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Cystic Diseases of the Liver and Bile Ducts

Kaitlyn Kelly · Sharon M. Weber

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Introduction

Hepatobiliary cystic lesions are more common than previously thought. With advances in modern cross-sectional imaging, the prevalence of hepatobiliary cystic lesions has increased from a presumed 2–3 % historically to 18 %.[1] The differential diagnosis of hepatobiliary cystic lesions is broad and it ranges from benign, asymptomatic lesions to infectious lesions to aggressive malignancies. Hepatobiliary cystic lesions can also be classified as congenital or acquired. Congenital lesions include simple cysts alone or in the setting of polycystic liver disease (PCLD), ciliated hepatic foregut cysts, and bile duct cysts (Caroli Disease). Acquired lesions include infectious cysts (abscesses and parasitic cysts) and neoplastic cysts, although some evidence suggests the latter may also be congenital. These lesions can be similar in presentation, but their management varies depending on the diagnosis. This evidence-based review will focus on the symptoms, diagnosis, treatment, risks, and quality and outcomes of therapy for hepatobiliary cystic lesions.

Symptoms

In many cases, hepatobiliary cysts are asymptomatic and are found incidentally on imaging for other reasons. This is especially true for simple hepatic cysts, although large simple cysts may produce abdominal pain, vague discomfort or fullness, early satiety, palpable mass, or abdominal distention. In the setting of PCLD, patients often present with a protuberant abdomen from massive hepatomegaly with or without associated autosomal dominant polycystic kidney disease.

Infectious hepatic cysts, including parasitic, amebic, and pyogenic abscesses, often present with fever, malaise, right upper quadrant pain, and even sepsis. Additionally, there is often a history of antecedent intra-abdominal infection or biliary tract manipulation. Pyogenic liver abscesses can result from hematogenous seeding from extra-abdominal infections, especially in immunosuppressed individuals. Parasitic liver abscesses are most commonly due to echinococcal infection, which is endemic in the Middle East, Asia, Australia, New Zealand, and South America.

Bile duct cysts are most frequently diagnosed in the pediatric population, but approximately 20 % will be present in adulthood. When present, symptoms usually mimic those of cholelithiasis/choledocholithiasis and include pain, tenderness, nausea, vomiting, fever, pruritus, and jaundice. Adults with bile duct cysts may also present with complications of the disease such as recurrent pancreatitis, hepatic fibrosis, cirrhosis, malignancy, and even portal hypertension.

Neoplastic hepatic cysts which include cystadenoma, cystadenocarcinoma, and intraductal papillary mucinous neoplasm of biliary origin (IPMN-B) are rare and are thought to account for <1 % of hepatobiliary cystic neoplasms.[2] Diagnosis is typically delayed for these lesions because symptoms are indolent and nonspecific. They include abdominal pain or discomfort, abdominal swelling, and jaundice. The mean time interval between symptom onset and diagnosis for these lesions is 1 year.[3] Solid hepatobiliary neoplasms and metastases to the liver can rarely undergo necrosis or cystic degeneration and present with the appearance of a cystic lesion. In these cases, patients may also present with fever/pyrexia and weight loss in addition to the symptoms described above. There is no defined staging

K. Kelly · S. M. Weber (✉)
University of Wisconsin Madison, Madison, WI, USA
e-mail: webers@surgery.wisc.edu

system available for cystic hepatobiliary neoplasia, and it is not possible to determine the presence or absence of malignancy based on preoperative imaging unless distant metastases are present. Cystadenomas/cystadenocarcinomas with ovarian stroma (OS) in the cyst wall occur exclusively in women and are thought to be the pathologic counterpart of pancreatic lesions of the same name. Biliary cystadenomas/cystadenocarcinomas without OS occur in both genders, but more commonly in men (2:1). It is unclear whether lesions without OS are truly a cystadenoma/cystadenocarcinoma or whether they are a cystic variant of cholangiocarcinoma or IPMN-B.

Symptomatology often overlaps among the different types of hepatobiliary cysts, and therefore, is usually not enough to establish a diagnosis. However, symptomatology combined with physical examination, imaging characteristics, and laboratory investigation are often enough to establish a diagnosis or at least significantly narrow the differential.

Diagnosis

Physical Examination

In cases of solitary hepatic cystic lesions not involving the biliary tree, physical examination may be entirely normal. Particularly large cysts may be palpable on exam if they extend below the costal margin, but this is unusual. A palpable mass that moves freely with respiration is a classic presentation of biliary cystadenoma. Patients with PCLD often have palpable hepatomegaly. Patients with pyogenic liver abscess may demonstrate abdominal tenderness from a site of intra-abdominal infection that lead to the abscess. Those with bile duct cysts may harbor sequelae of chronic liver disease or cirrhosis such as ascites, periumbilical varices, or spider angioma. Although benign cystic lesions may rarely exert pressure on the biliary tree, the presence of scleral icterus or jaundice on physical examination should raise concern for malignancy.

Imaging

The armamentarium for diagnosis and characterization of hepatobiliary cystic neoplasms includes ultrasound, cross-sectional imaging [computed tomography (CT) and magnetic resonance imaging (MRI)], magnetic resonance cholangiopancreatography (MRCP), and endoscopic retrograde cholangiopancreatography (ERCP). Cyst size is generally not helpful in establishing a diagnosis as the various types of hepatobiliary cystic lesions can all vary dramatically in size. There are other imaging characteristics that can be helpful however. General imaging characteristics of different hepatobiliary cystic lesions are outlined in Table 1.

Ultrasound is the imaging modality most often employed first for characterization of hepatobiliary cystic lesions. Simple cysts appear as smooth, well-circumscribed, anechoic lesions. They have a characteristically well-defined tissue-fluid interface with surrounding hepatic parenchyma that produces accentuated echogenic shadowing beyond the cyst itself. This is known as *acoustic posterior enhancement*, but is only observed when the tissue deep to the cyst transmits US. Septations are not present, but when multiple cysts are present, adjacent cyst walls may give a false appearance of septation. Liang and colleagues demonstrated a sensitivity and specificity of Doppler sonography in differentiating cystic malignancies from abscesses and simple cysts of 85 and 96 %, respectively.[4] Simple cysts complicated by intracystic hemorrhage or prior sclerotherapy present a diagnostic dilemma because they develop imaging characteristics similar to pyogenic abscesses, hydatid cysts, or cystic neoplasms.

The appearance of bile duct cysts also varies with cyst type, but most lesions can be well-characterized on US. The presence of wall thickening or nodularity is suggestive of malignancy. US can also demonstrate associated choledocholithiasis, strictures, and intrahepatic ductal dilatation if present. Extrahepatic bile duct cysts do not typically contain internal septations. This finding would suggest an extrahepatic cystadenoma or cystadenocarcinoma. Intrahepatic bile duct cysts appear as multiple cystic lesions immediately adjacent to major intrahepatic bile ducts and may be unilobar or bilobar.

Cross-sectional imaging with CT or MRI can be very informative in addition to US for evaluation of hepatobiliary cystic lesions. These studies can reliably demonstrate whether cystic lesions are solitary or multiple and are not as operator-dependent as US. CT is able to detect nodularity or septations within cysts but is not able to clearly detect intracystic blood clot or hemorrhage like US can. Discordant findings on US and CT regarding nodularity or septation therefore suggest hemorrhage and clot whereas concordance in these findings is suggestive of neoplasm.[5,6] Cross-sectional imaging also clearly defines the anatomic relationships of cystic lesions to the portal pedicles and hepatic veins. An imaging characteristic on contrast-enhanced CT or MRI important in the diagnosis of Caroli disease is the "central dot sign." This sign corresponds to vascular structures in bulbar protrusion of the cyst wall.[7]

Most hepatobiliary cystic lesions are T1-hypointense and T2-hyperintense on MRI. MRI is particularly helpful for characterization of hydatid cysts, especially when US is equivocal. On T2-weighted images, hydatid cysts exhibit a low-signal intensity rim that is characteristic of the outer, collagenous cyst membrane. Additionally, the presence of intracystic fat density on MRI suggests communication with the biliary tree.[8] MRI can also be helpful in distinguishing simple hepatic cysts with intracystic hemorrhage from neoplastic cysts. In a recent review of cystic liver lesions,

Table 1 Imaging characteristics of hepatobiliary cystic lesions

	Ultrasound	CT	MRI	ERCP/MRCP
Simple cyst/PCLD	Smooth, anechoic lesion without septations; thin-walled; posterior acoustic enhancement; often multiple[a]	Water-dense; smooth and round or oval; avascular	Homogenous; T1 hypointense/T2 very hyperintense	NA
Pyogenic abscess	Smooth, often hypoechoid lesion; thin or thick-walled; with variable internal echoes and debris; air within appears hyperechoic 80 % in right lobe	Single or multiloculated lesion with central hypodensity and peripheral contrast enhancement	T1 hypointense/T2 hyperintense lesion with hyperintense enhancing rim	NA
Hydatid cyst	Well-circumscribed lesions, content pattern varies with stage[34]; early: similar to simple cysts; advanced: laminated, concentric halos±hyperechogenic material (hydatid sand); late/inactive: amorphous and with wall calcification	Daughter cysts in rosette pattern	Low-signal intensity rim on T2-weighted images; daughter cysts hypointense to intracystic fluid on T1 and hyperintense on T2[35]	Useful for demonstration of biliary complications
Cystadenoma/ cystadenocarcinoma/ IPMN-B	Anechoic cystic lesion w/echogenic internal septations and internal papillary projections and mural nodularity	Multiloculated cystic lesion with well-defined, contrast-enhancing wall, often with wall nodularity	Multilocular cyst, T1-hypo- or isointense/T2 hyperintense; wall and internal septations enhance with contrast	No communication w/bile ducts unless fistulization for cystadenoma/ cystadenocarcinoma; communication more consistent with IPMN-B
Bile duct cyst	Peribiliary cystic lesions ± wall thickening/nodularity	Peribiliary cystic lesions ± wall thickening/ nodularity; central dot sign in Caroli Disease	Peribiliary cystic lesions ± wall thickening/ nodularity	Confirms communication with biliary tree; characterizes APBJ

[a] Diagnostic test of choice for pathology

hemorrhagic simple cysts consistently had hyperintense elements on T1- and T2-weighted MRI whereas neoplastic cysts were hypointense on both reflective of their proteinaceous content. In this series, no patients with hemorrhagic simple cysts had purely hypointense lesions on MRI.[5] Figure 1 demonstrates a T2-weighted MRI image showing two simple cysts (small arrows) and one complex hemorrhagic cyst (large arrow). The latter could be confused with a neoplastic cyst but does have hyperintense elements. Diffusion-weighted MRI can also be useful in distinguishing between simple cysts and hydatid cysts.[9]

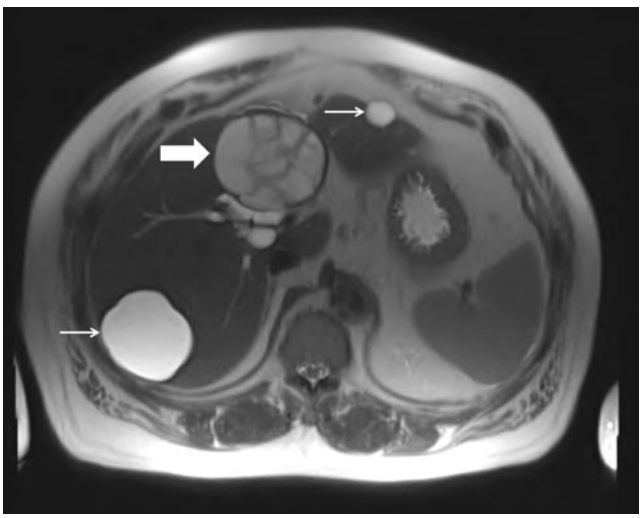

Fig. 1 T2-weighted MRI demonstrating a simple cyst with intracystic hemorrhage that could be confused with a neoplastic cyst (*large arrow*). In contrast, there are two simple cysts without hemorrhage visible in the same cut (*small arrows*)

When a bile duct cyst is suspected based on ultrasound or cross-sectional imaging, the diagnostic modality of choice is direct cholangiography. Cholangiography can elucidate the extent of the cyst and can identify other biliary pathology that is often associated with cysts including stenosis, stones, and nodules within the bile ducts. Additionally, cholangiography can define the anatomy of the common bile duct and pancreatic duct junction which is of utmost importance to avoid injury to the pancreatic duct during surgery for extrahepatic bile duct cysts.

The preferred method of cholangiography must be determined based on characteristics of the patient. ERCP is often favored because it can allow for clear definition of the pancreatic duct junction; brushings and biopsies can be performed for suspicious areas. Stricture dilatation, stone retrieval, and papillotomy can also be performed. Percutaneous cholangiography is the modality of choice for patients who have had prior roux-en-Y biliary-enteric anastomosis. It is also preferred in cases where stenoses or nodules in the extrahepatic ducts preclude imaging of the intrahepatic ductal system by ERCP.

Temporary external biliary drainage can also be placed at the time of percutaneous cholangiography if necessary for sepsis or relief of jaundice prior to surgery. Magnetic resonance cholangiography is an additional, noninvasive option that can accurately diagnose and characterize biliary cystic disease. It does not offer any therapeutic capabilities, however, and is inferior to ERCP in defining the anatomy of the pancreaticobiliary junction.

Laboratory Investigation

Patients undergoing evaluation for hepatobiliary cystic lesions should have a standard laboratory assessment including a complete blood count, basic metabolic panel, liver function tests, and prothrombin time/internationalized ratio. Additional lab tests should be obtained based on the suspected diagnosis. Simple hepatic cysts and PCLD will often have no associated laboratory abnormalities. Even with massive cyst burden in PCLD, the hepatic parenchymal volume is usually preserved.[10] Cyst fluid is typically acellular and is not helpful in distinguishing simple cysts from other cysts, including neoplastic cysts. Multiple recent studies have demonstrated that cyst and serum concentrations of CEA and CA19-9 in patients with simple cysts and cystadenoma are comparable.[11,12] Similarly, tumor marker concentrations are highly variable in cystadenocarcinoma and as in cases of pancreatic mucinous cystic neoplasms have not proven to be able to distinguish between cystadenoma and cystadenocarcinoma.[12,13]

Patients with infectious cysts may have leukocytosis and elevated alkaline phosphatase, gamma glutamyl transferase, and transaminases. Erythrocyte sedimentation rate and C-reactive protein are also frequently elevated. In a series of patients with PLA, 92.8 % had decreased serum albumin.[14] Another series demonstrated that low serum albumin, elevated creatinine, and prolonged PT were predictive of mortality in PLA.[15] Patients with parasitic cysts often have mild eosinophilia. Blood cultures should be obtained ideally before administration of antibiotic therapy in patients with suspected infectious cysts. Enzyme-linked immunosorbent assays (ELISA) aid in the diagnosis of parasitic cysts and can be highly sensitive. ELISA for galactose-inhibitable adherence protein has been shown to be >95 % sensitive for diagnosis of amebic liver abscess,[16] and sensitivity for diagnosis of hydatid disease varies depending on the antigens used.

Treatment, Risks, Quality, and Outcomes

In many cases, it is not possible to arrive at a diagnosis preoperatively. It can be difficult or impossible to determine whether a lesion is a simple cyst with hemorrhage, a ciliated hepatic foregut cyst containing mucin or with wall thickening, a hydatid cyst, an abscess, or a neoplastic cyst. If there is any uncertainty as to the diagnosis of a cystic lesion, surgical excision is recommended either by enucleation or formal resection. This will result in complete removal of the lesion and will allow for pathologic assessment of the cyst wall and contents. Resection should be performed without spillage of cyst contents into the peritoneal cavity. If a clinical diagnosis is able to be determined preoperatively, the treatment varies with the diagnosis.

Simple Hepatic Cysts

Asymptomatic simple cysts identified incidentally on imaging for other reasons do not require treatment or surveillance. If a simple cyst is identified on work up of vague abdominal pain and no other cause is identified, aspiration is recommended. If symptoms resolve after aspiration, it can be established that the cyst was the cause of pain and formal treatment can be undertaken. The primary treatment modalities for simple hepatic cysts include sclerotherapy and laparoscopic or open fenestration. Aspiration is not recommended as definitive therapy due to a nearly 100 % recurrence rate. Sclerotherapy involves cyst aspiration to achieve collapse, followed by injection of a sclerosing agent. The most commonly used agent is 95 % ethanol although others have been investigated (minocycline hydrochloride, ethanolamine oleate, and tetracycline). A key step prior to performing sclerosis is injection of the cyst with water-soluble contrast media to ensure no communication with the biliary tree or peritoneal cavity. Once such communication is ruled out, the cyst is injected with sclerosant and the patient is rolled in different positions to achieve maximal surface contact. Sclerosant retention times vary with different protocols but 10 to 20 min is standard, and the sclerosant is then aspirated.

An alternative to sclerotherapy for symptomatic cysts is laparoscopic or open fenestration or resection. In fenestration procedures, the cyst is opened to the peritoneal cavity, and a portion of the wall is excised flush with the adjacent liver parenchyma. This can be done with a GIA stapler, electrocautery, or a bipolar device. The remaining cyst epithelium can be treated with argon beam coagulation (ABC). One benefit of this approach is that at least a portion of the cyst wall can be sent for pathologic assessment. Cysts located superficially and in the anterior segments (III, IV, V, and VIII) are best suited to fenestration.

No randomized trials have been conducted comparing sclerotherapy and fenestration for simple cysts. Studies comparing the two treatment methods are small and include heterogenous groups of patients, but reported recurrence rates are higher after sclerotherapy (2 versus 75 %).[17] Many of these are only radiographic recurrences and do not require further therapy however. Sclerotherapy is a reasonable option for patients who are high risk for surgical complications and for those with posteriorly located or deep parenchymal cysts that

are not amenable to fenestration. A meta-analysis of retrospective studies comparing open versus laparoscopic fenestration demonstrated decreased blood loss and operative time, quicker return of bowel function, and shorter hospital stay after laparoscopic fenestration without an increase in perioperative complications.[18] The most common complications include wound infection, ileus, bile leak, and bleeding. A retrospective review of patients undergoing open fenestration, laparoscopic fenestration, or resection for hepatic cysts showed a superior perioperative outcome for laparoscopic fenestration but the lowest recurrence rate with resection.[19] We recommend laparoscopic fenestration as primary treatment for simple hepatic cysts, with open approach and resection being reserved for patients with deep or posterior lesions not amenable to laparoscopy. For patients at high risk for surgery, sclerotherapy is a reasonable option.

PCLD

In symptomatic patients with PCLD, the goal of therapy is to reduce liver volume to the greatest extent possible. In this setting, cyst recurrence is the rule. Medical therapies include octreotide, lanreotide, and sirolimus, all of which have been shown to achieve modest reductions in liver volume by inhibiting cyst fluid secretion. Additional options include aspiration and sclerotherapy, cyst fenestration, or liver resection in cases where one area of the liver is predominantly affected, and there is little to no functional parenchyma in that area. Liver transplant is rarely indicated for PCLD as the uninvolved liver parenchyma usually functions normally.

Operative treatment for PCLD must be individualized to each patient. Evacuation of superficial cysts often needs to be performed as an initial step to allow liver mobilization. If formal hepatic resection is performed, any cysts in the liver remnant should be opened and drained, and the lining should be treated with ABC. Recurrence rates of up to 70 % have been reported after sclerotherapy and fenestration for PCLD, but not all recurrences are symptomatic and long-term symptom relief is usually achieved with these procedures.

Pyogenic Liver Abscess

In patients presenting with PLA, blood cultures should be obtained immediately, and broad-spectrum IV antibiotics should be administered as soon as possible. If the abscess is secondary to an intra-abdominal infection, the source should be addressed (i.e., appendicitis, diverticulitis, cholangitis, etc.). As with any abscess, the key to successful treatment of PLA is drainage. Image-guided percutaneous drainage can be achieved by simple aspiration or catheter drainage and is generally recommended as the first therapeutic approach for PLA. Aspiration is well-suited for multiple small abscesses whereas catheter drainage is recommended for large abscesses with loculation. If percutaneous drainage fails, surgical drainage is required and can be performed via a laparoscopic or open approach.[19] Surgical drainage is recommended as the initial treatment strategy in patients who present with rupture into the peritoneal cavity, those undergoing laparotomy for source control, and those presenting with large, multiloculated abscesses in whom percutaneous drainage is likely to fail. Open drainage of PLA is difficult, however, because the majority of abscesses does not contain purulent material, but rather infected necrotic liver tissue. Therefore, open drainage is not preferred in most instances since it usually results in part.

In the past, liver abscesses were uniformly fatal. The mortality rate has decreased with development of modern drainage techniques, but is still high at 6–14 %. Potential complications of percutaneous drainage include biliary fistula, bleeding, or inadequate drainage.

Hydatid Cysts

Recommended treatment for hydatid cysts include chemotherapy combined with surgery or percutaneous-aspiration-injection and re-aspiration (PAIR). The technique involves image-guided percutaneous cyst aspiration which achieves collapse and confirms whether or not a communication with the biliary tree exists, injection with scolicidal agent (hypertonic saline or absolute alcohol), re-aspiration, and final irrigation. A drainage catheter may be left in place for larger cysts, and superinfected cysts and are removed once output is nil (PAIR-D). PAIR/PAIR-D is also recommended for patients with significant underlying liver pathology. Chemotherapy alone is not recommended unless patients are not candidates for surgery or PAIR or unless systemic hydatidosis is present. Abendazole is the mainstay chemotherapeutic agent for echinococcus, and the mechanism of action is glycogen depletion. Mebendazole is an alternative option, but is not as effective in vivo.[20] Chemotherapy is given anywhere from 2 weeks to 6 months prior to intervention (surgery or PAIR). There is no consensus on the duration of preoperative medical therapy. It is also advisable to administer 100 mg hydrocortisone at the time of intervention for hydatid cyst disease to decrease the risk of anaphylaxis.

The preferred surgical approach is pericystectomy or complete cyst excision with a rim of surrounding hepatic parenchyma. The abdomen is packed with laparotomy pads soaked with Betadine or hypertonic saline to isolate the cyst. The cyst is then aspirated with a large-bore needle to evaluate for cyst-biliary communication and to decrease intracystic pressure. If there is no biliary communication, the cyst can then be injected with 10 % Betadine solution to sterilize the contents prior to resection. For cysts with biliary communication, formal hepatic resection or localization of the communication

with suture ligation is required. Removal of excess hepatic parenchyma for benign disease should be avoided if possible however. A more conservative surgical option for cysts close to major pedicles is controlled drainage and partial cystectomy. Post-procedure chemotherapy is only indicated in cases where cyst contents have spilled.

Randomized, prospective trials to determine optimal management of hydatid disease are feasible in areas where *Echinococcus* is endemic. Unfortunately, there are no randomized trials to date comparing PAIR and surgery or conservative versus more radical surgical techniques, but all are effective therapies with comparable recurrence and complication rates.[21] Perioperative complication rates with resectional techniques vary from 18 to 25 % and common complications include biliary fistula, abscess, bleeding, and ileus. A large meta-analysis showed a statistically significantly higher recurrence rate (6 versus 2 %) and higher rate of major complication (25 versus 8 %) with resection versus PAIR, but this data is heavily biased by selection.[22] Another retrospective study comparing conservative surgery (unroofing, drainage, and/or marsupialization) with radical surgery (pericystectomy or hepatic resection) showed no increase in perioperative morbidity or mortality with more radical surgery. The authors recommend more aggressive surgical treatment of these lesions to decrease recurrence, biliary fistulization, and other cavity-related complications.[23] Choice of therapy should be tailored on an individual patient basis. Gharbi types I and II cysts can be effectively managed with PAIR. For more complex lesions where resection is required, current data suggest that more radical resection is preferred. A randomized trial comparing open versus laparoscopic resection has been completed in Turkey (NCT01643018NCT01643018). The primary endpoint of this study is cyst recurrence at 24 months. Secondary endpoints include intra- and perioperative complications, mortality, and postoperative pain score. Results are not yet available. A recently published trial from Turkey compared different techniques for management of the cyst cavity following surgical cystectomy.[24] Fifty patients were randomized to external tube drainage of the cavity ($N=28$) or omentoplasty ($N=22$). Omentoplasty resulted in decreased complications (23 versus 43 %), decreased postoperative pain, shorter hospital stay, and decreased time to resumption of activities of daily living. The authors concluded that omentoplasty is the preferred option when possible.

Neoplastic Cysts

Neoplastic hepatic include biliary cystadenoma, cystadenocarcinoma, and the more recently recognized IPMN-B. There is as of yet no way to distinguish between cystadenoma, a premalignant lesion, and cystadenocarcinoma in the absence of metastatic disease. Regardless, both lesions should be treated with complete resection by enucleation, partial hepatectomy, or bile duct resection with bilioenteric reconstruction for extrahepatic lesions. Partial resection and drainage is inadequate due to high recurrence rates.[25,26] If cystadenocarcinoma is known or highly suspected, partial hepatectomy with a margin of normal tissue is recommended over enucleation. For IPMN-B, intraoperative cholangiogram is recommended to evaluate for the presence of mucin in the bile ducts and to determine superficial spread of papillary epithelial growth beyond the cystic lesion and into adjacent bile ducts. Formal hepatectomy should be performed to ensure complete clearance of the lesion.

While reported surgical series are small, only one recurrence after complete resection of biliary cystadenoma has been reported in the literature.[27] Cystadenocarcinomas do exhibit recurrence and metastasis even after complete excision with negative margins. The variant without OS appears to be more aggressive than that with OS.[28,29] Surveillance is therefore indicated after resection of biliary cystadenocarcinoma, but given the rarity of this disease, an optimal surveillance schedule has not been defined. Additionally, no adjuvant therapy has been shown to be effective. Approximately, 70 % of patients with resected cystic IPMN-B have in situ or invasive adenocarcinoma. After complete resection, 5-year survival ranges from 60 to 80 % depending on histologic subtype.[30] Optimal surveillance schedule after resection of IPMN-B also remains to be defined.

Bile Duct Cysts

Management of bile duct cysts depends on the cyst type. The recommended treatment for type I cysts is complete excision of the extrahepatic cyst to and including an associated abnormal pancreaticobiliary junction (APBJ) and hepaticojejunostomy or hepaticoduodenostomy. If the posterior cyst wall cannot be separated from the portal vein due to inflammation or scarring, the wall can be left in situ and mucosectomy performed to remove all cyst epithelium. Type II cysts can simply be excised in the same fashion as a cholecystectomy if no APBJ is present. In the case of associated APBJ, type II cysts should be resected to and including the APBJ with hepaticojejunostomy like type I cysts. The recommended treatment for type III cysts is endoscopic papillotomy alone given the very low risk of malignancy in these lesions.

For type IV cysts, the extrahepatic component is approached like a type I cyst with excision and hepaticojejunostomy to the bile duct confluence. Type IVA cysts are frequently complicated by intrahepatic ductal strictures, stones, abscess formation, and malignancy. A thorough evaluation of the intrahepatic ductal system for anomalies is of paramount importance for successful treatment. Hilar stenoses must be recognized and excised circumferentially at their bases prior to bilioenteric anastomosis. The intrahepatic component is

often limited to individual hepatic segments and can be managed with segmentectomy. Type IVB cysts are approached like type I cysts with the addition of transduodenal sphincteroplasty for the choledochocele component. There are multiple reports describing laparoscopic and robotic resection of choledochal cysts, including primarily type I and type IV lesions.[31–33] The laparoscopic approach appears to be safe and effective in highly selected patients.

Management of type V cysts, or Caroli Disease, is based on the distribution of intrahepatic cysts and on the degree of underlying liver dysfunction. Liver transplantation is the only definitive treatment for patients with diffuse disease, and it should be undertaken at an early stage before complications of cholangitis, liver abscess, and cholangiocarcinoma develop. For patients with disease isolated to one lobe or segment and with sufficient hepatic functional reserve, resection of the affected lobe or segment may be effective.

Summary

Hepatobiliary cystic lesions encompass a range of benign and malignant conditions. Clinical presentation, laboratory investigations, and imaging can often lead to a diagnosis prior to surgical intervention. In more complex cases, patients should be presented and discussed in a multidisciplinary setting. When a diagnosis cannot be ascertained preoperatively, resection of the lesion (by enucleation or formal hepatectomy) should be performed to prevent peritoneal seeding of echinococcus or malignancy.

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CME/MOC Questions – 8 multiple choice (A-D)

1. What is the single most useful imaging modality for characterization of hepatic cystic lesions:

A) MRI
B) CT
C) ERCP
D) Ultrasound

2. What is the recommended treatment for an incidentally-identified, asymptomatic 12 cm simple cyst of the liver in segment V?

A) Laparoscopic fenestration
B) Open fenestration
C) Observation
D) Aspiration and sclerotherapy

3. What is the recommended treatment for an incidentally-diagnosed type I choledochal cyst in a 38 year old man?

A) Complete resection with hepaticojejunostomy
B) Observation
C) ERCP and sphincterotomy
D) ERCP and stent

4. What is the first step at operation for hydatid cyst disease?

A) Evacuation of superficial cysts to allow for liver mobilization
B) Packing around the liver with hypertonic saline-soaked pads
C) Injection of cysts with proscolicidal agent
D) Liver mobilization

5. What is the best imaging modality to distinguish between a biliary cystadenoma and a cystadenocarcinoma?

A) Ultrasound
B) There is no way to distinguish between these entities based on imaging
C) CT
D) MRI/MRCP

6. What is the incidence of in situ or invasive cancer in IPMN-B lesions?

A) 20 %
B) 70 %
C) 100 %
D) 50 %

7. CT and ultrasound are discordant on the presence of mural nodularity within a hepatic cyst. What does this indicate?

A) Intracystic hemorrhage/clot
B) Mural nodule not visualized on ultrasound
C) Calcification
D. Presence of a daughter cyst

8. What subtype of biliary cyst has the lowest risk of malignancy?

A) Type I
B) Type II
C) Type III
D) Type IV

Answers:
1. D
2. C
3. A
4. B
5. B
6. B
7. A
8. C