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Case Report

Mucinous Cystic Neoplasm of Liver: A Case Report from a Center in Kachchh District

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ABSTRACT

Mucinous cystic neoplasms of the liver (MCN-L) are rare cystic tumors that occur within the liver parenchyma, or less frequently, in the extrahepatic bile ducts. Epidemiologic data are limited due to indolent nature of the tumor and recent changes to the diagnostic criteria proposed by World Health Organization (WHO). The etiology remains unclear, however female predominance, age of onset, and hormonally responsive ovarian-type stroma, suggest ectopic organogenesis during embryologic development. MCNs are typically identified on imaging. Differentiation from common cystic lesions of liver is important for proper management. Complete excision is recommended for all lesions as malignant transformation has been reported.

Keywords: Ectopic organogenesis, Liver, Mucinous cystic neoplasm

INTRODUCTION

Hepatic cysts are common lesions easily detectable by modern imaging modalities, nearly affecting one of four adults, mainly in fourth decade of life.[1,2] The hepatic cysts are heterogenous lesions (Table-1) which differ in etiopathogenesis, prevalence and clinical manifestations. Majority of these cystic lesions are incidental and follow a benign course, while few are associated with serious morbidity and mortality.[1,2] Although most are unilocular, thin walled simple cysts, few show multiloculation with thickened walls.[3-5] The multiloculated cysts were previously referred to as cystadenomas, while those with collagen rich stroma and spindle cells were labelled as cystadenoma with mesenchymal stroma.[6,7] However, with the immunohistochemical identification of estrogen and progesterone receptors (ER & PR) in the stromal cells, the consensus descriptor changed to cystadenoma with ovarian like stroma.[8,9] In 2010, WHO adapted the nomenclature as Mucinous cystic neoplasms (MCNs), making it a unique entity, with presence of ovarian type stroma as a prerequisite for the diagnosis.[10,11]

MCNs account for less than 5% of all the hepatic cysts with an incidence of 1 in 20000-100000, with predominance (80-90% of reported cases) in Caucasian females in the age group of 55-65 years.[12,13] The risk of malignant transformation has been reported to be 20-23%.[12] The precise etiology of MCNs is unclear.[5] The postulated theories are (a) ectopic rests of embryonic bile duct and primitive mesenchyme,[6] (b) ectopic tissue deposited as part of endometriosis.[8] The latter theory was hindered by lack of uniform expression of CD10 as evident in endometrial tissue.[14] The former was supported by recent transcriptome sequencing based identification of sex cord stromal markers, correlating with the existing pancreatic MCNs theory of process secondary to ectopic primordial germ cells of gonads implanted during embryogenesis.[15]
We hereby, present a case of MCNs of liver (extrahepatic), radiologically suspected as ovarian cyst.

CASE REPORT

A 48-year-old female presented with abdominal mass and no other specific symptoms. Ultrasound imaging (Figure-1) showed a well-defined anechoic cystic lesion, 20 x 12 cm

with thin and thick septations in the right abdomino-pelvic cavity, possibility of ovarian cyst was suggested. The findings corroborated with the computed tomography (CT) scan imaging as well. Laboratory investigations revealed mild anemia, normal liver function tests and CA-125 levels. Viral markers (HIV, HCV, HBsAg) were non-reactive. Intra-operatively, the cyst was seen in the sub-hepatic region with adhesion to the liver and biliary duct. Right adnexa were free from the cyst and seen within the right pelvic region. Uterus and Left adnexa were unremarkable. The cyst was removed in toto with preservation of liver and biliary duct. On histopathological examination, the mass was tense, grey white, cystic in gross appearance with no breach in the capsule. On sectioning, straw colored jelly like material oozed out, displaying multiloculation within the cavity. The thickness of the cyst wall and septae ranged from 0.1-0.2 cm. There were no solid areas (Figure-2). Microscopic examination showed a cyst wall composed of fibro-collagenous tissue lined by columnar epithelium with basally placed round nuclei, fine chromatin, inconspicuous nucleoli and pale eosinophilic cytoplasm. There were no dysplastic features. The cellular population just beneath the epithelium was composed of sheets of compactly arranged spindle cells with hyperchromatic nuclei and eosinophilic cytoplasm, resembling ovarian stroma. The rest of the stromal cells were loosely arranged spindle shaped with no atypia and collagen deposition. A band of sparingly arranged hemosiderin laden macrophages were seen in-between the above two layers. There was no liver parenchyma or other ovarian elements noted (Figure-3). The stromal cells showed immunopositivity for estrogen receptor (ER) and progesterone receptor (PR) on immunohistochemistry, supporting the ovarian stromal cell origin (Figure-4 a & b). The histological and immunohistochemical features were consistent with MCN-L (extrahepatic). The patient has been on regular follow up with no post-surgery morbidity.

Table-1: Classification of hepatic cysts

Non-neoplastic cysts	Neoplastic cysts
Simple (solitary) cyst	Primary . Cystadenoma . Cystadenocarcinoma . Squamous cell carcinoma Secondary . Carcinoma of ovary, pancreas, colon, kidney . Neuroendocrine carcinoma
Polycystic disease	
Parasitic . Hydatid (echinococcal)	
Duct related . Caroli's disease . Bile duct duplication	
False cyst . Traumatic intrahepatic haemorrhage . Intrahepatic infarction . Intrahepatic biloma	
Ciliated foregut cyst	

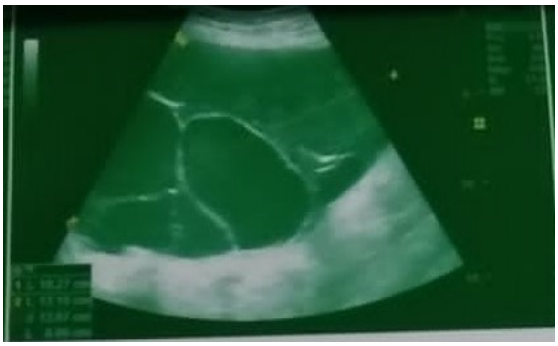

Figure-1: Ultrasonography showing multiloculated cyst with no obvious solid area

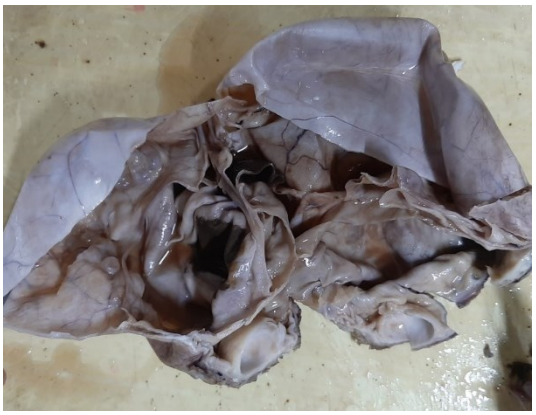

Figure-2: Gross examination showing a multiloculated cyst with variable size septae. Outer and inner surface is smooth with no solid areas

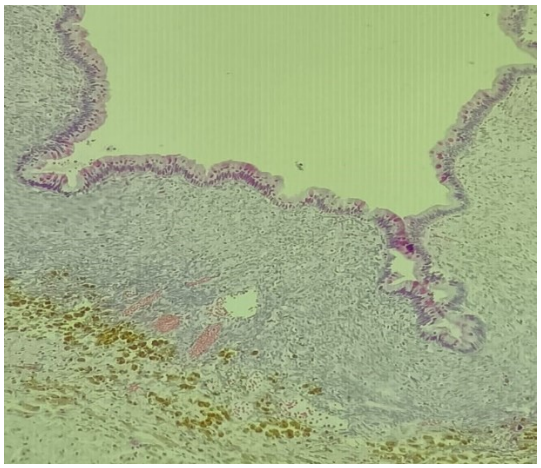

Figure-3 (H & E, 100 X): Cyst wall lined by mucinous secreting columnar epithelium with subepithelial dense stroma, scattered hemosiderophages and collagen rich stroma

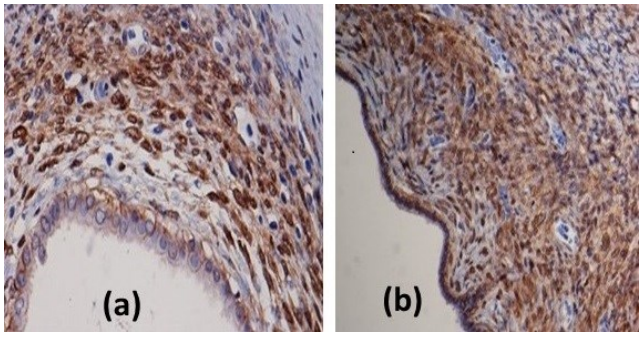

Figure-4: Immunohistochemistry highlighting the ER (a) and PR (b) positive stromal cells

DISCUSSION

Mucinous cystic neoplasms of the liver are rare cystic tumors that occur within the liver parenchyma, or less frequently, in the extrahepatic bile ducts.[16,17] They are reported to account for < 5% of all liver cysts with predominance in females during fourth to fifth decade of life.[12] Clinical presentation of patient with MCN is largely vague ranging from asymptomatic to abdominal pain, abdominal fullness, early satiety, jaundice, or weight loss.[18] On ultrasonography, an MCN typically appears as a hypoechoic lesion with thickened, irregular walls and occasional internal echoes representing debris and wall nodularity. On a CT scan, an MCN appears as a low attenuation mass, which may be uni- or multilocular or may have septations. The cyst wall is usually thickened and/or irregular. Magnetic resonance imaging (MRI) with contrast enhancement remains the best modality for characterization and surgical planning. MCNs typically appear with loculations and septations.[19] The three imaging features that accurately differentiated MCNs from benign hepatic cysts were (a) the presence of a solid enhancing nodule (100% specific), (b) lack of septations arising from external macrolobulations, and (c) the presence of a solitary lesion.[20]

The WHO classification (2010) has defined MCN-L as a counterpart of MCN of the pancreas (MCN-P),[21] comprising of a multiloculated cystic tumour lined by mucus-secreting cuboidal or columnar epithelium with septae, cellular (mesenchymal) stroma resembling ovary, beneath the epithelium, usually showing no communication with the bile duct. The presence of mural nodules and papillary projections is considered to be evidence of malignancy.[21,22]

Shiono et al. reported MCN in various organs (pancreas, liver, spleen, mesentery), with ovarian-like stroma as the common histopathological feature.[23] The common differential diagnosis of MCN-L is tabulated in Table-2 with mention of the diagnostic features. Immunohistochemistry plays a role in diagnosis of MCN-L by demonstration of expression of ER and PR in ovarian type stroma. However, role of immunohistochemistry in differentiation of benign from malignant MCN has been guarded, with expression of MUC5A in borderline to invasive malignant MCN, as seen in IPNB.[24] The expression of CDX2, goblet-cell specific MUC2, MUC5AC, and CK20 in the dysplastic epithelium of MCN, while absence in benign lesion was further correlated with KRAS mutations. The latter were linked with late malignant changes in MCN.[25]

Table-2: Common differential diagnosis of MCN-L

S. No.	Diagnosis	Features
1	Simple cyst	Absence of septations and papillary projections, presence of serous cystic fluid
2	Echinococcal cysts	Calcifications, positive serology
3	Intraductal papillary neoplasm of the bile duct (IPNB)	Communication with the bile ducts, bile duct dilatation, and papillary projections in the bile ducts

The consensus on the treatment of hepatobiliary MCN has been complete surgical resection.[26] The recurrence rate is reported to be high in patients with partial surgical resection, percutaneous aspiration, surgical fenestration, or marsupialization.[19]

CONCLUSIONS

MCN of liver are rare neoplasms presenting with vague clinical symptoms and causing radiological dilemmas with ovarian neoplasm in females. Histopathology remains the gold standard for diagnosis of MCNs with presence of ovarian type stroma as a mandatory criterion. Malignancy in these is rare occurrence and complete surgical removal treatment of choice.

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