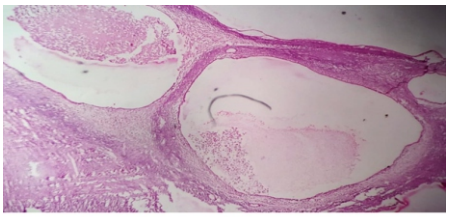

Figure 7 low power view revealing multiloculated cystic lesion filled with mucinous secretions(H&E X 10)

DISCUSSION

The differential diagnoses of hepatic cystic lesions include simple cysts, parasitic cysts, mucinous cystic neoplasms, congenital cystic dilatation, degenerated metastatic tumors, mucin producing metastatic tumors, cystic hemangioma, lymphangioma, hepatic foregut cyst, and mesenchymal hamartoma and teratoma [6-8]

These relatively uncommon tumors may be misdiagnosed as simple cysts, but it is important to make the correct diagnosis, given that the treatment of choice for biliary cystic neoplasms is complete excision to prevent recurrence[9].

These cystic neoplasms do not usually communicate with the bile ducts.The presence of biliary communication indicates intraductal papillary mucinous neoplasm (IPMN) rather than HBCAs [10,11].

Biliary cystic tumors are typically slow-growing lesions with a reported size that can range in diameter from 1.5 to 35 cm [11,12].

Hepatobiliary cystadenomas are generally symptomatic but have been accidentally discovered on rare occasions during radiological imaging for other reasons [13,14] When symptomatic, the clinical presentation is nonspecific. Patients may present with symptoms of mass effects, including abdominal pain, fullness in the upper abdomen, abdominal distension, nausea, vomiting, indigestion, and a palpable abdominal mass [13,15,16,17]. The tumour may occasionally cause biliary compression, resulting in jaundice and possibly cholangitis [17, 18] Rarely, the tumours may rupture, bleed, become infected, or cause vena caval compression and obstruction.

In 2010, the World Health Organization (WHO) had categorized the biliary cystadenoma into Biliary Mucinous cystic neoplasms (BMCN) with low-, intermediate-, or high-grade intraepithelial neoplasia and the biliary cystadenocarcinoma into BMCNs with an associated invasive carcinoma [19].

The presence of an ovarian stroma was accepted as a requirement for the diagnosis of BMCNs. All types of BMCN can become quite large; they range from 1.5 cm to 30 cm in diameter (20,21). Approximately 50%-55% of cystadenomas are located in the right lobe with the

remaining located in the left lobe or in both lobes (30%-40% in the left lobe and about 15%-20% in both lobes) while few arise from extrahepatic ducts[20,22,23].

Radiologic features such as papillary projections, internal septations, wall thickness, irregularities, and mural nodules suggest the possibility of a BCA, all of these except papillary projections may be observed in simple cysts as well albeit at a lower frequency [24, 25].

Preoperative cyst fluid aspiration for diagnosis has been advocated in the published literature. Cyst fluid CA19-9 and CEA levels can be helpful to enhance the accuracy of diagnosis of BCAs and cystadenocarcinomas from other cystic lesions [26, 27]. However, it is not accurate in differentiating BCAs from cystadenocarcinoma, as inadequate sampling may miss the microscopic foci of carcinoma in a cystadenoma [28-30]. Preoperative differentiation of BCAs from cystadenocarcinoma is extremely difficult and can only be done after pathological examination [31-33]. FNA and needle biopsy may risk dissemination of tumour cells and it is not generally recommended, especially when surgery is planned

CONCLUSION

Despite improved imaging modalities and increased abdominal imaging ,histopathological examination of the resected cyst remains the mainstay diagnostic modality to differentiate this benign enity from other benign entities as well from its malignant counterpart

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