Endoscopic management of a large choledochocele associated with choledocholithiasis

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SUMMARY

Choledochocele or type III choledochal cyst is a very rare lesion, defined as a cystic dilatation of the distal common bile duct protruding into the duodenal lumen. Abdominal pain, biliary disorders, and acute pancreatitis are frequently observed but malignant degeneration is rare. A 70-year-old man had a history of epigastralgia associated with abnormal liver function tests suggesting gallstones. During laparoscopic cholecystectomy, intraoperative cholangiography showed a 40-mm-diameter choledochocele associated with choledocholithiasis. A transpyloric drain was placed after cholecystectomy had been completed. Endoscopic retrograde cholangiopancreatography confirmed the diagnosis and a 45-mm-long endoscopic sphincterotomy successfully treated both lesions as confirmed by a transpyloric cholangiogram showing a thin-walled common bile duct with no residual stones. This case illustrates that the diagnosis of choledochocele remains difficult in clinical practice and confirms that endoscopic retrograde cholangiopancreatography is the best available diagnostic tool. Coexistent choledocholithiasis is observed in about 20% of choledochoceles. Endoscopic sphincterotomy is feasible and effectively treats both lesions even in larger choledochoceles.

RÉSUMÉ

Prise en charge endoscopique d’un cholédochocèle de grande taille associé à un empierrement cholédocien

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Le chélydocèle ou kyste du cholédoque de type III est une lésion très rare définie par la dilatation kystique de la partie distale de la voie biliaire principale qui se prolève dans la lumière duodénale. Des douleurs abdominales, des symptômes biliaires ou une pancréatite aigüe sont fréquemment associés mais la dégénérescence maligne est exceptionnelle. Nous rapportons le cas d’un homme de 70 ans ayant des douleurs épigastriques associées à des perturbations biliaires et hépatiques supposées en relation avec des calculs vésiculaires. Au cours de la cholangiopancréatographie rétrograde, la cholangiographie peropératoire a montré un cholédochocèle de 40 mm de diamètre associé à un empierrement cholédocien. Après la cholecystectomie, un drain transpylorique a été laissé en place. Une cholangiopancréatographie rétrograde endoscopique a confirmé le diagnostic et permis la réalisation d’une sphinctérotomie endoscopique de 45 mm de longueur qui a traité les deux lésions avec succès. Une cholangiographie réalisée ultérieurement par le drain transpylorique a montré une voie biliaire fine sans aucun calcul résiduel. Ce cas confirme que le cholédochocèle est difficile à diagnostiquer et que la cholangiopancréatographie rétrograde endoscopique reste le meilleur examen. Des calculs du cholédoque sont observés dans 20% des cholédococèles, même en cas de cholédochocèle large, la sphinctérotomie endoscopique est faisable et efficace pour traiter les deux lésions.

Case report

A 70-year-old man treated with antiarrhythmic drugs and oral anticoagulants for cardiac dysrhythmia had had two attacks of epigastralgia and nausea in the past 6 months. The most recent attack was associated with abnormal liver function tests: aspartate transaminase 85 IU/mL (normal 5-40), alanine transaminase 90 IU/mL (normal 5-40), alkaline phosphatase 182 IU/mL (normal 25-90), γ-glutamyl transpeptidase 432 IU/mL (normal <40), and total bilirubin 1.5 µmol/L (normal <17). Upper abdominal ultrasonography disclosed a normal thin-walled gallbladder with stones but without common bile duct dilatation. The gallstones were assumed to be responsible for biliary symptoms and a laparoscopic cholecystectomy was planned. Intraoperative cholangiography showed a 40 mm diameter cystic dilatation of the distal common bile duct with coexistent choledocholithiasis (figure 1) and impaired flow of contrast into the duodenum. Five gallstones were extracted via the cystic canal and a transpyloric drain was left in place after cholecystectomy. The postoperative course was uneventful. Postoperative endoscopic ultrasonography was performed: duodenoscopy showed a dilated and protruding pancreatobiliary ampulla but ultrasonography did not help to confirm diagnosis due to obstructive stones preventing evaluation of cystic dilatation of the ampulla (figure 2). The diagnosis was confirmed at ERCP. Duodenoscopy showed the major duodenal papilla at the lower part of the choledochocele. After catheterization of the papilla, opacification showed a choledochocele with numerous stones (figure 3). A 45 mm-long endoscopic sphinctero-
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tomy was performed, allowing opening of choledochocele and total stone extraction via a Dormia basket. Biopsies were also performed revealing normal biliary mucosa on the inner wall of the choledochocele. The patient made an uneventful recovery from sphincterotomy. A transcystic cholangiogram performed at postoperative week 4 showed a thin-walled common bile duct with no residual stones (figure 4). The transcystic drain was taken off postoperative week 6 without any complications. At 6-months follow-up, the patient was clinically asymptomatic.

Discussion

Choledochoceles are usually diagnosed in adults (80% of cases) [3, 12] with no predominance for sex [10]. The initially reported mean age of 40 [3] was raised to 67 in the latest series [4]. Abdominal pain is the most common clinical feature. It is reported in 91% of cases and usually mimics biliary colic with nausea and vomiting as observed in our patient [3, 10]. Biliary disorders with predominant obstructive jaundice are present in 10-34% of cases [3, 4, 6, 10, 13]. Pancreatic disorders, mainly acute pancreatitis are reported in 30-38% of cases [3, 4, 6, 10]. Spontaneous rupture of larger choledochal cysts has been reported in childhood and postpartum but, to the best of our knowledge, this has never been recorded in Type III choledochal cysts [14]. As in 17-21% of cases [3, 4, 6, 10, 15], the choledochocele in our patient was associated with common bile duct lithiasis (figure 1 and 2). The risk of malignant degeneration initially reported to be as high as 15%, like in other choledochal cysts [2] is now thought to be much lower and estimated at about 2.5% [4, 16]. This lower risk is probably due to the frequent presence of a duodenal epithelium in the choledochocele [3] or to the absence

Fig. 1 – Intraoperative cholangiogram showing the dilated common bile duct with choledocholithiasis.

Cholangiographie peropératoire montrant le cholédoque dilaté siège d’un empierrement.

Fig. 2 – Endoscopic ultrasonogram showing numerous stones preventing analysis of cystic dilatation of the ampulla.

Échoendoscopie montrant les calculs qui empêchent l’analyse satisfaisante de la dilatation kystique de l’ampoule biliopancréatique.

Fig. 3 – ERCP showing choledocholithiasis associated with large choledochocele. Numerous stones are observed in the common bile duct.

CPRE montrant l’empierrement cholédocien associé à un large cholédochocèle. De nombreux calculs sont mis en évidence dans la voie biliaire principale.

Fig. 4 – Transcystic cholangiogram four weeks after ERCP and endoscopic sphincterotomy showing a thin common bile duct with no residual stones.

Cholangiographie par le drain transcystique 4 semaines après la CPRE et la sphinctérotomie montrant une voie biliaire fine sans image de calcul résiduel.
of massive pancreatic reflux into the biliary tree [17]. Moreover, the fact that nearly half of choledochocele patients undergo cholecystectomy before being diagnosed with choledochocele probably contributes to the lower malignant degeneration rate [3, 4, 6, 17].

Our case illustrates the difficulty of diagnosing choledochocele in patients, because this case was incidentally diagnosed during intraoperative cholangiography which is not generally used as a diagnostic procedure. It has already been reported that 30-58% of cases of choledochocele had a history of cholecystectomy [3, 4, 6]. Sarris et al. [3] have noted that choledochocele can be diagnosed at cholecystectomy if an intraoperative cholangiogram is performed. As in other reports, preoperative upper abdominal ultrasonography failed to demonstrate choledochocele in our patient [4, 18] and postoperative endoscopic ultrasonography did not confirm the diagnosis because multiple stones prevented visualization of the common bile duct.

However, this endoscopic examination has been reported to be diagnostic in some cases of choledochocele [19]. In the present case, duodenoscopy and ERCP corroborated the diagnosis, which confirms the previously reported diagnostic value of both of these procedures [20]. Nevertheless, noninvasive tools such as computed tomographic cholangiography [18], helicoidal CT scan [21] and magnetic resonance cholangiography which provides excellent mapping of the complete bile duct system [22] tend to be used. The etiology of choledochocele is still controversial. The classic notion of a congenital biliary anomaly has been questioned by some authors who argue that choledochocele is an acquired lesion from papilla dysfunction [7, 15]. Likewise, Elton et al. [9] has suggested that the dilated common channel syndrome, which is considered to be an acquired lesion related to papilla dysfunction, is an incomplete form of choledochocele. Indeed, most symptoms associated with choledochocele, such as obstructive jaundice or pancreatitis are obstructive, even in the absence of stones [4] and could be related to papillary obstruction. It has also been suggested that the stones observed in 20% of cases of choledochoceles develop in the common bile duct due to bile stasis and not in the gallbladder where stones are only observed in 12-30% [4]. However, the frequent finding of duodenal mucosa on the inner wall of other choledochoceles suggests a congenital duodenal abnormality, which rules out congenital biliary dilatation [3, 18]. In the present case, biopsies showed normal biliary mucosa, which supports a biliary origin. For these reasons, the existence of two forms of choledochocele i.e. congenital and acquired has been hypothesized [7]. Anatomical types of choledochoceles were defined by Sarris and Tsang [3]. In the most common type A (67% of cases), the pancreaticobiliary ampulla opens into the choledochocele, which communicates with the duodenum via another small opening, as observed in our patient. There are three anatomical variants with either a common opening (type A1), or two separate openings of the pancreatic and common bile ducts into the cyst (type A2 or A3). In type B (21% of cases), the choledochocele represents only a diverticulum of a normal pancreaticobiliary ampulla protruding into the duodenal lumen [3]. In the present case, the ampulla was dilated and communication with a common opening of the pancreatic and bile ducts, and the papilla at the lower part of the choledochocele was observed, corresponding to type A1.

Suggested surgical treatment included radical or partial excision of the cyst or transduodenal sphincteroplasty to allow pancreaticobiliary outflow and eliminate the risk of malignant degeneration [2, 3, 8]. This therapeutic strategy is now rarely used because the risk of malignant degeneration is very low, and surgery is being progressively replaced by endoscopic management. Initially described by Dehyle in 1974 [23], endoscopic treatment has become the procedure of choice [4, 7, 11] as it allows drainage of choledochocele treatment of presenting symptoms and relief of biliary stagnation, which is supposed to be partly respon-

sible for cyst degeneration [5]. Treatment includes either endoscopic sphincterotomy [4, 7] or cyst unroofing [11]. After these conservative treatments, the need for long-term endoscopic control of the choledochocele wall to detect malignant degeneration is controversial [5]. Nevertheless, surgical management of larger choledochoceles is still proposed by some authors and cystodudodenotomy remains the most adequate procedure [10]. However, in our opinion, the endoscopic sphincterotomy used in the present case is the best treatment for most choledochoceles, including larger ones. In addition, endoscopic treatment allowed concomitant treatment of choledocholithiasis with good morphological results confirmed by a postoperative transpyloric cholangiogram. This supports the widely approved treatment of choledochocele with endoscopic sphincterotomy and cholecystectomy [17].

In conclusion, choledochocele is a very rare lesion causing abdominal pain and frequent biliary and/or pancreatic disorders. The present case illustrates the difficulty of diagnosing this condition, which was diagnosed by chance during cholangiography and laparoscopic cholecystectomy and confirms the feasibility and efficacy of endoscopic sphincterotomy since choledochocele and choledocholithiasis were treated concomitantly with good clinical and morphological results confirmed by a postoperative transpyloric cholangiogram.

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REFERENCES


