Mucinous Cystic Neoplasm of the Liver: Case Report and Review of the Literature

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Abstract

Mucinous cystic neoplasms of the liver (MCN-L) are a rare benign neoplasm of the biliary system that grows slowly with vague clinical symptoms. The malignant transformation in these tumors is high, and preoperative diagnosis using radiography or endoscopy is challenging. We present a case report of a 58-year-old female patient who presented with diffuse abdominal pain and nonprojectile vomiting. Abdominal ultrasound revealed characteristics that point to a hydatid cyst. The enhanced computed tomography revealed a well-defined multiloculated thin-walled fluid density lesion in the left lobe of the liver. After a left hemi-hepatectomy, a multiloculated cystic lesion in the liver with mucinous material as the content was discovered. Histopathology showed multiple cystic spaces lined by mucin-secreting columnar epithelium without nuclear atypia, the wall of the cysts showed ovarian-like stroma, and the diagnosis of MCN-L was made. There was no recurrence after 6 months of regular follow-up.

Keywords

► biliary cystadenoma
► liver cyst
► mucinous cystic neoplasm
► ovarian-like stroma

Introduction

Mucinous cystic neoplasms of the liver (MCN-L) are a rare disease with an ovarian-type stroma present and are slow-growing neoplasms that have a benign course. They were earlier termed as biliary cystadenocarcinoma and bile duct/biliary cystadenoma. But in 2010, the World Health Organization (WHO) reclassified and divided this type of lesion into noninvasive and invasive categories. It is now described 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.”¹

Surgical excision is the preferred course of treatment for both noninvasive and invasive MCN; however, resectability varies according to the tumor’s anatomic location, functional liver reserve, and comorbidities. Nevertheless, because of its uncommon presentation, it is frequently misdiagnosed.

Case Report

A 58-year-old female presented with complaints of pain in the abdomen and vomiting for 1 month. The abdominal pain was diffuse, dull, and nonradiating; the vomiting was nonprojectile, nonbilious, and contained undigested food particles. Abdomen examination revealed a nontender, enlarged liver in the right hypochondrium, palpable 8 cm inferior to the right subcostal margin and 2 cm lateral to the midline. There was no evidence of ascites. Her past history was uneventful.

Laboratory investigations revealed low hemoglobin, which was 10.4 mg/dL. Liver function tests were within normal limits and serology for hepatitis B virus was negative. The serum marker cancer antigen (CA) 19–9 was elevated, while α-fetoprotein and carcinoembryonic antigen levels were within the normal range.

Ultrasonography of the abdomen revealed an enlarged liver with a multilobulated cystic lesion measuring 12.2 cm by
The lesion showed multiple enhancing septations and loculated cyst measuring 7.5 cm in diameter. The gallbladder was unremarkable.

The postoperative course was uneventful. A specimen left hepatectomy and a specimen was sent for histopathological evaluation. The section showed multiple enhancing septations and calcifications along the septa and capsule. No solid-enhancing component was noted (Fig. 1).

The patient underwent a left hepatectomy and cholecystectomy, and a specimen was sent for histopathological evaluation. The postoperative course was uneventful. A specimen left hepatectomy with a gallbladder measuring 20 cm in diameter was received, and the gallbladder measured 6 cm in diameter by 1 cm. The cut surface of the liver showed a large multiloculated cyst measuring 7.5 cm by 7 cm. The cyst contained mucinous material (Fig. 2). The gallbladder was unremarkable.

Microscopy of the liver showed multiloculated cysts lined predominantly by mucinous epithelial cells having basal nuclei and supranuclear vacuolations; at places, the cyst wall is lined by denuded, low-cuboidal to flattened epithelium. The subepithelium showed thick, densely packed, bland spindle-shaped cells, much like an ovarian stroma. On immunohistochemistry, the ovarian stroma will be positive for estrogen receptor, progesterone receptor, and inhibin A.

8.0 cm that was noted to arise from the left lobe of the liver and was suggestive of a hydatid cyst. The enhanced computed tomography (CT) of the liver, gallbladder, and spleen revealed a well-defined multiloculated thin-walled fluid density lesion measuring 14.4 cm by 14.2 cm by 9.1 cm noted in the left lobe of the liver. The lesion showed multiple enhancing septations and calcifications along the septa and capsule. No solid-enhancing component was noted (Fig. 1).

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Histology of the MCN-L shows cyst lined by mucus-secreting columnar, cuboidal, and flattened epithelial cells which are grouped in a single, flat row with sporadic polyploid or papillary projections on the inner face of the cysts. Beneath the epithelium has hypercellular stroma encircled by a thick layer of fibrous collagenous tissue. The mesenchyme had an oval nucleus and a compact arrangement of spindle-shaped cells, much like an ovarian stroma. On immunohistochemistry, the ovarian stroma will be positive for estrogen receptor, progesterone receptor, and inhibin A.

Discriminating features from classical MCNs is the hormone-secreting capability of MCN-L. There were no signs of recurrence until 2 years. The patient is doing well without any complaints. There was no evidence of recurrence of tumor.

Discussion

MCN-L are extremely rare benign cystic tumors that can arise in the extrahepatic bile ducts or the liver parenchyma accounting for <5% of all hepatobiliary cystic neoplasms. These predominantly occur in adult females. The WHO classification of 2010 defined MCN-L as a counterpart of MCN of the pancreas (MCN-P). Similar to MCN-P, MCN-L is a multiloculated cystic tumor with a pathognomonic hormone receptor–positive ovarian-like stroma. Whether it represents an ovarian remnant, or a recapitulation of periductal fetal mesenchyme seen in these organs, or some other phenomenon is not known.

The majority of individuals clinically presents with vague abdominal symptoms and vomiting. Imaging modalities like CT scan/magnetic resonance imaging aid in the clinical diagnosis. The MCN-L displays as a low attenuation mass that can be uni- or multilocular, with or without septations. The cyst walls are typically thickened or irregular. The radiological differential diagnosis includes simple cyst, echinococcal cyst, intrahepatic cholangiocarcinoma with cystic change, and intraductal papillary neoplasm of the bile duct (IPNB). However, it is difficult to differentiate MCN-L and cyst-forming IPNB. In such cases, the presence of ovarian-like stroma on biopsy is essential for the diagnosis of MCN-L. Echinococcal cysts are frequently associated with calcifications and positive serology. Simple cysts contain serous cystic fluid and it lacks septations and papillary projections.

The laboratory investigations usually show elevated levels of serum markers CA 125 and CA 19–9. In our case no abnormalities were seen, with the exception of elevated CA19–9 level; however, recent study found no statistical support for this correlation. Complete surgical resection is the usual course of treatment for MCNs, other treatments like surgical fenestration and sclerotherapy are also available. The percutaneous aspiration has a high recurrence rate.

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Fig. 1 Computed tomography (CT) image showing a thin-walled cystic lesion with fluid density with septations.

Fig. 2 Left hepatectomy specimen with multiloculated cystic lesion containing mucin.
The epithelium of MCN shows positivity for CK7, CK19, CK8, and CK18 (biliary) or mucicarmine (mucinous areas).² ⁷,⁸

The degree of architectural complexity and cytologic atypia of the lining epithelium determine whether the noninvasive lesions are categorized as low or high grades. While neoplasms without ovarian-like stroma have a poor prognosis, those with this type of stroma are thought to be benign with malignant potential.⁸ Following complete surgical resection, the prognosis for MCN-L with extrahepatic involvement is excellent. Since they may be malignant, incomplete resection can result in local recurrence and malignant transformation.

**Conclusion**

MCN-L is a rare, slow-growing tumor characterized by an ovarian-like stroma in women. It has the potential to become malignant. The best course of action for preventing malignant transformation and recurrence is a complete surgical resection.

**Note**

The manuscript has been read and approved by all the authors, that the requirements for authorship have been met, and each author believes that the manuscript represents honest work.

**Conflict of Interest**

None declared.

**References**


**Fig. 3** (A) Histology showing multiloculated cystic spaces (hematoxylin and eosin [H&E], ×100). (B) Cysts are lined by mucin-secreting epithelium with underlying ovarian-like stroma (H&E, ×200). Immunohistochemical (IHC) study showed nuclear expression of (C) estrogen receptors, ×100; (D) progesterone receptors, ×100.