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Case Report

Pathology Section

Mucinous Cystic Neoplasm of the Liver: A Case Report

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ABSTRACT

Mucinous Cystic Neoplasms (MCNs) of the hepatobiliary tract constitute <5% of all hepatic cysts. Present case report is of MCN of the liver with radiological dilemma and special emphasis on histopathology combined with immunohistochemistry. The present case is a 36-year-old female who presented with pain in the upper abdomen for five months. The pain was insidious in onset, mild in intensity, dull aching, and non radiating. There was a history of intermittent loose stools. Routine laboratory investigations such as complete blood count, liver function tests, and renal function tests were within normal limits. Viral markers including Human Immunodeficiency Virus (HIV), Hepatitis B Surface antigen (HbsAg), and anti-HCV were negative. Ultrasound abdomen showed a well-defined cystic lesion measuring 6.5×4.8 cm in the left lobe of the liver (segment II/III) with thin internal echoes. A radiological diagnosis of hydatid cyst was made. Laparoscopic deroofing of the cyst with drainage was performed, and the specimen was sent for histopathological examination. Grossly, the specimen consisted of grey-brown tissue fragments measuring 6.5×3 cm. On microscopic examination, the cyst was lined by cuboidal to columnar epithelium with mucinous cytoplasm and basally oriented nuclei. The subepithelium showed ovarian-like stroma. A diagnosis of benign MCN was made on histopathology. Immunohistochemistry using Oestrogen Receptor (ER) showed strong nuclear positivity in the stromal cells, confirming the diagnosis. The postoperative period was uneventful, and the patient improved symptomatically.

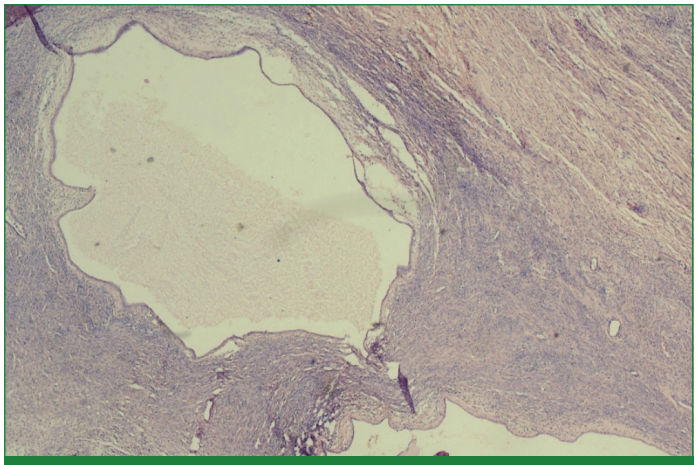
Keywords: Cystic lesions, Mucin secreting tumours, Ovary like stroma

CASE REPORT

A 36-year-old female presented with pain in the upper abdomen for five months. The pain was insidious in onset, mild in intensity, dull aching, and non radiating. She also reported intermittent loose stools. There was no history of vomiting, fever, jaundice, or loss of weight. No associated co-morbidities were noted. Routine laboratory investigations such as complete blood count (Hb- 11.8 g/dL, total leukocyte count- 9,900 cells/mm^3, platelet count- 1.65 lakh/mm^3), liver function tests (total bilirubin- 0.40 mg/dL, direct bilirubin- 0.13 mg/dL, Aspartate Aminotransferase (AST)- 55 U/L, Alanine Aminotransferase (ALT)- 41 U/L, AST/ALT ratio- 1.36), and renal function tests (blood urea- 13 mg/dL, serum creatinine- 0.41 mg/dL) were within normal limits. Viral markers such as HIV, HBsAg, and anti-HCV were negative.

Ultrasound abdomen revealed a well-defined cystic lesion measuring 6.5×4.8 cm in the left lobe of the liver (segment II/III) with thin internal echoes. The radiological diagnosis suggested a liver hydatid cyst. Laparoscopic deroofing of part of the cyst with drainage was performed, and the excised specimen was submitted for histopathological evaluation. Grossly, the specimen consisted of a grey-brown tissue fragment measuring 6.5×3 cm [Table/Fig-1]. No papillary excrescences, solid areas, or cyst contents were identified. Microscopic examination revealed a cyst lined by cuboidal to columnar epithelium with mucinous cytoplasm and basally located nuclei [Table/Fig-2]. The subepithelium showed closely arranged spindle-shaped cells with elongated nuclei and cytoplasm outlined by collagen fibres, resembling ovarian-type stroma [Table/Fig-3]. A diagnosis of benign MCN of the liver was made. Immunohistochemistry for ER showed strong nuclear positivity in the stromal cells, confirming the diagnosis [Table/Fig-4]. The postoperative course was uneventful, and the patient showed symptomatic improvement.

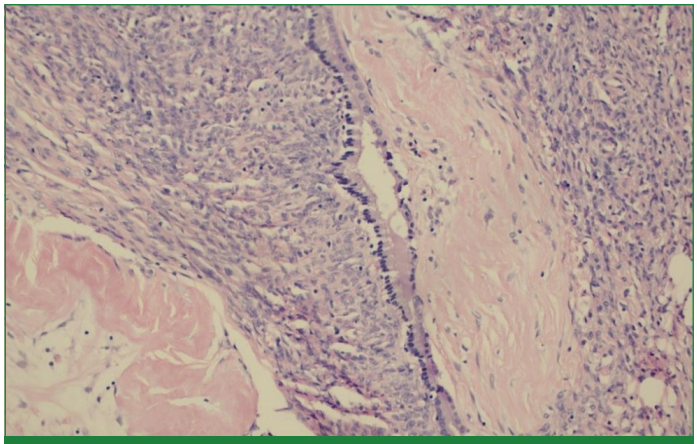
[Table/Fig-1]: Gross appearance- Consists of grey brown tissue fragment measuring 6.5×3 cm. No solid areas or papillary excrescences noted.



[Table/Fig-2]: Section show a cyst lined by cuboidal to columnar epithelium with mucinous cytoplasm and basally oriented nuclei (H&E,4x).

DISCUSSION

The MCNs of the hepatobiliary tract constitute <5% of all hepatic cysts [1]. According to the 5th edition of the World Health Organisation (WHO) classification, MCN-L is a rare benign entity with an incidence



[Table/Fig-3]: Section show basally oriented nuclei with subepithelial ovarian type stroma (H&E, 40x).

[Table/Fig-4]: Immunohistochemistry using Oestrogen Receptor (ER) highlighting ovarian like stroma which showed strong nuclear positivity (IHC, 4x).

of one case per 20,000-100,000 persons. Previously, it was referred to as cystadenoma or cystadenocarcinoma of the liver or extrahepatic bile ducts. MCN-L is a cyst-forming epithelial neoplasm composed of mucus-secreting epithelial cells and is characterised by ovarian-type stroma, which distinguishes it from other cystic lesions of the liver. Furthermore, the International Classification of Diseases for Oncology classifies MCNs as MCN with low-grade intraepithelial neoplasia, MCN with high-grade intraepithelial neoplasia, and MCN with associated invasive carcinoma [2]. The potential for malignant transformation ranges from 3-6% [3]. Some studies suggest that ovarian-like stroma in MCN-L may be due to the close proximity of the liver and gonads during embryonic development, which may allow transfer of gonadal cells to the hepatic surface, resulting in ovarian-type stroma [4].

MCN-L predominantly affects middle-aged women between 40 and 70 years of age, although rare cases in teenagers have been reported [5]. In the present case, the patient was a middle-aged female. Ultrasound and computed tomography imaging typically demonstrate a multilocular fluid-filled cyst. The cyst wall may reveal smaller cysts, septations, calcifications, and less commonly, surface nodules. Approximately 76-79% of cases occur in the left hepatic lobe, with a predilection for segment IV [6]. Radiologically, present case showed a cystic lesion with thin internal septations in the left lobe of the liver, similar to a case reported by Anu D et al., [7].

The differential diagnoses of MCN-L mainly include Intraductal Papillary Neoplasm of the Bile duct (IPNB) with cystic change, simple hepatic cyst, and echinococcal cyst. Findings in IPNB include communication with the bile ducts, ductal dilatation, and papillary projections within the bile ducts. In echinococcal cysts, calcification is commonly present and serology is usually positive. Simple cysts contain clear fluid with absence of septations or papillary projections [Table/Fig-5] [8,9].

Parameters	Soni S et al., [8] 2021	Ha SW et al., [9] 2022	Index case, 2025
Age (years)/ sex	55/ Female	32/Female	36/Female
Radiological findings	Unilocular cyst in the hilum of the liver-suspicious of hydatid cyst. Mild intra hepatic biliary radicles dilatation.	Multiloculated cystic lesion, pedunculated at liver capsule-right lobe of liver	Multiloculated cyst in the left lobe of the liver. No bile duct involvement
Treatment	Laparoscopic deroofing of the cyst	Resection with partial hepatectomy	Laproscopic deroofing of the cyst
Outcome	Postoperative period uneventful. Asymptomatic	Postoperative period uneventful. Asymptomatic.	Postoperative period uneventful. Asymptomatic.

[Table/Fig-5]: Literature review of comparing clinical and radiological features of reported cases and present case of MCN-L [8,9].

Grossly, MCN-L appears as a well-circumscribed cyst with a thick fibrous capsule, clearly demarcated from the surrounding liver parenchyma. These lesions do not communicate with the biliary system. Features that favour malignancy include loss of capsular integrity, presence of solid areas, and papillary excrescences on cut section. In present case, the cyst was multiseptated, thin-walled, and showed no solid areas or papillary excrescences.

Microscopically, the cyst wall consists of three distinct layers. The inner layer is lined by columnar or cuboidal epithelial cells, with occasional squamous-like cells. The middle layer is the characteristic subepithelial ovarian-type stroma, composed of closely arranged spindle-shaped cells with elongated nuclei and cytoplasm outlined by collagen fibres. The outermost layer forms a fibrous capsule [2,9]. On immunohistochemistry, the ovarian-like stroma of MCN-L shows positivity for ER, progesterone receptor (PR), and WT1 [1]. In present case, the stroma exhibited strong nuclear positivity for ER.

Resection is the main treatment modality for cystadenoma whenever possible, as malignant transformation of the lining epithelium may occur with time. Partial excision of cystadenocarcinoma or cystadenoma is associated with recurrence and poorer prognosis [10]. In present case, as the radiological diagnosis suggested hydatid cyst, only partial deroofing of the cyst with drainage was done due to the large size of the lesion.

CONCLUSION(S)

MCNs of the hepatobiliary tract are rare tumours compared to MCNs of the pancreas and usually have an indolent clinical course. Due to the rarity of these tumours, there remains limited understanding regarding disease aetiology, pathogenesis, and prognosis. Preoperative evaluation of cystic liver tumours should include MCN as a differential diagnosis. Complete surgical resection remains the mainstay of treatment.

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