Operative strategy based on imaging characteristics

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Lecture Choledochal cysts (CCs) have been considered congenital anomalies that present as abnormal cystic dilations of the intra and/or extra hepatic bile ducts. However, recently, the number of adult patients with CCs has been increasing due to improvements of noninvasive hepatobiliary imaging, with the reported incidence reaching up to 70%. In the western world, CCs is usually diagnosed in childhood. However, in Asian countries, adult CCs are particularly frequent, with frequency of 1 per 5000 in China and 1 per 1000 in Japan. Given the high risk of complications associated with adult CCs, including the development of cholangiocarcinoma, early diagnosis and treatment is very important. Definitive treatment of CCs is surgery and operative strategy is based on the classification of CCs. Therefore, in the present presentation, I'd like to describe classification of CCs and the operative strategy based on the classification.

Classification of choledochal cysts: current status

The most widely used classification system for CCs is the modified Todani system, which classifies CCs into categories I–V. Type I CCs (solitary extrahepatic cyst) are the most common type comprising approximately 50%–80% and are subdivided into 3 subtypes: Type IA CCs are the most common and are characterized by cystic dilation of the extrahepatic common duct; type IB CCs demonstrate focal, segmental dilation of the extrahepatic common duct and type IC CCs are characterized by smooth, fusiform dilation of the common bile duct (CBD) extending into the common hepatic duct (CHD) Ziegler et al. have suggested that choledochoceles should not be included in CCs classification due to their unique duodenal histology, location, and associations.
Type IV CCs are the second most common type, comprising 15-35% of all cysts and can be further subdivided into two subtypes based on their involvement of the intrahepatic and/or extrahepatic biliary ducts. While type IVA CCs are characterized by multiple cystic dilations of the both intrahepatic and extrahepatic bile ducts, type IVB cysts refer to multiple dilations of the extrahepatic common duct. It has been shown that preoperative imaging is unable to accurately predict true intrahepatic involvement in CCs, thus, it may be better to wait to make the distinction between type I and IVA cysts until after the cyst has been excised and postoperative imaging can then be used to determine which patients have true intrahepatic involvement.
Recently, several additional subtypes have been proposed to the Todani system. Type ID and type VI CCs are the newly proposed additions to the Todani system. Type ID is characterized by dilation of the cystic duct in addition to dilated CBD (type I) resulting in a bicornal configuration of the cyst. Type VI CC is manifested as an isolated dilation of the cystic duct without CBD or CHD involvement; this is extremely rare with only few reported cases.
There has been significant discussion regarding advantages and disadvantages of classifying CCs in a complex, alphanumerical system, and surgeons are moving toward a simplified classification scheme of CCs that is more directly relevant to guide management. For example, Visser et al. proposed that type I and type IVA cysts are simply variations of the same disease, citing that in their experience all type I cysts had some element of intrahepatic involvement. Furthermore, they argue that type II cysts are just diverticula of the CBD, more closely resembling gallbladder duplication than true CCs and choledochoceles should be thought of as variants of duodenal duplication cysts. Although Caroli disease resembles CCs morphologically, it has an difference pathogenesis and Caroli disease should no longer be thought of as subtypes of CCs. Martin et al. simplified classification, which is predominantly based on management approaches, classifies CCs as intrahepatic cysts, extrahepatic cysts, and intraduodenal cysts.

Operative strategy

Surgical resection, interventional therapy, and hepatic transplantation are the available treatment options for CCs and choice of management depends on the subtype and extent of biliary tract involvement. Complete excision of the cysts with some form of biliary reconstruction has become the gold standard for the treatment of the extrahepatic CCs and it is believed that incomplete cyst excision is associated with poorer clinical outcomes and a greater risk of malignancy. However, frequently there is no histological boundary between the lesion and normal bile duct, and the so-called radical excision is based on the morphology assessed by visual inspection during surgery. Furthermore, complete excision is not always achievable, especially in patients with widespread intrahepatic cysts. Recently, it is proposed that the establishment of proper bile flow exerted a greater impact on long-term outcomes than did complete excision. Thus, establishing proper bile flow can ensure good long-term outcomes in complicated cases of bile duct cysts and when complete excision is not feasible.

For type II CCs, some surgeons prefer excision with choledochoduodenostomy, whereas others prefer simple cyst excision with T-tube drainage. However, in patients with a type II cyst and anomalous pancreaticobiliary junction, the gallbladder should be removed because of the high risk of gallbladder malignancy, stressing the importance of accurate preoperative imaging.

For type III CCs, because of the rarity of this anomaly, standard methods for diagnosis and treatment have not been established. ERCP helps in diagnosis as well as management; endoscopic sphincterotomy followed by long-term endoscopic surveillance is the management of choice in these patients. However, some surgeon proposed cyst wall biopsy, and, if intestinal mucosa is present, they may be treated with cyst resection and sphincteroplasty. However, if a patient is found to have biliary epithelium within the choledochocele, a pancreaticoduodenectomy could be considered.

For type IVA CCs, a customized approach is needed, with a segmental hepatectomy and wide hilar hepaticojejunostomy for localized intrahepatic involvement and transplantation for symptomatic, diffuse intrahepatic involvement. Similarly, type V cysts are treated with segmental resection for unilobar involvement and liver transplantation for diffuse bilobar involvements complicated with cholangitis and/or portal hypertension.

Irrespective of subtype, all postsurgical patients require permanent, meticulous, long-term surveillance given the risk of cholangiocarcinoma and anastomotic strictures involving bilioenteric anastomosis

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