A Large hepatic Peribiliary Cyst - A Case Report

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Abstract:
Peribiliary cyst is a poorly recognized and under-reported clinico-pathological entity around the biliary tree. Peribiliary cysts are cystic dilatations of obstructed peribiliary glands, which are normal elements of the biliary tract. They are generally asymptomatic and rarely cause biliary obstruction. They are usually discovered incidentally at autopsy. A 77-year-old female patient presenting with abdominal pain and abdominal distension for 1 month duration due to a large hepatic peribiliary cyst is re-reported here. Radiological (CECT) imaging demonstrated a large cystic lesion with septation occupying Right hypochondrium and Right lumbar region, arising from right lobe liver displacing the liver medially but not communicating with the bile duct. Our pre op diagnosis was either hydatid cyst or simple liver cyst. Laprotomy with de-roofing of the cyst was performed and histological examination of the cyst wall revealed a Non-Neoplastic Cyst - Peribiliary gland cyst. The patient made an uneventful recovery and remained asymptomatic with normal liver function tests 8 months postoperatively.

Keyword: Peribiliary glands, peribiliary cyst, liver cyst

Introduction:
Peribiliary cysts (PBCs) are inflammatory retention cysts that evolve from cystic dilatations of obstructed small peribiliary glands (PBGs) of larger extra-hepatic and intra-hepatic bile ducts (1). These cysts are usually multiple, ranging in size from a few millimeters to 1 cm, and rarely up to 3 cm in diameter. They are located around the larger intra-hepatic bile ducts, termed as “hepatic peribiliary cysts” or “intra-hepatic peribiliary cysts” (1). Histologically, PBGs are classified into intramural and extramural according to their location. The intramural glands are scattered within the bile duct walls, few in number, and are non-branching simple tubular mucinous glands. In contrast, the extramural glands are more abundant, located in the periductal connective tissue and are branching tubulo alveolar seromucinous glands that contain enzymes to digest protein and lipids (2-4).

Histologically, the wall of the PBC consists of cuboidal or low columnar or flattened epithelium and connective tissue (3-4). PBCs are generally asymptomatic and rarely cause biliary obstruction. Only 4 cases presenting with obstructive jaundice have been reported (4-7). More frequently they are discovered incidentally during radiological screening examination or on autopsy (2). We report a case of large intra-hepatic peribiliary cyst, initially thought to be a large hydatid cyst or simple liver cyst.

CASE REPORT:
A 77-year-old woman was admitted with symptoms of upper abdominal pain and abdominal distension that had existed for 4 weeks. H/O fever and vomiting for 10 days. She had no history of co-morbid medical illness. The symptoms settled after a short course of intravenous antibiotics except abdominal distension. Physical examination revealed a huge mass occupying Right hypochondrium, epigastrium and umbilical regions. Mass was cystic in consistency with smooth margins. Laboratory investigations revealed as an increase of the total leukocyte count 13000 cells/mm³ (normal : 4000-11,000 mm³), serum total bilirubin-1.1 mg/dL (normal : up to 1.2 mg/dL), serum alanine transaminase-19 U/L (normal : up to 45 U/L), serum aspartate transaminase-12 U/L (normal : up to 45 U/L), serum alkaline phosphatase-56 IU/L (normal : 108-306 IU/L), and serum gamma-glutamyl transpeptidase- 30 U/L (normal : 10-50 U/L). Her coagulation profile was normal and the panel of hepatitis viral markers was negative. Repeated blood cultures detected no microbial organisms.

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Abdominal ultrasonography (US) showed a huge large cyst in right hypochondrium - ? Right renal cortical cyst.

CECT- ABDOMEN
Contrast enhanced computed tomography (CECT) revealed a large cystic lesion with septation occupying right hypochondrium and right lumbar quadrant arising from right lobe of liver, displacing the liver to left side and right kidney medially. The final impression was suggestive of ? Hydatid cyst. Patient was not affordable for magnetic resonance cholangio-pancreatography (MRCP). Since we thought of Hydatid cyst Ultrasound-guided percutaneous fine needle aspiration of cyst fluid was not performed pre-operatively. Our pre-operative diagnosis was ?Hydatid cyst/ ?Simple liver cyst.

INTRA-OPERATIVE PICTURES
Surgical exploration and resection was planned and all precautionary measures required for hydatid cyst exploration were taken in to the account.
At laparotomy, the liver was hugely enlarged below upto the level of the umbilicus. Liver surface was smooth, with a huge cyst occupying over the right lobe of the liver, the surface of which was bluish black in colour and a clear straw-coloured fluid was aspirated from the cyst, thereby excluding hydatid cyst. Then de-roofing & marsupialisation of the cyst was done.

Internal examination of the cyst showed 3 cysts in number, largest measured 20 x 15 cm. Other 2 cysts measured 10 X 8 cm & 3 X 2 cm. All the cysts occupied the postero-superior aspect of right lobe liver with about 1.5L straw coloured fluid. All the 3 cysts were communicating with each other. Tube drainage of the right sub-hepatic space was then performed with uneventful recovery. The de-roofed cyst showed a blind ending trilobar cystic space, not communicating with bile duct lumen. Histological examination of Liver Cyst Wall section showed fibro-collagenous cyst wall containing hepatocytes arranged in nodular pattern with bile duct. Cyst wall in occasional areas lined by cubo-columnar epithelium with uniform nuclei. Impression was Non-Neoplastic Cyst – Peribiliary gland cyst. Liver cyst fluid cytology showed eosinophilic material only with no cells or structures. During the follow-up of 8 months, the patient remained asymptomatic with normal liver function tests.

DISCUSSION:
Peribiliary cysts have also been termed as hepatic hilar cysts [1] and mucinous hamartoma of the bile ducts [5]. These cysts were first described in 1984 by Nakanuma et al. [1] who described the pathologic findings in eight cases found incidentally at autopsy, in that series, the cysts were from 0.2 to 2.0 cm in diameter and were found in the peribiliary connective tissues in the center and periphery of the porta hepatis. The cysts occurred in the same distribution They are located in the connective tissue of hepatic hilum and within the larger portal tracts. The cyst are typically unilocular and contain serous fluid. They are lined with a single layer of cuboidal/columnar epithelium similar to bile duct epithelium. Cystography has shown that these cyst are isolated without communication with bile ducts and necrosis and inflammation of the peribiliary glands can be seen in 23% of cases.
suggesting intrahepatic circulatory disturbance. Rarely, they can cause obstructive jaundice and cholangitis (4,7). For the most part, they are found incidentally at surgery or autopsy, in fact, 50% of cirrhotic livers and 73% of polycystic liver may demonstrate cystic dilation of the peribiliary glands, whereas the incidence is only 3% in normal livers (9). There is a potential genetic predisposition with autosomal dominant polycystic kidney disease with liver involvement. Kida et al. (10) reported that PBGs of the extra hepatic bile ducts showed no cystic dilations in most cases, unlike the intra-hepatic peribiliary glands. On ultrasound examination, they appear as round or tubular anechoic areas or clustered small cysts around the large portal tracts. On CT imaging, there can be multiple small cysts <1 cm in diameter or larger discrete cysts >1 cm in diameter, scattered along the portal vein. On MRI imaging, they appear as hypointense areas surrounding the portal vein on T1-weighted images and hyperintense areas on T2-weighted images. They also can be identified using MR cholangiography, appearing as a string of beads along the hepatic hilum or larger bile ducts (11). Extrinsic compression of the bile ducts can be seen on ERCP or percutaneous transhepatic cholangiography. These cysts may enlarge over time and increase in number. There has been one case in which these peribiliary cysts were associated with bile duct carcinoma (11). Therefore, long-term observation seems warranted.

Table: Features of Simple Liver Cyst and Peribiliary Cyst

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<tr>
<th>Features</th>
<th>Simple Liver Cyst</th>
<th>Peribiliary Cyst</th>
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<tbody>
<tr>
<td>Gross</td>
<td>Solitary or multiple unilocular cysts, parenchymal or subcapsular</td>
<td>Multiple unilocular cysts, in hepatic hilum</td>
</tr>
<tr>
<td>Biliary tree connection</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>Epithelial lining</td>
<td>Cuboidal or low columnar epithelium</td>
<td>Flat or cuboidal epithelium</td>
</tr>
<tr>
<td>Cyst wall stroma</td>
<td>Fibrous</td>
<td>Fibrous</td>
</tr>
<tr>
<td>Cyst content</td>
<td>Clear serous fluid</td>
<td>Clear serous fluid</td>
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A diagnostic dilemma can exist when cystic lesions are limited to the larger bile ducts and the hepatic hilum. Under these circumstances, a cholangiographic contrast enhanced multi-detector CT performed immediately after drip-infusion cholangiography (DIP), i.e. CT cholangiography (11), direct biliary imaging, i.e. ERCP with intraductal ultrasound (IDUS) by using a 20 MHz over-the-guide wire catheter probe, may help to delineate an extra luminal origin. The prognosis of PBC is uncertain. Although the majority of cases have been discovered incidentally, malignant transformation is unreported, the natural history is unknown. Reported series are small, long-term follow up data are limited and the conditions are mostly under reported.

Conclusion:
We reported a patient with a large peribiliary cyst arising from the intrahepatic bile duct. The cyst was a very large, multiple and absence of communication with bile duct with normal liver functions. Our pre operative diagnosis was ?

Hydatid cyst? Simple liver cyst. Surprisingly histopathology report revealed peribiliary cyst. It is one of the rarest presentation of the cystic lesion of the liver.

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