Choledochal cysts

Part 2 of 3: Diagnosis

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Much about the etiology, pathophysiology, natural course and optimal treatment of cystic disease of the biliary tree remains under debate. Gastroenterologists, surgeons and radiologists alike still strive to optimize their roles in the management of choledochal cysts. To that end, much has been written about this disease entity, and the purpose of this 3-part review is to organize the available literature and present the various theories currently argued by the experts. In part 2, we explore the details surrounding diagnosis, describing the presentation and imaging of the disease.

Dans une large mesure, l'étiologie, la pathophysiologie, le cours naturel et le traitement optimal de la maladie kystique de l'arbre biliaire continuent de faire l'objet de débats. Les gastroentérologues, les chirurgiens et les radiologues cherchent toujours à optimiser leur rôle respectif dans la prise en charge des kystes du cholédoque. C'est pourquoi les chercheurs ont beaucoup écrit à propos de cette entité morbide, et le présent examen en 3 parties a pour objet d'organiser les études publiées et de présenter les diverses théories que font actuellement valoir les experts. Dans la deuxième partie, nous examinons les détails entourant le diagnostic, en décrivant la présentation et l'imagerie de la maladie.

Choledochal cysts (CCs), which are cysts that can occur anywhere along the biliary tree, can cause clinically important mortality and morbidity unless diagnosed early. They can mimic many other conditions clinically, and can often be missed unless vigilantly included in differential diagnoses of certain common presenting symptoms such as abdominal pain and jaundice. This second part of our 3-part comprehensive review explores the common presentations and imaging strategies of biliary cystic disease to facilitate a correct diagnosis.

Presentation

Clinical presentation can occur at any time, but 80% of patients present before the age of 10 years. The classic triad of symptoms, consisting of abdominal pain, jaundice and a palpable abdominal mass, occurs in less than 20% of patients, although almost two-thirds of patients present with 2 of the 3 symptoms.[1–16] Symptomatically, patients should be divided into 2 groups: neonatal (age < 12 mo) and adult (age > 12 mo, as defined in the literature). Neonatal patients generally present with obstructive jaundice and abdominal masses, whereas adult patients present most commonly with pain, fever, nausea, vomiting and jaundice.[4–15,17–19]

Symptoms associated with CCs are usually due to the associated complications of ascending cholangitis and pancreatitis.[1–15,17–20] Complications associated with all types of CCs result from bile stasis, stone formation, recurrent superinfection and inflammation. Both dilated cysts and ductal stricture caused by chronic inflammation lead to proximal bile stasis, which in turn leads to stone and sludge formation and infected bile. Both of these factors lead to ascending cholangitis and further obstruction, resulting in the classic symptoms of episodic abdominal pain, fever and obstructive jaundice.[9,15] Stone and protein plug formation in the distal common bile duct and pancreatic duct causes

obstruction and resultant pancreatitis.[21–26] Protein plug formation may be due to chronic inflammation and the formation of albumin-rich exudate or the hypersecretion of mucin from dysplastic epithelium.[27] Recurrent cholangitis in patients with type-IV A and type-V cysts is thought to be due to persistent bacterial colonization of the intrahepatic dilations and exacerbated by the presence of bile stasis, sludge and stones.[28–35] As the cysts are difficult to eradicate short of total excision and liver transplantation, these complications tend to be lifelong and may progress to liver abscess and life-threatening sepsis.[36–38] The obstruction and infections in all CCs, especially those with intrahepatic involvement, also lead to secondary biliary cirrhosis in 40%–50% of patients, such that patients can also present with signs and symptoms of portal hypertension such as upper gastrointestinal bleeds, splenomegaly and pancytopenia.[33,35,39–41] Portal hypertension can also occur without cirrhosis, in which case the cyst can mechanically obstruct the portal vein.[42,43] Bile stasis can also lead to acalculous cholecystitis.[44]

About 1%–12% of patients with CCs present with spontaneous rupture and symptoms and signs of abdominal pain, sepsis and peritonitis.[45] The condition can be diagnosed when bilious paracentesis fluid is observed, bile-stained ascites are found intraoperatively and there is peritoneal entry of contrast seen on hepatobiliary iminodiacetic acid (HIDA) scan.[46–49] Ultrasounds may be misleading as the cyst may be decompressed from the rupture, and the biliary tree may thus appear normal. The cause of spontaneous rupture has been hypothesized to be caused by mural fragility from chronic inflammation, increased ductal pressure due to distal obstruction or raised intrabdominal pressure.[46] The site of rupture is often at the junction of the cystic and common bile ducts, as this is a site of poor blood flow.[46,49]

Although patients with choledochoceles can also present with the above complications, they are often asymptomatic. Type-III cysts can also cause gastric outlet obstruction either by directly obstructing the duodenal lumen or by intussusception.[50,51]

Diagnosis

When patients present with the symptoms described, the first step toward making the correct diagnosis is imaging. The first imaging modality generally used for the biliary tree is ultrasonography, which, with the exception of type-III and type-V cysts, will show a cystic mass in the right upper quadrant (usually at the porta hepatis) that is separate from the gallbladder. Diagnosis of a CC requires demonstration of continuity of the cyst with the biliary tree so that it can be differentiated from other intrabdominal cysts such as pancreatic pseudocysts, ecchinococcal cysts or biliary cystadenomas.[52] Although most authors recommend other imaging modalities for this purpose, Akhan and colleagues[53] demonstrated continuity with the bile duct in 93% of their patients and recommended other imaging only when the diagnosis cannot be made based on an ultrasound. Sensitivity of ultrasonography in making the diagnosis is 71%–97%.[54] Furthermore, given that ultrasonography is noninvasive and inexpensive, it is the modality of choice for follow-up surveillance. Reconstruction of 2-dimensional ultrasound images to form a 3-dimensional image has been advocated by some authors to view the cyst from different angles, allow full visualization of curved structures and estimate cystic volume, all of which may be important for preoperative planning.[55] Unfortunately, all ultrasonography is limited by body habitus, bowel gas and overlying structures.[55] Furthermore, the size of the cyst may be underestimated by suboptimal probe pressure.[56] Endoscopic ultrasonography has been proven useful as it does not have any of these limitations and allows good visualization of the intrapancreatic portion of the common bile duct.[57]

Another commonly used technique is a technetium-99 HIDA scan, which is recommended for viewing continuity with bile ducts.[58] This type of scan will show an initial area of photopenia at the cyst, with subsequent filling and then delayed emptying into the bowel. The sensitivity of HIDA scans varies with type of cyst (100% for type-I and 67% for type-IV A cysts[54]) owing to the inadequacy of HIDA scans in visualizing the intrahepatic bile ducts. Neonatally, it is important to differentiate a CC from biliary atresia, both of which can present as an obstructive cyst in the porta hepatis. Biliary atresia requires urgent surgical correction via Kasai portoenterostomy within the first few weeks of life and carries a very poor prognosis of progression to cirrhosis, liver failure and death.[59,60] Although it is difficult to distinguish a CC from biliary atresia on an ultrasound, a HIDA scan will show emptying of contrast into the bowel with CC, whereas retention of contrast owing to the distal obstruction indicates artresia.[56,60] In addition, HIDA scans are useful for the diagnosis of cyst rupture, as this will show entry of contrast into the peritoneal cavity.[57]

Computed tomography (CT) scans are useful in showing continuity of the cyst with the biliary tree, its relation to surrounding structures and the presence of associated malignancy. It is superior to ultrasonography in imaging the intrahepatic bile ducts, distal bile duct and pancreatic head.[58] In patients with type-IV A cysts and Caroli disease, it is useful to delineate the intrahepatic dilations and the extent of disease such as diffuse hepatic involvement versus localized segmental involvement. This is important, as localized type-IV A cysts or Caroli disease can be treated with segmental lobectomy. Malignancy can be identified as a mass or a focal region of wall thickening on a CT scan.[61,62] Some authors recommend spiral CT to differentiate malignant cyst wall changes from reactive inflammation.[63] Computed tomographic cholangiography (CTCP) has been used to delineate the full anatomy of the biliary

tree to correctly plan surgery; this imaging modality is 93% sensitive for visualizing the biliary tree, 90% sensitive for diagnosing CCs and 93% sensitive for diagnosing lithiasis. Unfortunately, it was reported to be only 64% sensitive for imaging the pancreatic duct, as this depends on reflux of the contrast into the ducts.[61] Virtual endoscopy based on CT images has been used to evaluate the biliary tree anatomy and identify defects successfully.[64,65] Intravenous cholangiography and spiral CT can be combined to form a 3-dimensional image that very accurately delineates the postoperative anastamosis site.[66] Of course, the drawbacks to using CT and CTCP is the risk of nephro- and hepatotoxicity with contrast and the exposure to ionizing radiation.

Cholangiography via endoscopic retrograde cholangiopancreatography (ERCP), percutaneous transhepatic cholangiography (PTC) or intraoperative cholangiography is necessary for completely delineating biliary anatomy preoperatively.[31] Cholangiography is also useful for identifying an abnormal pancreaticobiliary duct junction or ductal filling defects, which may be stones or cancers.[65] Although the use of cholangiography was previously ubiquitous in patients with CCs, it is slowly falling out of favour for a variety of reasons. For one, it is an invasive procedure with inherent risks of cholangitis and pancreatitis, which has been reported to be as high as 87.5% in patients with CCs.[66–68] Given that many patients with cystic disease have long common channels, dysfunctional sphincter mechanisms and dilated ducts, this risk is greater in these patients than in the general population.[68] Cholangiography also exposes the patient to ionizing radiation.[58,69] Although ERCP has been reported to be the most sensitive imaging modality for CCs, this sensitivity does fall in certain situations. Recurrent inflammation and scarring may make cannulation of the ampulla difficult or impossible and may cause partial or complete obstruction at any point of the biliary tree, with no resultant biliary imaging.[58] Full visualization of large cysts requires high dye load, and a compromise needs to be made between complete visualization and the risk of cholangitis or pancreatitis with increased amounts of dye.[60,68,70–72] The use of a high volume of dye can also cause intense opacification, thus obscuring mucosal defects such as ulcers or malignancy, as well as dilate the cyst and overestimate its volume.[68,72–74] Cholangiography is also not useful for postoperative imaging, as contrast is drained into the bowel without continuity to the hepatic duct.[64] Additionally, although ERCP can be performed safely in pediatric patients, the procedure requires the administration anesthesia.[75] Finally, the sensitivity of ERCP and the quality of images is operator-dependent.[60,76]

Given the concerns regarding cholangiography, magnetic resonance cholangiopancreatography (MRCP) is now considered to be the gold standard.[70,76–81] Magnetic resonance imaging (MRI) and MRCP create images by differential signal intensity of stagnant pancreatic and bile secretions compared with surrounding structures. Unfortunately, intraductal air, blood, debris, stones or protein plugs, all of which are common in patients with CCs, can interfere with the signal and alter visualization.[58] Nevertheless, sensitivity for diagnosis has been reported to be as high as 90%–100%.[82] Although breath holding manoeuvres were previously necessary to negate the interference of motion artifact, new technology allows for quicker procedures and eliminates motion interference, such that breath holding is no longer necessary. This allows more convenient imaging for adults and obviates the need for anesthesia in children.[72] Magnetic resonance cholangiopancreatography is 84% sensitive for imaging of postoperative anastamosis.[64] Unfortunately, sensitivity for assessing the pancreaticobiliary junction is as low as 46%–60%.[61,70,76,79,82] Magnetic resonance imaging is poor at imaging ducts or stones smaller than 5 mm and tortuous ducts.[70,76,83] Some authors suggest that the low sensitivity of MRCP in visualizing pancreaticobiliary junction is related to the small caliber of this junction, and they advocate the preimaging administration of secretin, which will increase pancreatic secretion and dilate the duct.[60] Magnetic resonance cholangiopancreatography is 20% less expensive than ERCP, although both modalities are twice as expensive as PTC.[58] Further advantages of MRCP over ERCP are that it avoids ionizing radiation; it is noninvasive and operator-independent; there are no complications of cholangitis and pancreatitis; and it can be coupled with MRI to image surrounding structures, lithiasis and malignancy.[69,76,77,81,84] Endoscopic retrograde cholangiopancreatography allows for the performance of therapeutic procedures, but this is only necessary with type-III cysts.[58]

Although all the information we have discussed so far pertains to the diagnosis of most CCs, type-III and type-V cysts deserve special consideration. Owing to their intramural nature, imaging abnormalities in choledochoceles are subtle, and the correct diagnosis is made preoperatively as little as 30% of the time.[85] Generally, multiple imaging modalities are required to make the diagnosis. Upper gastrointestinal series (UGIS) may show a filling defect where the cyst bulges into the duodenal lumen. Endoscopy and ERCP will show smooth bulging of the papilla, and cannulation will opacify the dilated intramural common bile duct.[58,86] Magnetic resonance cholangiopancreatography and CTCP have been advocated by some authors for diagnosis, but these modalities do not offer the option of performing sphincterotomy for treatment of the choledochoceles.[69,87–90] In contrast to other CCs, ultrasonography is not useful for the diagnosis of choledochoceles. The cysts are usually too small to visualize, and the normal diameter of the common bile duct makes connection to the biliary tree difficult to identify.[90,91] Endoscopic ultrasonography, however, has been used with much success, as it achieves close proximity to the cyst and is not as hindered by surrounding bowel gas as traditional

ultrasonography.[91–93] Differential diagnosis for type-III cysts includes duodenal diverticuli and duplication cysts. Diverticuli fill up with contrast in an UGIS and fail to opacify with ERCP. Duplication cysts will have identical images to choledochoceles and are therefore very difficult to differentiate. Some authors claim that a muscular wall is present in duplication cysts and absent in choledochoceles.[85]

In patients with Caroli disease, ultrasounds and CT and MRI scans show multiple saccular dilations, which can be focal or diffuse and contain bile, sludge and stones.[94] Computed tomography and MRI scans can also be used to diagnose associated cirrhosis, portal hypertension and varices, cholangitis, liver abscesses, malignancy and renal abnormalities.[61,76,81,95,96] Bloustein and colleagues[97] described the "central dot sign," which is a dilated duct surrounding a portal bundle, as pathognomic for Caroli disease.[98] Initially found on ultrasounds, this sign can also be seen on MRI and CT scans.[61,97] Although the central dot sign does suggest Caroli disease, it is not pathognomic, as it is also seen in obstructive dilation.[76] Also suggestive of Caroli disease is intraductal bridging, which involves echogenic septa traversing the duct.[61] A beaded appearance of the intrahepatic bile ducts on HIDA scan can be diagnostic.[76,83] The differential diagnosis for Caroli disease includes recurrent pyogenic cholangitis, polycystic liver disease and primary sclerosing cholangitis. Recurrent pyogenic cholangitis manifests as intra- and extrahepatic nonsaccular dilations with cast-like stones filling the entire lumen. Polycystic liver disease will have cysts that do not communicate with the biliary tree. Primary sclerosing cholangitis manifests as mild, focal fusiform dilations with obvious distal obstruction and is associated with inflammatory bowel disease.[61,84] These differences can help differentiate Caroli disease from other conditions.

Conclusion

The initial imaging of the biliary tree should be a simple ultrasound, and in most patients this will lead to the diagnosis of CCs. Cholangiography should then be performed to delineate biliary anatomy for operative planning. Although ERCP is commonly used for this, the risk of cholangitis warrants the use of MRCP instead whenever possible. Technetium-99 HIDA scans are useful in the neonatal period to differentiate congenital CCs from biliary atresia, as the ultrasound images in both diseases are similar. Computed tomography and MRI are useful modalities to diagnose and determine the extent of intrahepatic disease, such as type-IV A and type-V cysts. Once CCs have been diagnosed, careful treatment decisions need to be made. The third and final installation of this review series describes the management of biliary cystic disease.

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