Endoscopic management of choledochocele

A case report and review of the English literature

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Summary. Choledochocele or type III choledochal cyst is a rare lesion of the biliary tree. It may be of acquired or congenital etiology and can assume one of two anatomic variants. Either the common bile duct and the pancreatic duct enter the choledochocele together, or it is formed by the common bile duct alone with the pancreatic duct entering as a separate opening. The most frequent presenting symptoms are abdominal pain, pancreatitis, and jaundice. Traditional therapy has been either marsupialization of the cyst into the duodenum or complete surgical excision. This paper cites the eighth patient reported in the English-language literature whose choledochocele was treated endoscopically. It is the authors' opinion that either an endoscopic papillotomy or an endoscopic fistulotomy with extension of the incision over the cyst is the simpler and preferred method of treatment.

Key words: Choledochocele – Choledochal cyst – Pancreatitis – Jaundice – Endoscopic papillotomy – Endoscopic fistulotomy.

Since first described by Wheeler in 1940 [11], there have been sporadic reports of choledochoceles. It is a rare lesion and an easily overlooked cause of abdominal pain, pancreatitis, or jaundice. With larger lesions, the diagnosis may be made by an upper gastrointestinal series or by fiberoptic duodenoscopy. Smaller lesions may require cholangiography for diagnosis. Our recent experience with a patient and the endoscopic management of her choledochocele prompted a review of the literature pertaining to the etiology, pathology, and treatment of this entity. This is the eighth patient reported in the English-language literature for whom endoscopic management has been utilized.

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Fig. 1. Gastrointestinal series demonstrating a filling defect on the medial wall of the second portion of the duodenum (arrows)
Case report

The patient was a 75-year-old white female who had had a cholecystectomy and common bile duct exploration 40 years earlier. There had been no stones present in the gallbladder, but a stone had been removed from the common bile duct. The patient was asymptomatic for 1 year but then began to develop recurrent right upper quadrant pain, occasionally associated with nausea and vomiting and occurring approximately once a week. The patient denied any history of jaundice, cholangitis, or pancreatitis. Work-up during hospitalization in 1978 included an upper gastrointestinal series, which revealed a smooth, round, filling defect 2.0 cm in diameter on the medial wall of the second portion of the duodenum (Fig. 1). Intravenous cholangiography revealed a choledochocele. The patient refused surgery at that time but was readmitted in September 1984 for increasing abdominal pain occurring 2–3 times per week.

The patient had a hemoglobin of 12.8 mg/dL, a hematocrit of 37.9%, 9800 leukocytes, and a normal differential blood count. The sequential multiple analysis was normal, as was the serum amylase. The upper gastrointestinal series revealed a round filling defect in the second portion of the duodenum. Computerized axial tomography of the abdomen was not helpful. Abdominal sonography revealed a nontubular biliary tree.

The patient underwent an esophagogastroduodenoscopy, which revealed a 2-cm bulge on the medial wall of the second portion of the duodenum (Fig. 2). Endoscopic retrograde cholangiopancreatography was then performed. The pancreatic duct was first cannulated and appeared to be within normal limits and without communication into the cyst. Several unsuccessful attempts were made to cannulate the common bile duct. At this time, an endoscopic fistulotomy was performed over the most prominent and distal portion of the cyst. Bile flowed freely into the duodenum, and the choledochocele was decompressed. The common bile duct was then easily cannulated through the fistulous opening. The distal common bile duct measured 20 mm (Fig. 3). The fistulotomy was then extended to 1 cm over the most prominent portion of the cyst. There was no bleeding or other complication. The patient was placed on prophylactic antibiotics and intravenous fluids for 24 h. Oral feedings were started on the following morning. The patient was discharged on the 4th day after hospitalization. The patient has done well since and has been entirely pain free.

Discussion

Choledochocele was first described by Wheeler in 1940 [11]. He reported a patient whose biliary obstruction was caused by a diverticular lesion of the distal portion of the common duct and coined the term choledochocele for this entity. Alonso-Lej proceeded to classify abnormal dilatations of the biliary tree into three types in 1959 [1]. Type I is a choledochal cyst – a cylindrical or fusiform dilatation of the common bile duct usually associated with a distal narrowing of the common bile duct. The biliary radicles are of normal size (Fig. 4). Type II is a diverticulum of the common bile duct located above the intramural portion of the duct. This type is not associated with protrusion into the duodenal lumen. Type III is a cystic dilatation of the distal common bile duct with protrusion into the duodenum and is referred to as choledochocele. In 1975, Flanigan added Type IV – a cystic lesion in which there is dilatation of both the intrahepatic and extrahepatic portion of the biliary tree [2]. More recently, this type has been referred to as Caroli’s disease.

In reviewing the English literature, we have compiled the following statistics:

1. Choledochoceles represent 4.1% of all choledochal cysts.
2. Abdominal pain was the most common presenting complaint (94%), followed by pancreatitis (30%), jaundice (24%), anorexia and vomiting (14%), and gastrointestinal bleeding (10%).
3. Histologically, choledochoceles may be lined with duodenal mucosa (71%), biliary mucosa (25%), and both biliary and duodenal mucosa (4%).
4. Choledochoceles are of two anatomic variants: the first involves entry by only the common bile duct (79%), and the second by both the common bile duct and the pancreatic duct (21%).

The etiology of choledochoceles is unknown. Some investigators postulate a congenital origin, while others [9] believe it to be an acquired lesion. The majority of authors [3–5] believe that both explanations may be possible. An acquired origin is suggested by the presence of biliary mucosa lining the choledochocele. Biliary obstruction caused by either impacted stones or ampullary strictures leads to increased intraluminal pressure and herniation of the distal duct into the duodenal lumen. The argument for a congenital origin is supported by the presence of duodenal mucosa in the choledochocele lining. In such cases, a duplication of the duodenum or a walling-off of a communicating biliary cyst may cause the choledochocele.

The traditional therapy of these rare lesions has been surgical. The procedure most often recommended includes incision of the intraduodenal portion of the choledochocele, marsupialization of the wall, and a sphincteroplasty if obstruction to bile flow is suspected. In 1981, Powell recommended total excision of the cyst with reanastomosis of the pancreatic and common bile ducts to the duodenum [7]. A few recent reports have recommended endoscopic management [8, 10, 12]. We believe an endoscopic papillotomy should be performed, if feasible, for most of these lesions. If the papillotome cannot be inserted into the common bile duct, an endoscopic fistulotomy and extension of the incision over the cyst is the treatment of choice. Endoscopic surgery has proven to