Case Report

A case of peribiliary hepatic cysts in a cirrhotic liver: A mimicker of Klatskin Tumor

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ABSTRACT

Patients with end-stage liver disease may present to healthcare facilities with features of obstructive jaundice and a picture of hilar cholangiocarcinoma on radiological imaging. Careful observation and knowledge of the presence and higher prevalence of peribiliary hepatic cysts in a cirrhotic liver can aid in differentiating this benign entity from malignancy that may halt or delay the patients’ eligibility for receiving a liver transplant. We present a case of a patient with liver cirrhosis initially diagnosed as Klatskin tumor on imaging then as a simple case of multiple peribiliary hepatic cysts with the patient eventually undergoing successful liver transplantation.

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Introduction

Obstruction of the intrahepatic extramural periductal glands can lead to peribiliary hepatic cyst formation [1,7]. Peribiliary hepatic cysts are seen exclusively at the hepatic confluence and on both sides of the larger portal tracts [2]. Nevertheless, they can uncommonly also be found around the extrahepatic bile ducts [9]. Peribiliary hepatic cysts show no direct communication with the biliary tree and are progressive in nature showing a gradual increase in number and size over time [1,2,3]. These thin walled round cysts contain serous fluid and usually face the hepatic parenchyma in their peculiar distribution [2]. They are often lined by cuboidal or columnar epithelia with surrounding thin fibrous tissue admixed with non-dilated extramural glands and conduits [2,3].

Case Report

A 43 years old man, known case of alcoholic liver disease with decompensated liver cirrhosis, esophageal varices and en-
Cephalopathy presented to a nearby outpatient clinic with increasing yellowish skin discoloration, itching and dark urine.

On clinical examination, the patient was oriented to time and place but appeared jaundiced. His abdomen was soft with an enlarged palpable liver and shifting ascites. The patient was admitted to the hospital for further investigation and management.

His blood workup showed increased total bilirubin and direct bilirubin. Indirect bilirubin was within the normal range. Alkaline phosphatase and Alanine aminotransferase (ALT) were also elevated and inflammatory markers ESR (84) and CRP (37) were moderately high. CA19 was measured (214.6 U/ml) and was found to be elevated. AFP tumor Marker was within normal limits. Urine analysis revealed positive urine bilirubin and urobilinogen.

An urgent sonogram of the upper abdomen was requested to rule out biliary obstruction. It showed extensive intrahepatic dilatation which was more in the left than the right lobe. No mass was visualized at the hilar region. Common bile duct appeared within normal limits. A patent portal vein was depicted on doppler.

Further imaging with contrast-enhanced triphasic CT scan of the abdomen revealed cirrhotic changes in the liver parenchyma with upstream right and left intrahepatic biliary dilatation. No evidence of a hilar mass or significant lymphadenopathy was seen. However, extensive collaterals with ascites suggestive of portal hypertension were observed. The patient underwent MRCP examination upon suspicion of Klatskin tumor, which revealed bilobar intrahepatic dilatation and a stricture at the hilum of the intrahepatic ducts (Fig. 1).

The apparent dilatation of the biliary tree in both lobes raised the suspicion of an occult malignant stricture situated at the hilum. The patient underwent endoscopic retrograde cholangiopancreatography which revealed a stricture at the proximal main and left hepatic duct with evident upstream biliary dilatation in both lobes and beading of the smaller ducts (Fig. 2). The picture of smaller branch beading raised the suspicion of primary sclerosing cholangitis or IgG4 cholangiopathy. The common bile duct diameter was normal. Brush cytology of the stricture was taken, and an 8.5 F 15 cm plastic stent was placed after a sphincterotomy where drainage of the bloody bile secretions was attained. The patient’s bilirubin levels gradually decreased, and his symptoms improved.

The histopathology of the brushed sample showed flat large honeycomb sheets of uniform cells with few scattered groups of cuboidal epithelial cells and no features of atopy or malignancy. The patient underwent a change of stent as he later on developed fever and features of cholangitis.

Afterward, the patient presented to our health facility for further workup as his chronic liver disease was progressing and as hilar cholangiocarcinoma was suspected on top of his chronic liver disease.

MRI/MRCP of the liver was performed and showed multiple clustered cysts located circumferential to right and left portal vein tracts and the hilar confluence with compressive pressure effects on the adjacent biliary tree causing a beaded appearance on MRCP. These findings suggested peribiliary cysts along the intrahepatic biliary tree. (Fig. 3)
PET-CT was performed to rule out a malignant cause of the coexisting stricture. No abnormally increased FDG uptake was appreciated. (Fig. 4)

After the final imaging diagnosis ruled out biliary malignancy, a full liver transplant workup was completed and the patient underwent a successful orthotopic liver transplant for his chronic liver disease. Histopathology of the explant confirmed the diagnosis and showed multiple benign simple liver cysts as foci of dilated epithelial lined cystic lesions on a background of mostly nodular liver parenchyma (Fig. 5).

Discussion

Hepatic peribiliary cysts represent a benign entity that is observed more commonly in males than in females with a median age of occurrence of 63 years old [6]. Patients are usually asymptomatic [8]. They are often associated with chronic liver disease and may rarely present with obstruction in less than 2% of patients due to the external compression of the biliary

Fig. 3 – (A-D) MRI of the same patient. A. Axial T2 weighted image reveals portal vein signal void (short arrow) and peribiliary cysts circumferential to portal vein tract (long arrow). B. Coronal MIP MRCP image shows clustered cysts along the intrahepatic biliary tree. C. Postcontrast T1 fat saturated image confirms the portal vein enhancement (short arrows), and peribiliary cysts appear hypointense around the portal vein. D. Same findings are also demonstrated on coronal T2 weighted image.

Fig. 4 – PETCT axial image at the level of the liver reveals physiologic FDG uptake.
Fig. 5 – Histopathology images showed multiple benign simple liver cysts. A. Image shows mostly nodular liver parenchyma (left) and foci of dilated epithelial lined multiple cystic lesions (Original magnification x40). B. Cysts are lined by bland appearing biliary-like epithelium with no high-grade dysplasia (Original magnification x200). Haematoxylin and eosin staining.

tree [4,7]. Its frequency is said to be directly related to the degree of liver disease [6]. However, they do occur in less than 5% of patients with normal liver function [7].

Two different theories have been proposed as the likely mechanism for cyst formation. The first theory is that there is an inflammatory circulatory disturbance such as in cases of portal hypertension that leads to gland obstruction and cyst formation. The second suggested mechanism may be attributed to hereditary factors such as in autosomal dominant polycystic liver disease [2,7]. Hepatic peribiliary cysts are observed in all patients with autosomal dominant polycystic liver disease who usually show diffuse excessive glandular dilatation but no detectable perifocal inflammatory changes [7]. It is also seen in patients with polycystic kidney disease and is likely due to a specific gene expression in the intrahepatic glands that eventually leads to the cyst formation [5,6].

The incidence of cyst occurrence seems to be also increased in patients who underwent hepatic portoenterostomy strengthening the hypothesis of disturbed vascular flow as the main etiology of this disease [5].

Sato et al. detected glandular epithelial cyst changes and speculated that there may be a correlation between peribiliary hepatic cysts and the formation of intraductal papillary neoplasms and cholangiocarcinoma [10]. Although this remains unclear, an association with biliary neoplasms should be kept in mind during follow-up visits of these patients [2,6,10]. The synchronicity of peribiliary hepatic cysts with hepatic, biliary, and pancreatic tumors was seen in 20% of studied cases. Hepatocellular carcinoma was the most common associated neoplasm [6].

The differential diagnosis of this benign entity includes other cystic diseases of the liver parenchyma such as intraductal papillary mucinous neoplasms of the bile ducts, cystic metastasis, Caroli’s disease, abscess formation, and primary sclerosing cholangitis. Nevertheless, Klatskin tumor at the hilum causing biliary dilatation is the most common and most serious differential of peribiliary hepatic cysts [6,7].

Early detection and reaching the correct diagnosis of hepatic peribiliary cysts is crucial as a misdiagnosis may cause delays or even denial of the patients undergoing liver transplantation for their end-stage liver disease [1].

On radiological imaging, a liver sonogram may perceive hepatic peribiliary cysts as larger echogenic portal tracts, as an echogenicity beside or connected to the portal vein or as a hilar multicystic mass [2].

Two imaging modalities are proposed for the diagnosis of peribiliary cysts. Magnetic resonance cholangiography can demonstrate the cysts and can differentiate from mimickers with the accurate anatomic resolution of the biliary tree and periporal tracks noninvasively. Drip infusion cholangiographic computed tomography imaging can also clearly distinguish between biliary dilatation and peribiliary cysts with the enhancement of the intrahepatic bile ducts [1,5]. The latter imaging modality however, may not yield clean cut results in patients with hepatic dysfunction and is only used if the liver function is normal [1,6]. It is often difficult to differentiate these cysts from gross biliary dilatation on standard computed tomography due to the thin septa surrounding these cysts and if they are minute in size they may mimic periportal edema or periportal fat deposition [7].

On magnetic resonance imaging, these cysts have the same fluid signal as the dilated biliary tree thus they are also difficult to diagnose [4]. However, they may appear as a tubular cluster of cysts hyperintense on T2 weighted images [7,9]. Magnetic resonance cholangiography may reveal a lobulated fringe along the hepatic hilum of larger ducts with beading of the smaller ducts [5]. Grouped together cysts can give the appearance of a string of beads that may mimic the findings of primary sclerosing cholangitis [7].

Endoscopic retrograde cholangiography clearly shows that there is no communication of the cysts with the biliary tracts as they do not fill upon contrast injection [4]. However, the compressive effect of the peribiliary cysts may mimic primary sclerosing cholangitis and cholangiocarcinoma as in our case.
The complications of hepatic peribiliary cyst formation include biliary obstruction with associated jaundice, cholangitis due to bile stasis, hepatolithiasis, pancreatitis, and peribiliary cyst infection [6]. Collina et al. reported peribiliary cyst formation as an infrequent complication in post liver transplant patients [9]. These cysts may ultimately require medical or surgical management as they are progressive and the resolution of symptomatic biliary obstruction may be achieved by fenestration of the obstructing cysts [2,4].

Hepatic peribiliary cysts are not well recognized in the daily clinical practice [5,9]. The presence of these cysts may be of diagnostic value in suggesting an undiagnosed underlying chronic liver disease [9]. Careful observation and knowledge of the presence and higher prevalence of peribiliary hepatic cysts in a cirrhotic liver should be applied to differentiate this benign entity from malignancy that may halt or delay the patients’ eligibility for receiving a liver transplant [2,6].

In conclusion, we presented a case with peribiliary cysts in a cirrhotic liver. Careful evaluation of contrast-enhanced MRI with MRCP can correctly differentiate it from cholangiocarcinoma and enabled the cure of the patient by liver transplantation.

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