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Case Report

A Rare Tumor of the Liver: Mucinous Cystic Neoplasm

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Abstract

Mucinous cystic neoplasms (MCNs) are rare hepatic lesions with malignant potential, most commonly occurring in middle-aged women. Due to the lack of specific diagnostic tests and pathognomonic radiologic findings, establishing a preoperative diagnosis is challenging. The recommended primary treatment is complete surgical resection, while definitive diagnosis is typically made through histopathological evaluation.

A 24-year-old female patient, initially operated on with a preoperative diagnosis of hydatid cyst, was found on imaging to have a 9 × 4.5 cm cystic mass predominantly located in segment 4B of the liver, with partial extension toward segment 5. The patient underwent a left hepatectomy. Her postoperative course was uneventful, and histopathological analysis revealed a low-grade MCN.

There is limited information in the literature regarding MCNs of the liver. Accurate management of these patients is crucial due to their potential association with invasive carcinoma. Therefore, this entity should be considered in the differential diagnosis of hepatic cystic lesions, and curative surgical resection should be pursued whenever feasible.

Keywords: Mucinous Cystic Neoplasm, Liver, Ovarian-Like Stroma

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Mucinous cystic neoplasms (MCNs) of the liver are rare tumors that typically occur in middle-aged women, accounting for less than 5% of all hepatic cysts.[1,2] Although these tumors are generally benign, they carry a 3–5% risk of malignancy, which creates challenges in both diagnosis and management.[3] The exact origin of MCNs remains uncertain; however, histopathological studies suggest a possible association with ectopic ovarian-like stroma within the liver.[4]

Current knowledge about MCNs is primarily derived from case reports and small-scale studies, leaving no clear consensus on their optimal diagnosis and treatment strategies. Given their rarity and malignant potential, accurate clinical management is essential. In this report, we present the diagnostic and therapeutic management of a 24-year-old female with a hepatic MCN.

Case Report

A 24-year-old female patient who presented to another centre two months ago with complaints of abdominal pain and bloating was found to have a mass consistent with a hydatid cyst in the left lobe of the liver during the examinations performed. The patient underwent a partial cystectomy, and her postoperative course was uneventful.

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Histopathological examination of the surgical specimen removed from the cyst wall revealed it to be a MCN without dysplasia or invasive malignancy. The patient was then referred to us.

The patient had no significant comorbidities, and abdominal examination was unremarkable. Laboratory tests, including liver function tests and tumor markers, were within normal limits. Contrast-enhanced computed tomography and magnetic resonance imaging revealed a lobulated, multilocular cystic mass with thin septations, predominantly located in segment 4B of the liver, with partial extension into segment 5, measuring 9 × 4.5 cm at its widest dimension (Fig. 1).

A left hepatectomy was performed, ensuring tumor-free surgical margins. The postoperative period was uneventful. Histopathological examination of the hepatectomy specimen revealed MCN containing low-grade dysplasia measuring 9x7x4 cm (Fig. 2). The patient was subsequently placed under follow-up by the medical oncology department without adjuvant therapy (Fig. 3).

Discussion

MCNs were previously classified as biliary cystadenomas or cystadenocarcinomas. In 2010, the World Health Organization (WHO) reclassified these mucin-producing biliary tumors into two distinct entities: MCNs and intraductal papillary mucinous neoplasms (IPMNs) of the bile duct.[5] Hepatic IPMNs are considered the biliary counterpart of pancreatic IPMNs. Unlike MCNs, IPMNs tend to communicate with the bile ducts, are slightly more common in males, and typically lack ovarian-like stroma.

Histopathologically, MCNs are cyst-forming epithelial tumors lined by mucin-producing cuboidal or columnar epithelium, usually without communication with the bile ducts, and characterized by the presence of ovarian-like stroma. According to the WHO classification, the presence of ovarian-like stroma is a defining diagnostic criterion for MCNs. These tumors can exhibit low-, intermediate-, or high-grade dysplasia and, in some cases, progress to invasive carcinoma.

Nearly all MCNs occur in female patients and are most commonly located in the left lobe of the liver.[6] Our patient is similarly a female patient with a left lobe lesion. Interestingly, a significant proportion of cases originate from segment 4, as observed in our case, although the exact reason for this remains unclear. The average age at presentation is typically between 40 and 70 years, but, as in our case, MCNs

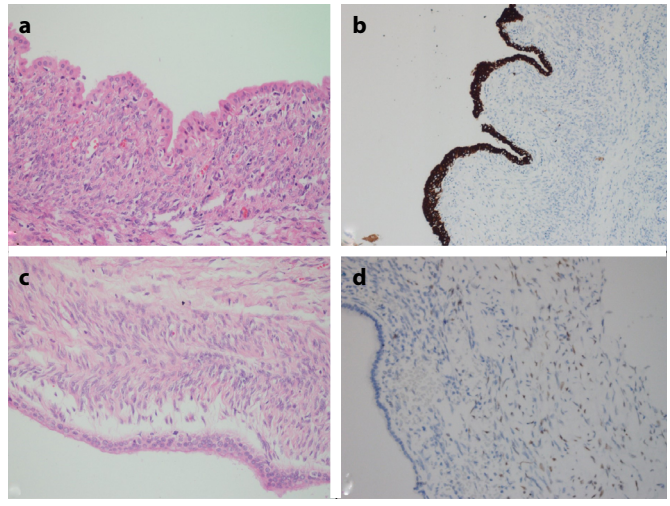

Figure 2. (a) Cyst surface surrounded by single-layered cuboidal cells (HE; 200X). **(b)** CK7 positivity in single-layered cuboidal cells (CK7; 200X). **(c)** Ovarian stroma with a spindle cell appearance beneath the epithelium (HE; 200X). **(d)** Estrogen receptor positivity in the ovarian stroma area (Estrogen receptor; 200X).

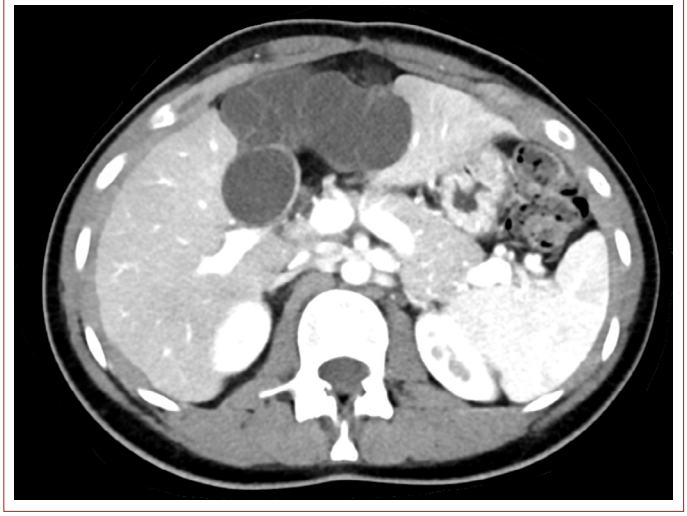

Figure 1. Preoperative septated cystic lesion in liver segment 4B-5.

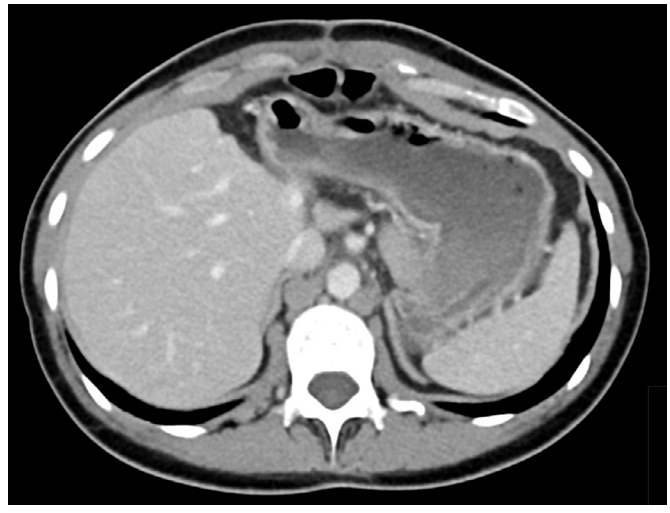

Figure 3. The post-operative CT scan at the 3-month follow-up.

may also present in younger patients, with only a few cases reported in individuals under 25.

In their early stages, MCNs are usually asymptomatic, and diagnosis often occurs when the lesions enlarge to approximately 10 cm.[7] When symptomatic, patients most commonly present with epigastric pain, abdominal fullness, and anorexia. Depending on the tumor's location, obstructive jaundice may also occur. Laboratory findings are often unremarkable unless there is biliary obstruction or communication with the bile ducts, and serum CA19-9 levels may be elevated. Our patient had abdominal pain and bloating, but no biliary problems, and her CA 19-9 was within normal limits.

On imaging, MCNs typically appear as multiloculated cystic tumors with septated walls that do not communicate with the bile ducts.[8] Due to these nonspecific imaging characteristics, they are frequently misdiagnosed as simple hepatic cysts, hydatid cysts, liver abscesses, or cholangiocarcinomas. In our patient, the lesion was initially misdiagnosed at another center, leading to an incomplete resection and potential risk of recurrence.

Definitive diagnosis relies on histopathological evaluation. The hallmark feature of MCNs is the presence of mucin-secreting biliary-type epithelium accompanied by dense subepithelial ovarian-type stroma that expresses female sex hormone receptors.

Although rare, MCNs carry a risk of malignant transformation. Due to the limited number of cases reported, the incidence of invasive carcinoma has been reported to range between 2% and 15.4%.[7,9] However, the absence of specific radiological features makes distinguishing benign from malignant lesions challenging.[10]

The recommended primary treatment for these tumors is curative surgical resection, aimed at preventing recurrence and malignant transformation.[11] Recurrence rates are high after partial resection (up to 80%). Prognosis largely depends on the presence or absence of invasion. In benign lesions, five-year survival after complete resection approaches 100%, with 18-year survival rates around 90%, whereas in cases associated with invasive carcinoma, five-year survival declines to 65–70%.[12,13]

Conclusion

In conclusion, MCNs of the liver are rare lesions with an uncertain etiopathogenesis and no specific diagnostic tests. Due to the limited number of reported cases, data regarding optimal management are scarce. Given their malignant potential, increasing awareness of these lesions and reporting more cases are essential to improve diagnostic accuracy and therapeutic strategies.

Disclosures

Informed consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Conflict of Interest: None declared.

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