Abdominal imaging PICTORIAL ESSAY

Peribiliary cysts: CT and MRI evaluation

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ABSTRACT

Introduction. The purpose of our study is to describe the characteristics of peribiliary cysts, a generally benign entity that may be confused with other entities, and to emphasize the relationship with other diseases.

Methods. We examined 29 people over 10 years with the modalities of Computed Tomography (CT), and Magnetic Resonance Imaging (MRI).

Results. 29 patients with peribiliary cysts were examined, aged between 6 months and 57 years. In one patient the peribiliary cysts were combined with Autosomal Dominant Polycystic Kidney Disease (ADPKD). One patient had also simple cysts, Von Meyenburg complex lesions, and ADPKD. In one patient peribiliary cysts were associated with Cholangiocarcinoma of the caudate lobe of the liver.

Conclusion. Peribiliary cysts may sometimes be confused with other entities. The knowledge of the pathology of peribiliary cysts and the establishment of the correct diagnosis will help patients to avoid unnecessary procedures.

Introduction

Peribiliary glands are present around intrahepatic and extrahepatic bile ducts, around the cystic duct, and sometimes around the neck of the gallbladder.

The peribiliary glands are classified into two groups (fig.1):

a) intramural without branches and b) extramural with branches.

The extramural peribiliary glands connected between them and consisted of serous and mucinous acini. The lobules of peribiliary glands are lined by simple columnar epithelium with some progenitor cells/biliary tree stem cells and small foci of pancreatic tissue. Peribiliary glands have their openings into the lumen of bile ducts or the commu-

KEY WORDS | Peribiliary cysts, Cholangiocarcinoma, Autosomal Dominant Polycystic Kidney Disease (ADPKD), Multiple hilar hepatic cysts

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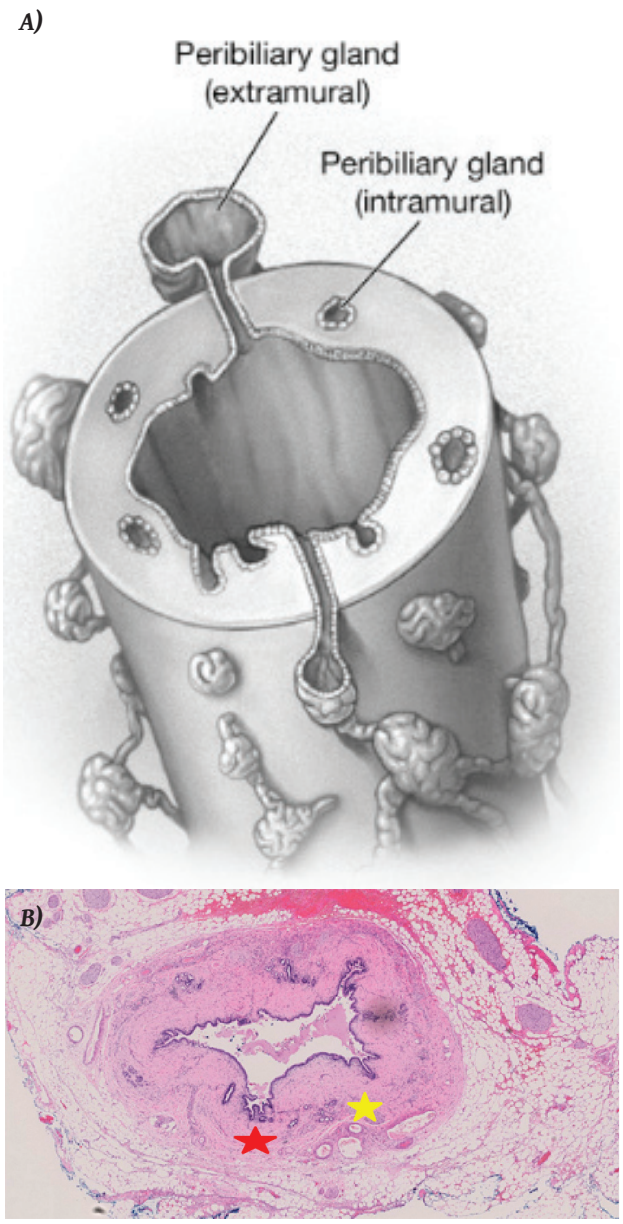

Fig.1 A) *Schematic representation of intramural and extramural peribiliary glands (bazerbachi F, et al.BMJ Open Gastroenterology 2018;5: e000204).*
B) *Histology with low power view showing a cross-section of the bile duct. Red asterisk= peribiliary gland, yellow asterisk = sacculi of Beale (NUS- Yong Loo Lin School of medicine).*

nication between peribiliary glands and the lumen of bile ducts succeeds via small intramural pits which are named sacculi of Beale.

Distortion of the communication between peribiliary glands and bile ducts lumen results in the origin of the peribiliary cysts.

Fig.2: *Schematic representation of the distribution of peribiliary cysts in the biliary system in 29 cases. In 26 cases the cysts are located in the right or left main hepatic duct and in 3 cases in the right or left main hepatic duct and smaller intrahepatic bile ducts.*

Methodology

29 patients had an exam by CT and MRI.CT scans were obtained by a SIEMENS Somatom Perspective 16 machine with a 2mm scan width before and after intravenous contrast material administration.

Also in some cases, we perform CT Cholangiography using intravenous administration of contrast material secreted by hepatocytes into the biliary system (biliscopin).

MRI images were obtained using a 1,5 Tesla PHILLIPS machine.

Results

From the 29 cases with peribiliary cysts which we studied, in 26 the cysts were located in the right and/or left main hepatic duct whereas in 3 cases they were located in the right and/or left main hepatic duct as well as in smaller intrahepatic ducts(fig.2). 2 of our cases were combined with polycystic kidney disease ADPKD type(fig.3-4) and 1 with Cholangiocarcinoma(fig.5). Regarding the imaging findings, in CT 19 of them had water densities whereas the other 10 had a little higher than that of water. In MRI all the cysts had the typical appearance of a cyst with a high signal on T2WI, a low signal on T1WI without enhancement after contrast material administration(fig.6).

Discussion

Peribiliary cysts (formerly called multiple hilar cysts of

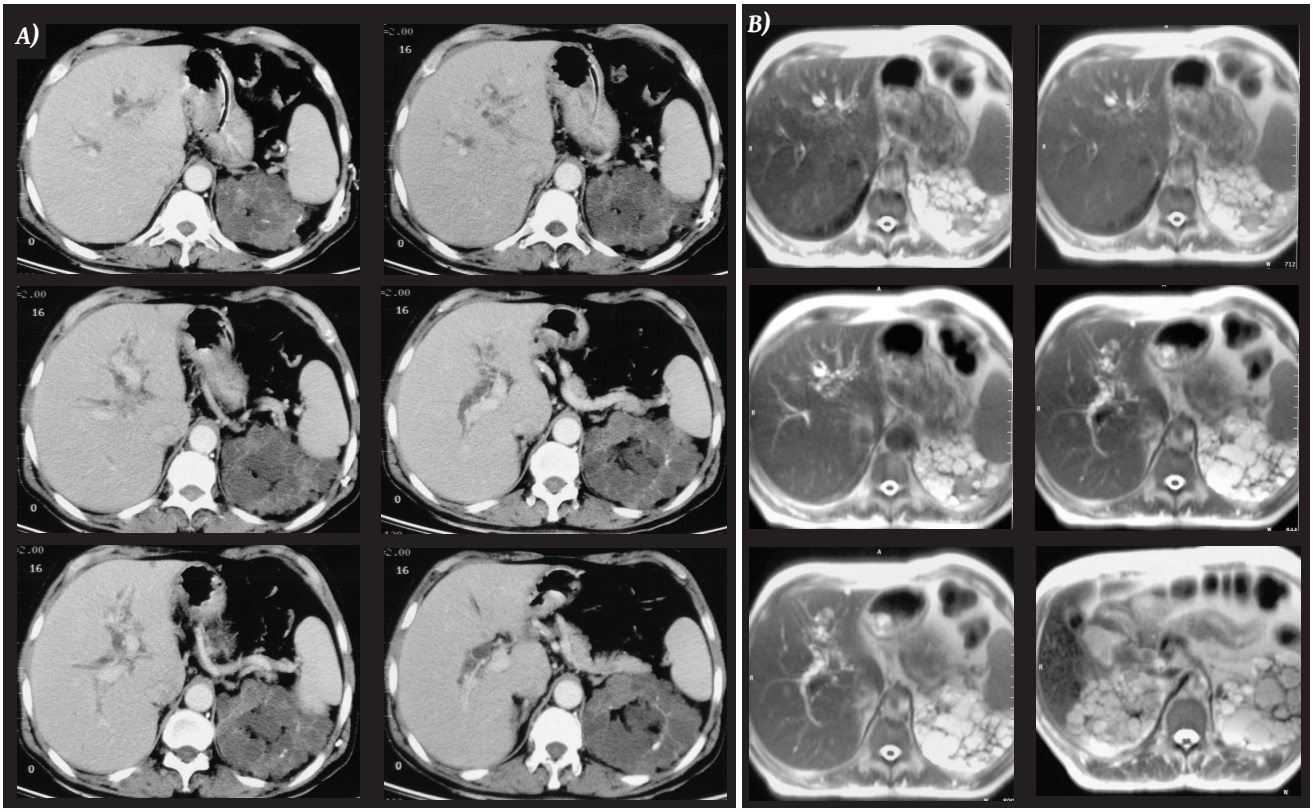

Fig. 3: *Intrahepatic peribiliary cysts in a patient with ADPKD.* ***a)*** *axial CT images and* ***b)*** *MR T2WI axial images reveal the peribiliary cysts and the polycystic disease of the kidneys.*

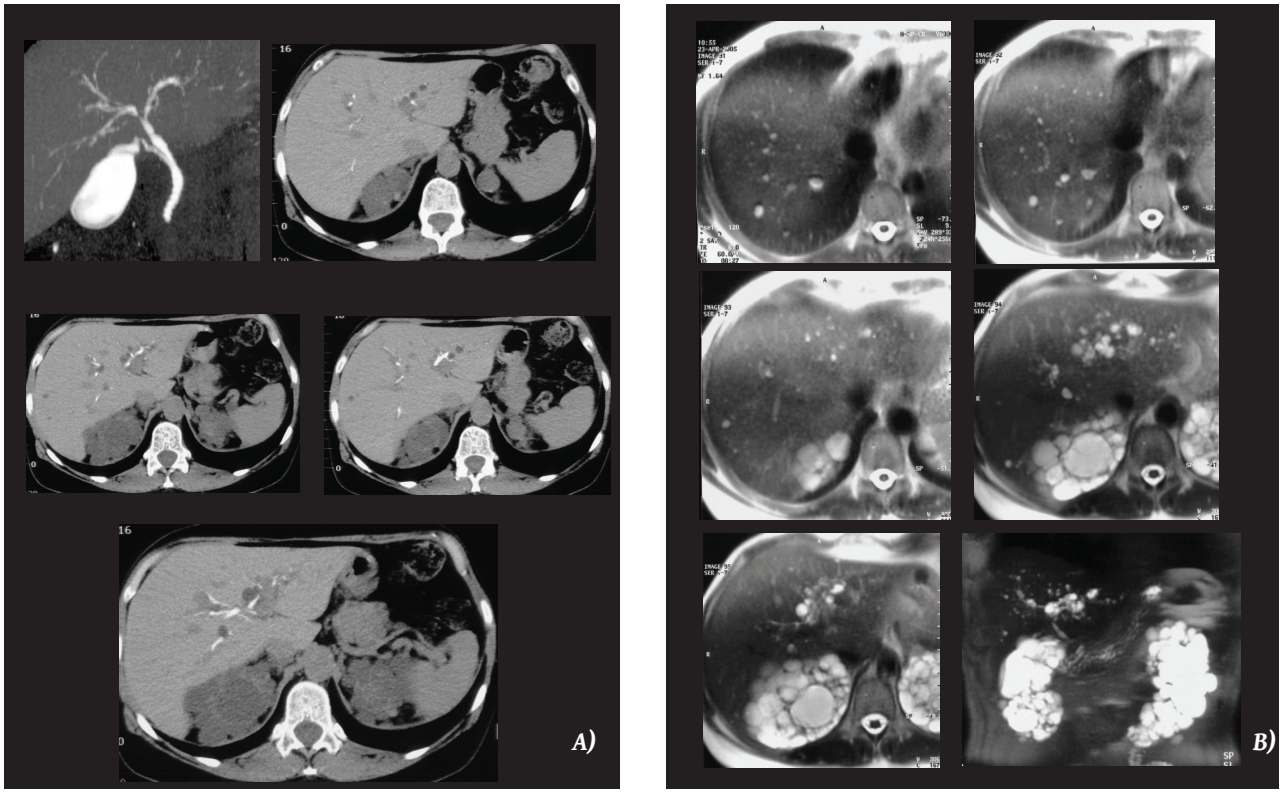

Fig. 4: *Intrahepatic peribiliary cysts, non-parasitic simple cysts, and Von Meyenburg complex lesions in a patient with ADPKD.* ***a)*** *CT Cholangiography and axial source images, which reveal the normal biliary tree, peribiliary cysts, simple cysts, and polycystic kidney disease.* ***b)*** *MRCP and MR T2WI axial images, which more clearly depict a normal biliary tree, peribiliary cysts, simple non-parasitic cysts, lesions of Von Meyenburg, and polycystic kidney disease.*

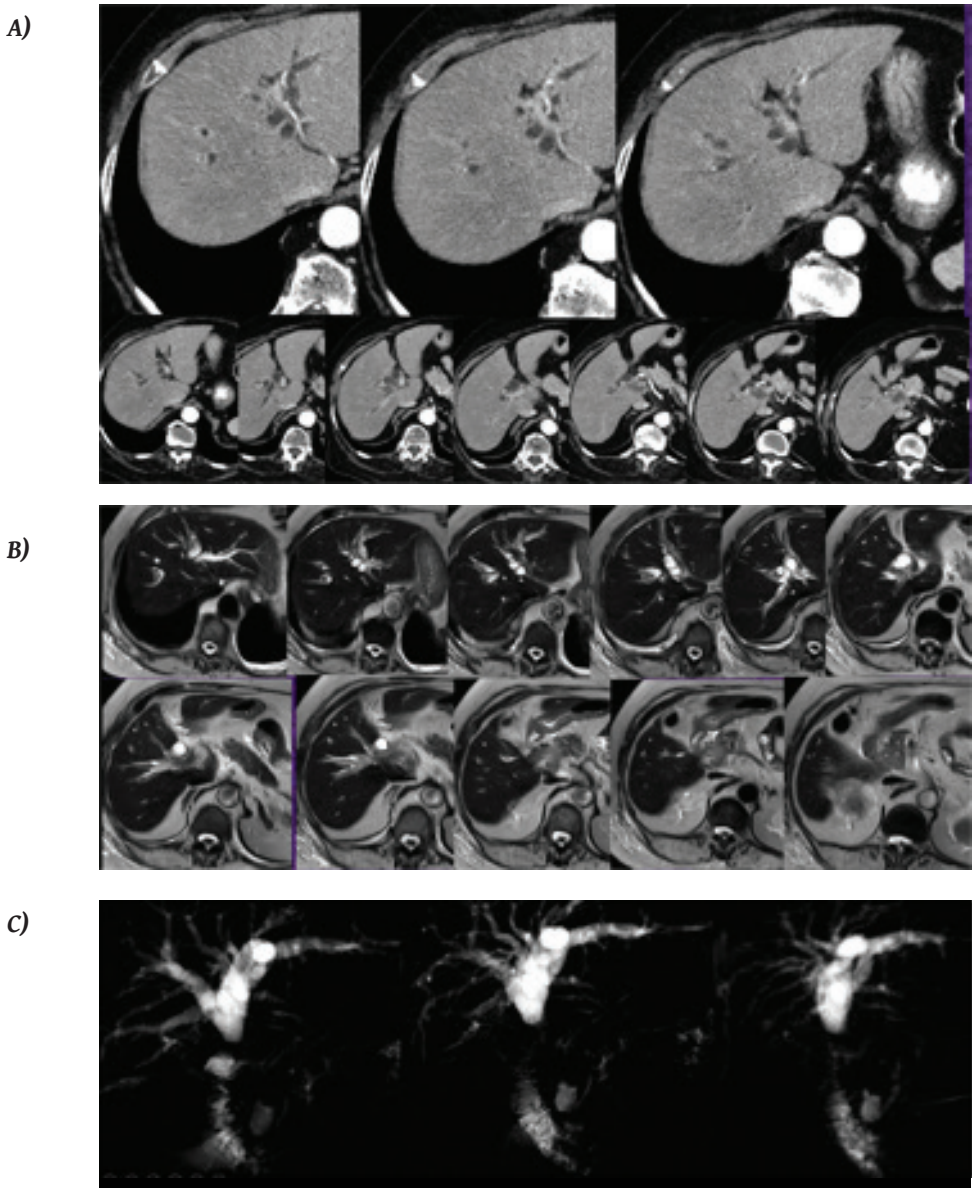

Fig. 5: *Intrahepatic peribiliary cysts and Cholangiocarcinoma.* ***a)*** *axial CT scans and* ***b)*** *MR T2WI axial scans, which reveal peribiliary cysts and a lesion in the caudate lobe of the liver, proved to be a Cholangiocarcinoma.* ***c)*** *MRCP which clearly depicts the level of the obstruction of the common hepatic duct and the peribiliary intrahepatic cysts.*

the liver) represent cystic dilatations of peribiliary glands, found in the connective tissue around the bile ducts without communication with the biliary tree [1]. They are multiple small cysts adjacent to the hepatic hilum and the larger ducts (from the main duct to the third-order branches) [2,3].

Peribiliary glands are tubuloalveolar seromucous glands around the extrahepatic and intrahepatic large bile ducts – relatively dense in the hilar bile ducts, cystic duct, and periampullary region. These glands may be associated with mucin hypersecretion and involved in the pathogenesis of stone formation (hepatolithiasis) or even Cholangiocarcinoma. There are two possible mechanisms underlying the development of peribiliary cysts [4,5,6,7].

The first is associated with adenitis and is found in acquired diseases, such as liver cirrhosis with or without hepatocellular carcinoma, alcoholic liver diseases, a malignant disease with obstructive jaundice, ascending cholangitis, systemic infection, septicaemia, portal hypertension, thrombosis. Due to inflammatory or ischemic processes,

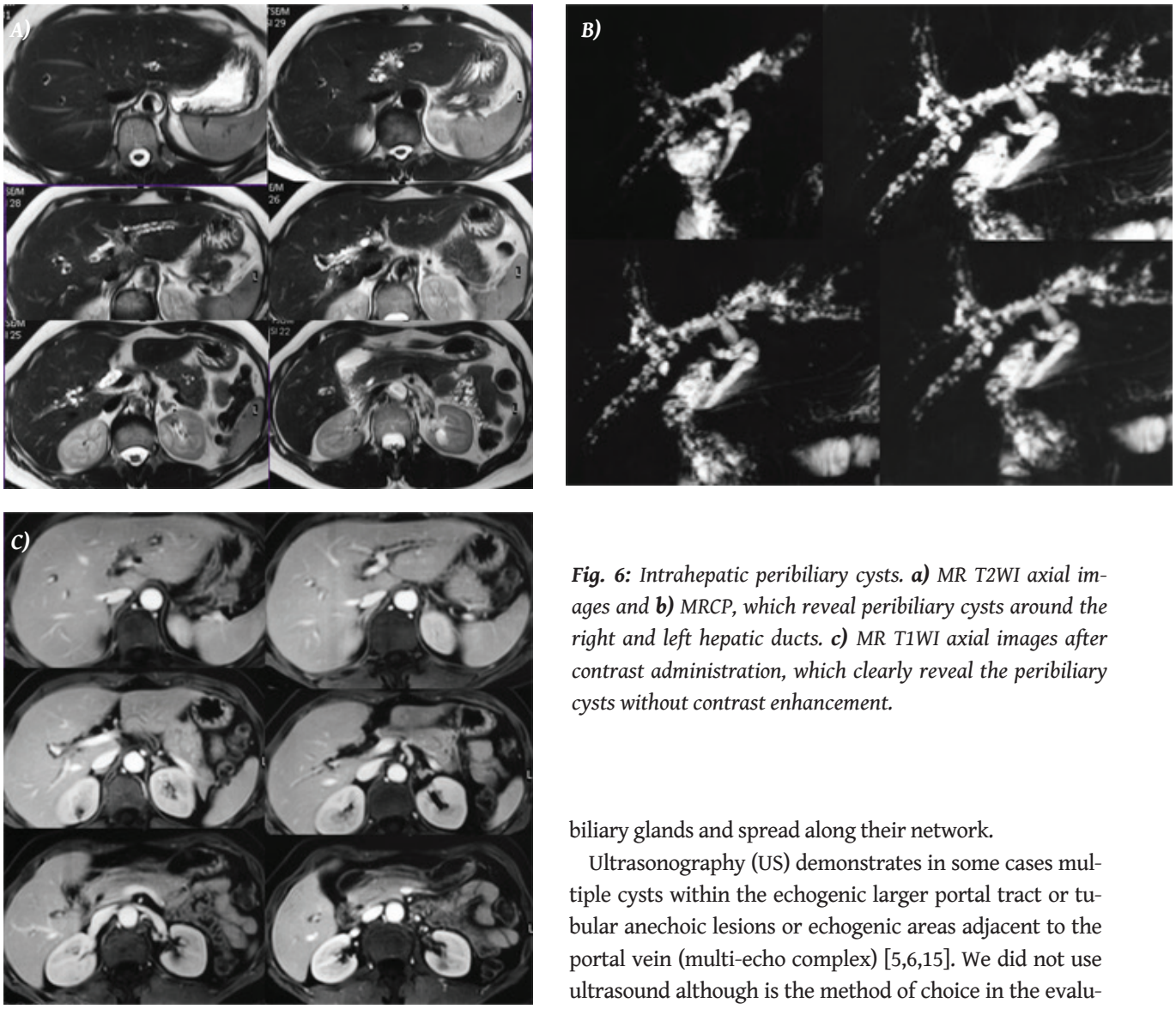

Fig. 6: *Intrahepatic peribiliary cysts.* ***a)*** *MR T2WI axial images and* ***b)*** *MRCP, which reveal peribiliary cysts around the right and left hepatic ducts.* ***c)*** *MR T1WI axial images after contrast administration, which clearly reveal the peribiliary cysts without contrast enhancement.*

the conduits of peribiliary glands are obstructed, thus forming retention cysts [1,2,8].

The second mechanism is related to congenital cystic diseases, such as autosomal dominant polycystic kidney disease (ADPKD) [3,7,9].

Although peribiliary cysts are usually asymptomatic, it has been found that they gradually enlarge in size and increase in number, paralleling the course of hepatic diseases, particularly alcohol-related processes [6,7,10]. They may even result in obstructive jaundice if the cysts are large in size and compression of the biliary tract becomes complete [7,11,13]. Peribiliary cysts are associated with cholesterol hepatolithiasis, Cholangiocarcinoma (CCA), and other neoplasms [7,14].

Some types of CCA may even arise primarily from peribiliary glands and spread along their network.

Ultrasonography (US) demonstrates in some cases multiple cysts within the echogenic larger portal tract or tubular anechoic lesions or echogenic areas adjacent to the portal vein (multi-echo complex) [5,6,15]. We did not use ultrasound although is the method of choice in the evaluation of cystic lesions because peribiliary cysts are usually small in size and very difficult to evaluate the details of their characteristics.

On Computed Tomography (CT) peribiliary cysts appear along either side or both sides of portal veins and bile ducts from the hepatic hilum to the third or fourth-order branches [15]. They may present as separate discrete near-water attenuation lesions adjacent to the hilum or the proximal biliary ducts, as a linear cluster (a string of beads), or as a low-density confluent linear structure [4]. CT Cholangiography (a technique using meglumine iotroxate preferably excreted into the bile duct) reveals the true ductal lumen, which is sometimes compressed by large peribiliary cysts [7.15]. The peribiliary cysts do not communicate with the biliary tract, so they do not fill with contrast material [8]. Nevertheless, the method requires

normal hepatic function [11].

Magnetic Resonance Imaging (MRI) and Magnetic Resonance Cholangiopancreatography (MRCP) show peribiliary cysts as multiple small T2WI hyperintense cystic structures along the portal vein in the porta hepatis to perihilar regions. MRCP demonstrates the string of bead-like lesions which lie adjacent to the biliary tree, even in patients with end-stage liver disease [4].

There are benign and malignant conditions that may mimic peribiliary cysts including primary sclerosing cholangitis, localized or diffuse mucinous cystic neoplasm, abscess, polycystic liver disease, von Meyenburg complex, Caroli disease, IPNB (Intraductal Papillary Neoplasm of Biliary ducts), Cholangiocarcinoma, solitary simple cysts, and periportal lymphoma.

In conclusion, a radiologist should be familiar with the diagnosis of peribiliary cysts, because it is crucial to distinguish this predominantly benign entity from malignancy or other diseases which require intervention. **R**

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