Journal of Gastrointestinal Surgery (2019) 23:874–876
https://doi.org/10.1007/s11605-018-3768-z

Biliary Mucinous Cystic Neoplasm: a Classic Presentation of a Rare Neoplasm

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Received: 23 March 2018 / Accepted: 2 April 2018 / Published online: 20 April 2018

Abstract
Biliary mucinous cystic neoplasms are rare parenchymal neoplasms with a considerable malignant potential. Due to a lack of diagnostic imaging criteria, histopathologic evaluation remains the definitive method of diagnosis. Resection is the treatment of choice. Here, the authors present a case of biliary mucinous neoplasm in a 39-year-old female with the associated radiographic and histopathologic findings.

Keywords Biliary mucinous cystic neoplasm · Biliary cystadenoma · Mucinous cystic neoplasm with invasive carcinoma

Clinical Case

Thirty-year-old female presents with year-long history of intermittent epigastric pain and more recent onset of jaundice. CT and MRCP reveal a 3.9-cm multiseptated cystic lesion in the left lobe. Upstream dilation of the left bile duct and lithiasis within the left and common bile ducts were also noticed. ERCP imaging confirms MRCP findings (Fig. 1). CA19-9 level was elevated at 416 U/mL (reference range < 36 U/mL). A left hepatectomy was performed (Fig. 2). Histopathology (Fig. 3) was consistent with biliary mucinous cystic neoplasm.

Discussion

Biliary mucinous cystic neoplasms (MCN) are rare neoplasms that comprise less than 5% of all hepatic biliary cystic lesions with a preference toward middle-aged women. Clinically, MCNs can present with a wide variety of symptoms depending on the size and location. While considered benign, MCNs carry a risk for malignant transformation to mucinous cystic neoplasms with associated invasive carcinoma, as high as 30% according to some reports.[1,3] Despite significant advances in imaging, and given the rarity of the disease, no imaging criteria have been validated that can reliably differentiate the benign lesions from the carcinomas. That said, MCNs and MCN with associated invasive carcinomas both tend to be large, well-defined, solitary lesions with thick fibrous capsules and internal septations. Reports suggest the carcinomas are usually more associated with the presence of nodular septations, mural nodules, and calcifications along the wall or septum of the lesion.[2] Thus, histopathology remains the definitive diagnostic modality.

Given the aforementioned lack of reliable pre-operative imaging and risk for malignant transformation, the treatment of choice for MCNs with or without associated invasive carcinoma remains surgical resection. The extent of resection, namely enucleation versus formal hepatectomy, remains a subject for debate with various methods and degrees of resection discussed in the literature. While outcomes following surgical resection are good, it should be noted that incomplete resection is associated with recurrence, albeit rare, and a worse prognosis.[1,3]

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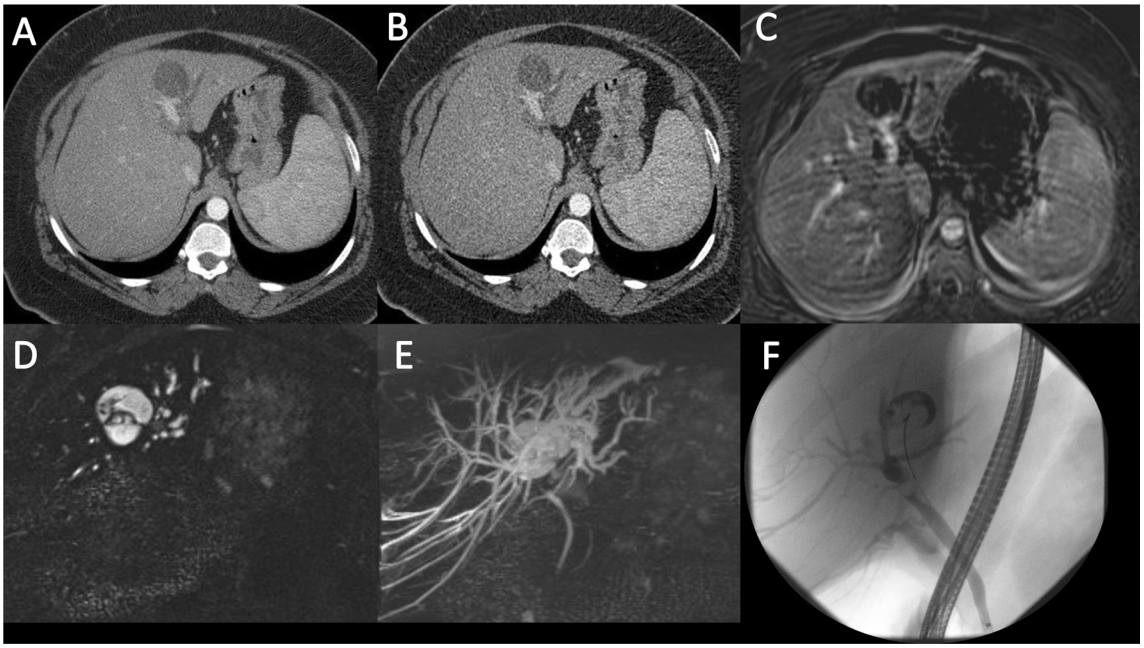

Fig. 1 Arterial phase CT (**a**, **b**) showing left lobe lesion relative to portal vein. Axial (**c**) and T2-weighted coronal (**d**) MRI images. MRCP (**e**) and ERCP (**f**) visualization of defect

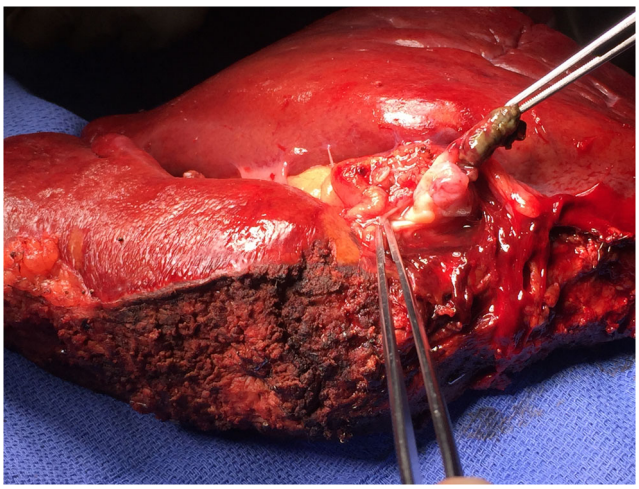

Fig. 2 Surgical specimen with a tumor, measuring 3.2 × 3 × 2.1 cm, highlighted by instrument

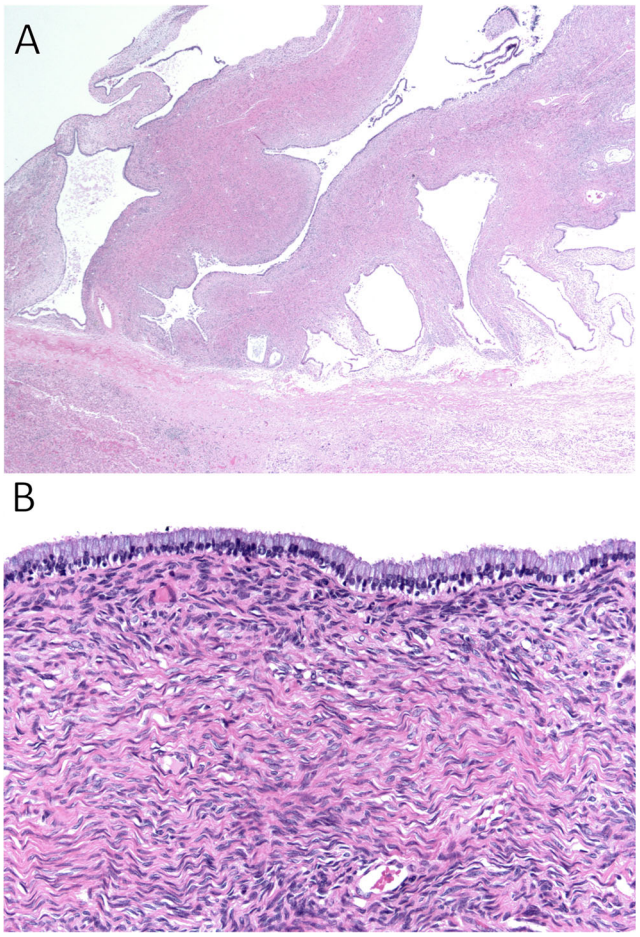

Fig. 3 Histopathological evaluation: **a** Multilocular cystic lesion lined by mucinous epithelium with basally located nuclei (H & E, × 100), and **b** high magnification demonstrates the characteristic ovoid cytological bland cells of the typical ovarian stroma diagnostic of these lesions (H & E, × 200)

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflicts of interest.

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