An unusual cystic liver lesion (ECR 2019 Case of the Day)

Clinical History:

A 69-year-old female patient with suspicion of cystic echinococcus recurrence in the liver with segmental biliary obstruction presented to the ER with cholestasis, leukocytosis and elevated CRP. She received a partial cystectomy and drainage operation for a liver cyst located in the left lateral segment 9 years ago.

Imaging Findings:

Fig 1. A mixed echogenic solid mass with unechogenic cystic components on the left liver lobe is seen on the preoperative ultrasound examination. Patent middle hepatic vein is seen (as blue) next to the mass on colour Doppler image. A tiny spot of colouration (seen as red) suggests that the mass is vascular.

Fig 2a. Coronal T2W MR image; a large mass that has nodular hypointense-solid areas with hyperintense fluid areas is filling the atrophic left liver lobe. There are tubular fluid-filled structures suggestive of dilated biliary ducts in the atrophic left liver lobe (between the lesion and stomach). Right liver lobe contours are regular.

Fig 2b. Axial T2W MR image; the lesion abuts the central portion of the left liver lobe biliary duct. Notice the dilated biliary ducts in the upstream areas secondary to ductal obstruction by the mass forming intraductal tumour.

Fig 3a. Coronal MRCP image; fluid component of the nodular mass and adjacent fluid-filled tubular structures suggestive of dilated biliary ducts can be seen.

Fig 3b. MRCP image; notice the dilated extrahepatic bile duct.

Fig 4a. Diffusion-weighted image; nodular restricted diffusion area is seen as hyperintense and it is hypointense on the ADC map (Fig 4b).

Fig 5a,b. Axial pre-contrast fat saturated T1W vibe sequence; a large hypointense mass on the left liver lobe is depicted.

Fig 6. Axial post-contrast fat saturated T1W vibe sequence; solid components of the lesion show some contrast enhancement (Fig 5). The arching middle hepatic vein (Fig 6a) and obliterated left portal vein (Fig 6b) are seen.

Fig 7. Intraoperative ultrasound confirms the preoperative findings.
Fig 8. Postoperative ex-vivo T2W MR image of the left lobectomy specimen reveals heterogenous mixed intensity nodular lesion with smooth contours. Adjacent liver parenchyma is atrophied and filled with dilated biliary ducts.

Fig 9. The macroscopic section of the pathologic specimen have shown similar findings with the ex-vivo MR image.

Discussion:

Cystic liver lesions are being encountered with increasing frequency due to the development of cross-sectional modalities, especially MRI. Intraductal papillary neoplasm of the liver is a rare cystic lesion that consists of the papillary growth of atypical epithelial cells with oncocytic changes. Non-tumorous intrahepatic bile ducts near the lesion may show epithelial dysplasia and contain a lot of mucus[1,2,3,4].

The patient was operated on 9 years before with a presumed diagnosis of cystic echinococcosis of the liver. After partial cystectomy and drainage, the surgeon noticed an increased viscosity of the cyst fluid and sampled it intraoperatively. The pathologist could neither find daughter cysts nor cestodes in the aspirate. The final diagnosis could deny the lesion as echinococcus cyst but was not ultimately diagnostic.

Although the lesion appeared cystic on T2 weighted images, contrast enhancement in the intracystic components and absence of a fibrotic pericyst raised the suspicion of other cystic entities rather than echinococcus granulosus[5]. The presence of portal and left hepatic vein occlusion/invasion by the lesion, puts echinococcus alveolaris in the differential diagnosis [6]. However, E. alveolaris was also unlikely due to the sharp margins of the lesion and the lack of calcification. Taken together, the presence of contrast enhancement in the cyst cavity, connection to the biliary system, ectasia of the biliary ductal system near the lesion and presence of malignant features such as vascular occlusion/invasion and parenchymal atrophy suggested a malignant process. Hence cystadenocarcinoma and intraductal papillary biliary neoplasm were included in the differential diagnosis. A transabdominal US examination confirmed all observations made by MRI.

After prophylactic albendazole treatment, a laparotomy was made. Upon the radiological suggestion of a likely malignant cystic lesion, the surgeon aspirated the cyst and verified the presence of mucin within the cavity. Hence a left hepatic lobectomy was performed without contaminating the peritoneal cavity with cyst contents to avoid dissemination. The final pathology report confirmed the diagnosis of intraductal-oncocytic papillary biliary neoplasm with malignant transformation. The lesion was resected with clear (R0) margins.

In summary, the bilateral information sharing between the clinicians and radiologists is of utmost importance for proper diagnosis and treatment of extraordinary cases. The presence of contrast enhancement within a cyst should always be regarded as a warning sign for a malignant lesion.

Differential Diagnosis List: Cystic echinococcosis recurrence, Liver abscess, Alveolar echinococcosis, Cystadenocarcinoma, Intraductal oncocytic papillary biliary neoplasm

Final Diagnosis: Intraductal oncocytic papillary biliary neoplasm

References:


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Origin: Department of Radiology, Koç University Hospital, Istanbul, Turkey, 2018
Description: Coronal T2W MR image; a large mass that has nodular hypointense-solid areas with hyperintense fluid areas is filling the atrophic left liver lobe. There are tubular fluid-filled structures suggestive of dilated biliary ducts in the atrophic left liver lobe (between the lesion and stomach). Right liver lobe contours are regular. Origin: Department of Radiology, Koç University Hospital, Istanbul, Turkey, 2018
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**Description:** The macroscopic section of the pathologic specimen have shown similar findings with the ex-vivo MR image. **Origin:** Department of Pathology, Koç University Hospital, Istanbul, Turkey, 2018