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Case Report

Peribiliary cysts: Two case reports ☆

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ABSTRACT

Peribiliary cysts are an incidental finding in patients with advanced liver disease. They were found to be common in fibrocystic disorders such as autosomal dominant polycystic kidney disease and polycystic liver disease, as well as cirrhotic livers. They result from obstruction-induced cystic dilatation of the peribiliary glands. We report in this article the case of 2 patients, aged 41 and 71, smokers and chronic alcoholics, admitted for febrile cholestatic jaundice, for which MRI revealed the presence of peribiliary cysts.

Introduction

Peribiliary cysts were first described by Nakanuma et al. in 1984. They are defined as cystic dilatations of the intrahepatic extramural peribiliary glands, located adjacent to the intrahepatic bile ducts. These ovoid or spherical cysts, ranging in size from 0.2 to 2.5 cm^2, are confined to the hepatic hilum and the enlarged portal tract, areas which contain the periductal glands. The pathogenesis of these cysts involves genetic factors in fibrocystic disorders and disruption of periportal blood flow and inflammation in cirrhotic patients, leading to obstruction of the peribilar glands and resulting cystic dilatation. The cysts are located outside the duct walls and do not communicate with the bile duct lumen. Detecting these cysts may reveal an underlying chronic liver condition in previously undiagnosed patients.

Case reports

Case one

A 43-year-old male, with a history of smoking and chronic alcohol consumption, was admitted following 2 episodes of angiocholitis. He had a prior myocardial infarction, treated with thrombolysis, and was on antiplatelet therapy. The patient presented with rectal bleeding, both defecatory and nondefecatory, persisting for 2 days. Physical examination revealed

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a distended abdomen with venous collaterals, cholestatic jaundice, and dusky urine, with normally colored feces. Two slightly erythematous and lichenified patches were noticeable on the lower limbs.

Laboratory tests indicated normocytic hypochromic anemia, elevated total and direct bilirubin levels (124 μmoL/L and 100 μmoL/L, respectively), and increased liver enzymes (PAL 5xN, GGT 13, GOT 2.7xN). Renal function tests showed mild renal failure (urea 0.7 mmoL/L, creatinine 22 μmoL/L). An emergency colonoscopy was unremarkable.

Abdominal ultrasound revealed a liver of normal size, but with significant dilatation of the intrahepatic bile ducts and right-left disconnection overlying a hepatic hilar tissue infiltrate.

A Bili MRI (Fig. 1) was performed, showing the presence of a 22 mm-long hilar sheet tumour causing significant dilatation of the upper intrahepatic bile ducts and disconnection of the right and left ducts, with compression of the right portal branch without infiltration. Multiple cystic lesions on either side of the portal branches, involving the entire liver, are described as T1 hyposignal, T2 liquid hypersignal, nonrestrictive in diffusion, and not modified after contrast, without communication with the bile ducts, reported to be peribiliary cysts.

Case two

A 71-year-old patient, smoker and chronic alcoholic for 20 years, who had been abstinent for 10 years, admitted for cholestatic jaundice with normo-colored feces, associated with abdominal pain more pronounced in the right quadrant, with no other abdominal or extra-abdominal clinical features, all this evolving in a context of fever and unquantified weight loss.

Laboratory results included hemoglobin 8.3 g/dL, WBC 21,000/mm³, PNN 20,140/mm³, PT 58%, and elevated liver enzymes (GOT 1.2xN, GPT 1.1xN, PAL 5xN, GGT 4.4xN). Renal

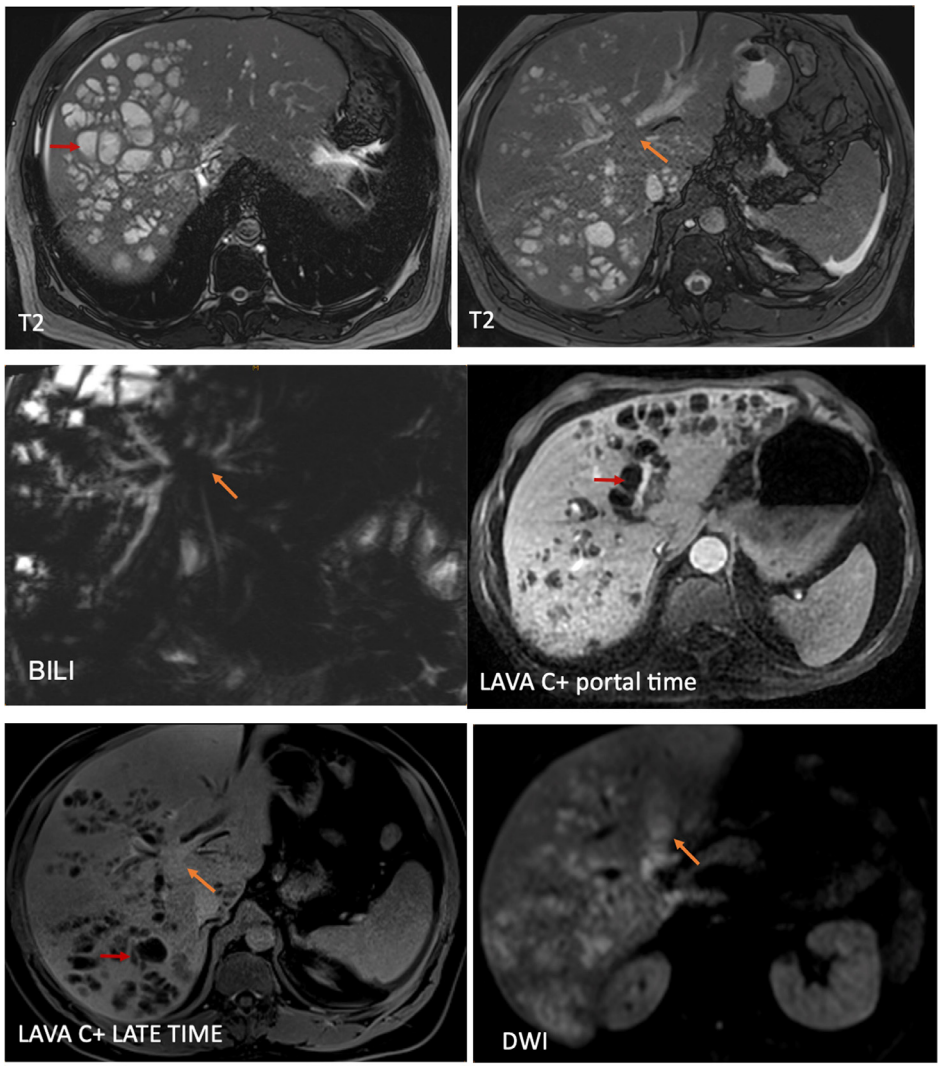

Fig. 1 – MRI in axial and coronal sections, showing the presence of multiple cystic lesions on either side of the portal branches (red arrow), described in T2 fluid hypersignal, non-restrictive in diffusion, and not enhanced after portal and late contrast. MRCP sequence: dilatation of the intrahepatic bile ducts with right and left disconnection, upstream of a tumor of the hilar plate (orange arrow).

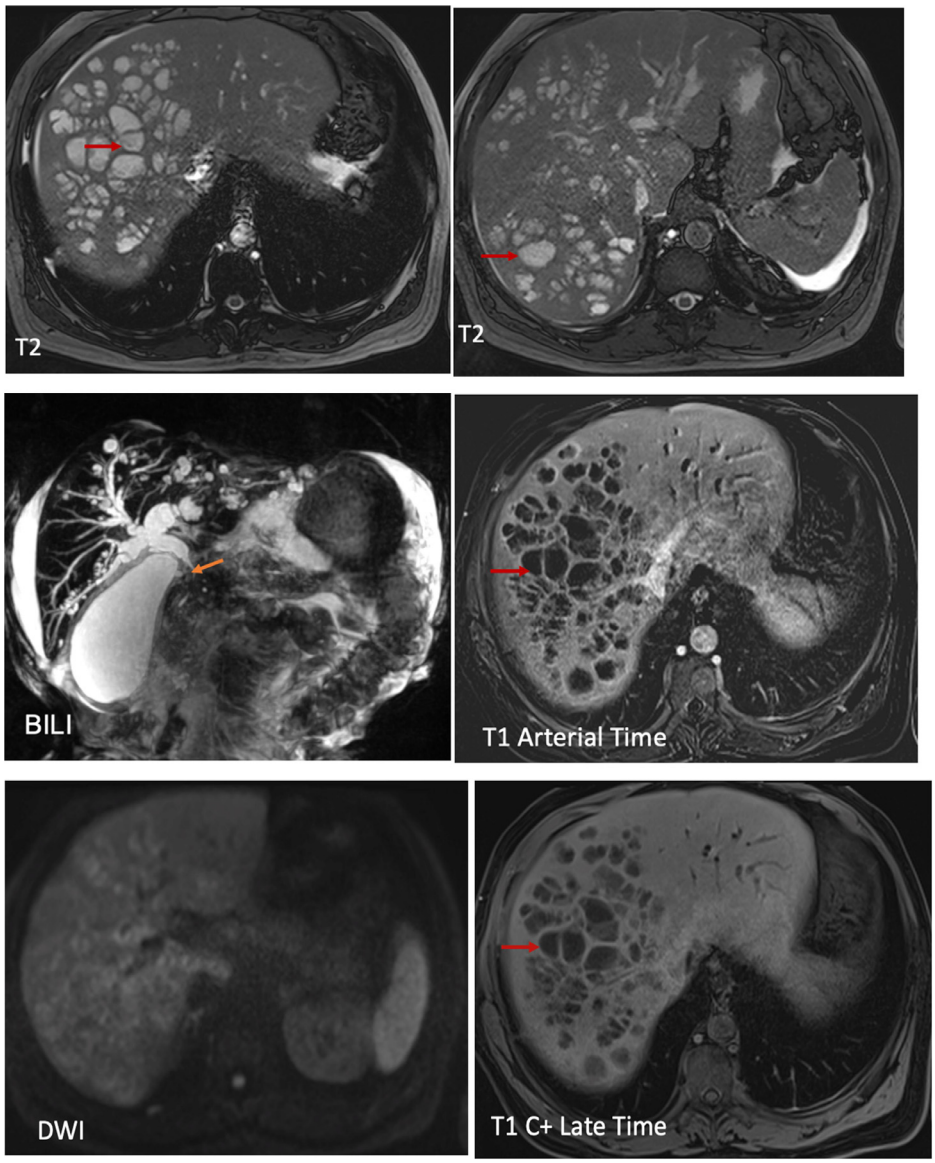

Fig. 2 – MRI in axial and coronal sections, showing multiple cystic lesions on either side of the portal branches (red arrow), described in T2 fluid hypersignal, nonrestrictive in diffusion, and not enhanced after arterial and late contrast. On the BILI sequence: dilatation of the intrahepatic bile ducts and the main bile duct upstream of a tumor-like stenosis of the middle part of the main bile duct (orange arrow).

function tests indicated renal failure (urea 2.3 mmoL/L, creatinine 17 μmoL/L).

Abdominal ultrasound revealed a liver of regular contours, normal in size and echostructure, and 1 or 2 cystic lesions in segments I and IV, with thin echogenic deposits and thickened walls, measuring 16 and 20 mm in length respectively, which were probably related to hepatic micro-abscesses, combined with dilatation of the intrahepatic and main bile ducts, with no visible obstruction.

A Bili MRI (Fig. 2) was performed, showing multiple small cystic lesions around the bile ducts with no clearly visible communication, described as T1 hyposignal, T2 fluid hypersignal, nonrestrictive in diffusion and unchanged after contrast, in relation to peribiliary cysts, associated with dilatation of the intrahepatic bile ducts, upstream of a tumour-like stenosis of the middle part of the main bile duct.

Discussion

Peribiliary cysts are also known as mucinous biliary hamartomas and hepatic hilar cysts. Nakanuma et al. [1] first reported these cysts in 1984, describing the pathological findings of 8 cases discovered during autopsy. The periductal tissues of the large bile ducts contain minimally dilated normal periductal glands, which are present in the same distribution as the cysts.

Several studies have identified several potential etiologies for peribiliary cysts. Most of them occur in the context of chronic liver disease, more frequently in cirrhotic patients, and are probably associated with disorders of the intrahepatic circulation, more likely the portal venous system [2]. In addition, peribiliary cysts are more common in patients with alcoholic liver cirrhosis than in those with other causes of cirrhosis [3].

In patients who were previously undetected, the detection of these cysts can help in the identification of an underlying chronic liver illness [4]. Additionally, a worsening of liver disease is indicated by the cysts' increasing size and quantity [4,5].

Based on these data, Nakanuma et al. [1] postulated that the glands' necks may have become occluded, leading to the formation of cysts, as a result of the disrupted portal circulation and related inflammatory alterations caused by thrombosis. There have been reports of both mucinous and senous [1,5] contents in these cysts.

A small percentage are linked to polycystic liver disease, with patients who also have concomitant autosomal dominant polycystic kidney disease being more likely to have this condition [6].

Cholangitis and obstructive jaundice are the most frequently reported complications. Due to the extrinsic compressive effect on the biliary tree, around 2% of patients may experience signs and symptoms of biliary obstruction [4]. Due to alterations in the epithelium of the peribiliary gland, these cysts may also behave as neoplastic precursors, potentially leading to cholangiocarcinoma and intraductal papillary neoplasm of the bile duct [5].

In an autopsy study, Sato et al. [7] found that up to 10% of patients had epithelial changes in the peribiliary glands. These changes may be precursors of cholangiocarcinomas and intraductal papillary neoplasms of the bile ducts, though it is unclear what their clinical significance is [8].

Multiple anechoic cystic lesions are commonly seen on ultrasounds next to the right and left branches of the portal vein, as well as the central intrahepatic bile ducts. But occasionally, in the same topography, only anechoic tubular lesions are found a result that mimics bile duct dilatation [6,7].

Multiple cysts or hypointense linear or tubular areas without contrast enhancement are seen on computed tomography (CT) scans, parallel to and on both sides of the portal vein branches. It is incorrect to interpret these results as bile duct ectasia [9,10].

Multiple clusters of tiny fluid-filled cavities with no interconnected biliary ducts are shown in periportal topography on MRI, with hypointense signal on T1-weighted sequences and hyperintense signal on T2-weighted sequences. Differential diagnosis of peribiliary cysts include biliary tract dilatation, primary sclerosing cholangitis, Caroli disease, biliary tract hamartomas and periportal edema [11]. High-morbidity procedures, unnecessary diagnostic tests and misdiagnosis can be avoided by taking imaging data and clinical background into consideration. As these lesions do not have contrast excretion on MRI using hepatobiliary contrast agents, it helps to demonstrate the noncommunication of these cysts with the lumen of the biliary tree, while biliary duct tracts show contrast excretion within their lumen.

Motoo et al. [12] demonstrated biliary stenosis due to extraluminal compression of enlarged peribiliary cysts using endoscopic retrograde cholangiopancreatography (ERCP). In fact, the growth of these cysts can result in extrinsic compression of the bile ducts, causing obstructive jaundice [13].

Conclusion

Peribiliary cysts can be identified by MRI, CT and ultrasound scans and are associated with a number of disorders. By correctly identifying the appearance of these cysts on imaging, it is possible to avoid the misdiagnosis of an abscess, cystic neoplasm or bile duct dilatation. On the other hand, these cysts may indicate one of the underlying related disorders when visualized on imaging.

Peribiliary cysts may be misdiagnosed, and their underlying predisposing factors may not be acknowledged.

Patient consent

Informed written consent was obtained from both patients for publication of the case report and all imaging studies.

REFERENCES

[1] Nakanuma Y, Kurumaya H, Ohta G. Multiple cysts in the hepatic hilum and their pathogenesis. Virchows Arch A Pathol Anat Histopathol 1984;404:341–50.

[2] Seguchi T, Akiyama Y, Itoh H, Tanaka H, Naganuma S, Nagaike K, Uchiyama S, Kataoka H. Multiple hepatic peribiliary cysts with cirrhosis. J Gastroenterol 2004;39(4):384–90. doi:10.1007/s00535-003-1307-4.

[3] Matsubara T, Sato Y, Igarashi S, Matsui O, Gabata T, Nakanuma Y. Alcohol-related injury to peribiliary glands is a cause of peribiliary cysts: based on analysis of clinical and autopsy cases. J Clin Gastroenterol 2014;48(2):153–9. doi:10.1097/MCG.0b013e318299c8c1.

[4] AlNuaimi D, Balci NC, AlDuaij A, AlKetbi R, Sierra MPR. A case of peribiliary hepatic cysts in a cirrhotic liver: a mimicker of Klatskin Tumor. Radiol Case Rep 2020;15(7):1039–43.

[5] Bazerbachi F, Haffar S, Sugihara T, Mounajjed TM, Takahashi N, Murad MH, et al. Peribiliary cysts: a systematic review and proposal of a classification framework. BMJ Open Gastroenterol 2018;5(1):e000204.

[6] Nakanuma Y. Peribiliary cysts have at least two different pathogeneses. J Gastroenterol 2004;39:407–8.

[7] Sato Y, Harada K, Sasaki M, Nakanuma Y. Cystic and micropapillary epithelial changes of peribiliary glands might represent a precursor lesion of biliary epithelial neoplasms. Virchows Arch 2014;464(2):157–63. doi:10.1007/s00428-014-1537-2.

[8] Uchida T, Yamamoto Y, Ito T, Okamura Y, Sugiura T, Uesaka K, Nakanuma Y. Cystic micropapillary neoplasm of peribiliary glands with concomitant perihilar cholangiocarcinoma. World J Gastroenterol 2016;22(7):2391–7. doi:10.3748/wjg.v22.i7.2391.

[9] Bächler P, Baladron MJ, Menias C, Beddings I, Loch R, Zalaquett E, et al. Multimodality imaging of liver infections:

differential diagnosis and potential pitfalls. Radiographics 2016;36(4):1001–23.
[10] Malekzadeh S, Widmer L, Salahshour F, Egger B, Ronot M, Thoeny HC. Typical imaging finding of hepatic infections: a pictorial essay. Abdom Radiol 2021;46(2):544–61.
[11] Mehta P, Prakash M, Khandelwal N. Radiological manifestations of hydatid disease and its complications. Trop Parasitol 2016;6(2):103–12.
[12] Motoo Y, Yamaguchi Y, Watanabe H, Okai T, Sawabu N. Hepatic peribiliary cysts diagnosed by magnetic resonance cholangiography. J Gastroenterol 2001;36:271–5.
[13] Maganinho P, Carvalho J, Coelho L, Amorim J. ARP case report n° 24: peribiliary cysts - an uncommon diagnosis. Acta Radiologica Portuguesa 2022;34:19–20. doi:10.25748/arp.26140.