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Diverticular Choledochal Cyst with a Large Impacted Stone Masquerading as Mirizzi's Syndrome

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Key Words
Choledochal cyst · Mirizzi's syndrome · Cholangitis

Abstract
Choledochal cysts are congenital anomalies of the biliary tract manifested by cystic dilatation of the extrahepatic and intrahepatic bile ducts. Choledochal cyst is not rare in far-East Asian countries. Type II choledochal cysts account for 2% of all such cysts. They are true diverticula of the extrahepatic bile duct and communicate with the bile duct through a narrow stalk. This condition is associated with significant complications, such as ductal strictures, stone formation, cholangitis, rupture and secondary biliary cirrhosis. We describe a case of a huge impacted stone in a diverticular choledochal cyst which masqueraded as an unusual cystic duct stone causing Mirizzi's syndrome.

Introduction

Choledochal cysts are congenital anomalies of the biliary tract manifested by cystic dilatation of the extrahepatic and intrahepatic bile ducts. Choledochal cysts are rare, and their incidence is higher in Asian than Western countries [1, 2]. The majority of them are diagnosed during the first decade of life, but around 20% are not diagnosed until adulthood [3]. Presentation of choledochal cysts in adults differs from their presentation in children in the way that these are more commonly associated with hepatobiliary pathology, such as cholangitis, cystolithiasis, pancreatitis, pancreatic duct abnormalities and malignancy. The widely accepted classification system for choledochal cysts, which was devised by Todani et al. [4], is based on cholangiographic morphology, location and number of intrahepatic and

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extrahepatic bile duct cysts. Type II choledochal cyst is a very rare true congenital diverticulum of the extrahepatic bile duct and a potential precursor of cholangiocarcinoma [3, 5–7]. Cystolithiasis and cholecystolithiasis are the most frequent conditions, occurring in 70% of adults with choledochal cysts [3]. However, the frequency of these conditions according to the type of choledochal cyst has not been estimated. Impacted stones are extremely rare in type II choledochal cysts and, to our knowledge, have not been reported in the English language literature.

We describe the unusual case of a huge impacted stone in a diverticular choledochal cyst which masqueraded as an unusual cystic duct stone causing Mirizzi's syndrome. The diagnosis in this case was confirmed by laparoscopic exploration.

Case Report

A 24-year-old male presented with recurrent abdominal pain that had started to worsen 2 months previously. He had no relevant family or past medical history. On admission, his vital signs were stable. Physical examination revealed slight tenderness in the epigastric area with no sign of peritoneal irritation. Biochemical investigations yielded the following findings: white blood cell count 6,300/mm^3, serum glucose 93 mg/dl, total bilirubin 1.1 mg/dl, aspartate aminotransferase 28 IU/l, alanine aminotransferase 13 IU/l, alkaline phosphatase 61 IU/l, gamma-glutamyltransferase 28 IU/l, amylase 115 IU/l, lipase 45 U/l, alpha-fetoprotein 2.07 ng/ml, carbohydrate antigen 19-9 23.68 U/ml, carcinoembryonic antigen 4.05 ng/ml and high-sensitivity C-reactive protein 0.3 mg/l.

Abdominal computed tomography showed a large stone in the common bile duct (CBD) with no marked upstream duct dilatation, and magnetic resonance cholangiopancreatography (MRCP) demonstrated the same stone at the same level with compression of the CBD, suggesting Mirizzi's syndrome. The location of the impacted stone was suspected to be the cystic duct rather than the CBD, which could cause compression of the CBD (fig. 1a). Subsequent endoscopic retrograde cholangiopancreatography (ERCP) showed that the large impacted stone was located in the central CBD. Injected contrast filled the dilated upstream cystic duct and CBD at the same time and demonstrated that the large stone was laterally compressing the CBD (fig. 1b). This finding strongly suggested that an impacted stone in the cystic duct of the gallbladder had caused Mirizzi's syndrome. Selective CBD cannulation was difficult due to the narrowing caused by extrinsic compression and variation in the common pathway of the CBD. However, the gallbladder was not distended and laboratory findings revealed no obstructive pattern typically seen in Mirizzi's syndrome. Following the insertion of a 7-Fr plastic stent (Cook Endoscopy, Winston-Salem, N.C., USA) into the CBD, laparoscopic cholecystectomy was planned.

Laparoscopic operative findings showed that the gallbladder was atrophic; thus, the surgeon planned to perform laparoscopic cholecystectomy and to remove the impacted stone via CBD exploration. Intraoperatively, the stone in the CBD was considered to be impacted due to the atrophic gallbladder. However, following removal of the gallbladder and the large impacted stone, surgical exploration revealed a bulging cystic lesion and the surgeon could not find the plastic stent inserted previously in the CBD (fig. 2). Thus, the surgery was converted to laparotomy, which revealed that the saccular dilated cystic lesion with a large impacted stone was a diverticular choledochal cyst. The type II choledochal cyst was excised and choledochoduodenostomy was performed. The final diagnosis was confirmed histologically, the specimen showing chronic inflammatory infiltration without dysplasia and denuded epithelium (fig. 3).

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Discussion

The estimated incidence of choledochal cysts in Western countries has ranged from 1/100,000 to 1/150,000 individuals. However, the incidence is higher in Asia, and these cysts occur more frequently in females (male:female ratio 1:4) [1, 2]. Type II choledochal cysts appear as diverticula of the CBD, with some cases closely resembling gallbladder duplication and others resembling rudimentary diverticular structures. Complications of choledochal cysts in adults include cholecystitis, recurrent cholangitis, biliary stricture, choledocholithiasis, recurrent acute pancreatitis and malignant transformation to cholangiocarcinoma [1–3]. Although abdominal pain is the most common presenting symptom in adults [3, 4, 8], most patients have nonspecific clinical symptoms [1, 4, 8, 9]. Choledocholithiasis is a common complication of choledochal cysts [2], but a diverticular choledochal cyst with an impacted stone causing diagnostic confusion is extremely rare and difficult to differentiate using imaging modalities, as described in our case.

ERCP has been regarded as the gold standard for the diagnosis of choledochal cysts and associated anomalies [10, 11]. Recently, MRCP has replaced ERCP for the assessment of many pancreaticobiliary diseases. MRCP allows visualization of the biliary and pancreatic ducts noninvasively and without contrast injection [11, 12]. Delineation of the precise anatomy of the pancreaticobiliary system is critical for optimal surgical management. Choledochal cyst types can be defined more accurately by MRCP than by ERCP [12]. However, the initial use of MRCP and ERCP in the present case led us to suspect that the patient had an unusual impacted cystic duct stone causing Mirizzi's syndrome. A large stone may be impacted at a low insertion point in a cystic lesion, which would not be captured by the basket in ERCP.

Currently, optimal treatment of a choledochal cyst is likely to involve complete excision of the extrahepatic bile duct, cholecystectomy and Roux-en-Y hepaticojejunostomy [8]. In the present case, laparoscopic cholecystectomy was first attempted based on radiologic and ERCP findings, but the surgery was converted to laparotomy due to unusual anatomic findings, including gallbladder atrophy above the level of the choledochal cyst, and failure to locate the previously inserted plastic stent. These findings eventually led to the diagnosis of a diverticular choledochal cyst with an impacted stone.

In summary, we experienced the very unusual case of a diverticular choledochal cyst with a huge impacted stone which masqueraded as a cystic duct stone causing Mirizzi's syndrome. The diagnosis was made via surgical exploration. In this case, preoperative diagnosis and treatment planning were difficult due to the limitations of imaging studies, including ERCP.

Disclosure Statement

All authors disclose no financial relationships relevant to this publication.

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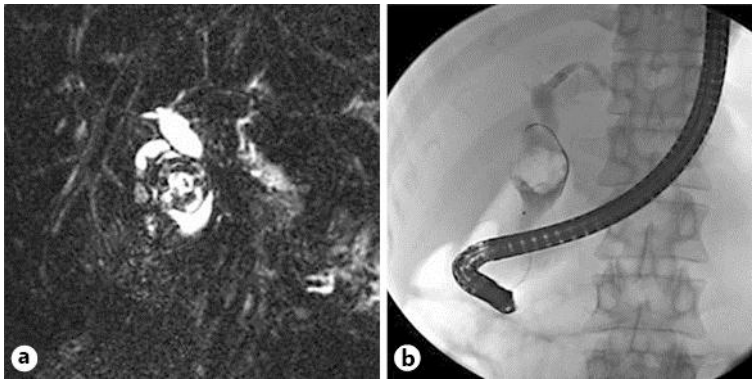

Fig. 1. **a** A 2.5-cm-sized heterogeneous SI lesion on the lateral side of the CBD which compressed the duct. **b** A huge lesion filled the middle level of the CBD and was suspected to be a portion of the cystic duct or gallbladder.

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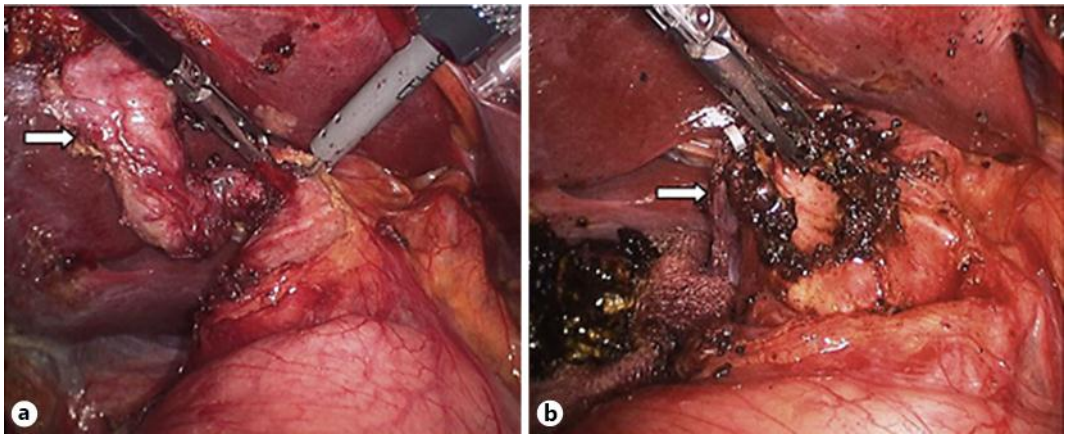

Fig. 2. **a** Laparoscopic view showing an atrophic gallbladder (arrow) and a bulging cystic portion with suspected impacted stones. **b** Following gallbladder removal, the cystic dilatation was incised to remove the impacted stone. Note the removed black stones and exposed choledochal cyst (arrow).

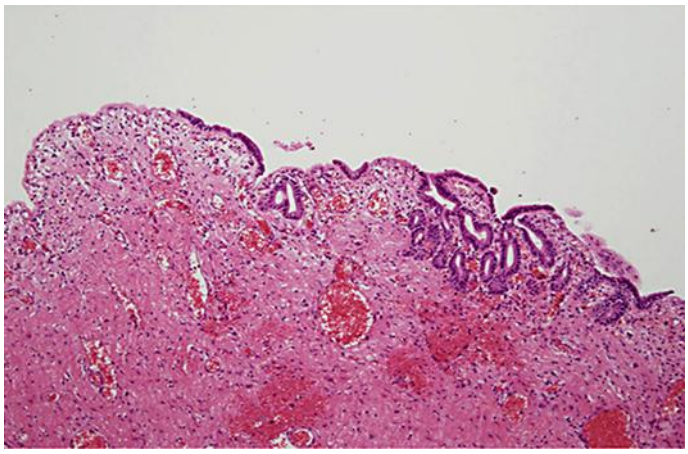

Fig. 3. Pathologic examination confirmed the diagnosis of a true choledochal cyst with no malignant change (H&E stain, ×100).