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A case report

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Choledochal cyst mimicking Mirizzi's syndrome. A case report

Choledochal cysts are cystic dilatations of the intra or extra-hepatic biliary tract with an incidence of 1 case per 150.000 live births. Cysts usually are diagnosed in childhood, but diagnosis can be delayed until adulthood in the 20-50% of cases. Clinical manifestations comprise abdominal pain with biliary or pancreatic features. Mirizzi's syndrome is a late and rare complication, that occurs in 1% of patients with cholelithiasis due to extrinsic compression of the common bile duct by stones impacted either in the gallbladder or in the cystic duct. Clinical symptoms include extrahepatic obstructive jaundice, ascending cholangitis, or, in the later course, cholecystocholedocal fistula. For both pathologies the Endoscopic Retrograde Cholangio Pancreatography and the Magnetic Resonance Cholangio Pancreatography should lead to the diagnosis with a sensibility and a specificity up to 100%. We report the case of a 66 year old patient admitted to the Emergency Department of our hospital for jaundice and abdominal pain, whom both the endoscopic and radiologic examination showed a Mirizzi's syndrome but surgery revealed a type I choledocal cyst.

KEY WORDS: Choledochal cyst, Mirizzi's syndrome, ERCP, MRI

Introduction

In patients presenting with obstructive jaundice, Mirizzi's syndrome (MS) and choledochal cyst (CC) are part of the differential diagnosis.
MS is a rare benign complication, seen in 1% of patients with long-standing cholelithiasis [1]. It has a wide spectrum of manifestations, ranging from gallstone impaction with biliary obstruction, to cholecystocholedochal fistula, and sometimes to complete erosion of the common hepatic duct [2].
The importance and implications of this condition are related to their associated and potentially serious surgical complications, such as bile duct injury, and to its management when encountered during laparoscopic cholecystectomy [3,4].
The pathophysiological process leading to the subtypes or stages of MS has been explained as an inflammatory phenomenon secondary to a pressure ulcer caused by an impacted gallstone at the gallbladder infundibulum. The impacted gallstone along with the inflammatory response, causes external obstruction of the bile duct first, and eventually erodes into the bile duct evolving into a cholecystocholedochal or cholecystohepatic fistula with different degrees of communication between the gallbladder and bile duct. The clinical presentation of MS is unspecific, being obstructive jaundice the most common clinical presentation of MS (60%-100%), accompanied

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by abdominal pain over the right upper abdominal quadrant (50%-100%) and fever, in the context of a patient with known or suspected gallstone disease [5,6].
CC is a cystic dilatation of the biliary tract with an incidence of 1 case for 100.000-150.000 live births [7]. It is usually diagnosed in childhood, but the diagnosis is delayed until adulthood in 25-50%, is more commonly seen in Asia, particularly in Japan, and has a female predominance [7,8]. The cause remains unknown and the most credible hypothesis presently presumes abnormalities in the pancreatobiliary junction [9]. The presence of a long common channel formed by the pancreatic duct and the common bile duct may activate pancreatic enzymes, causing inflammation and eventually dilatation of the biliary duct wall [9]. Great pressure in the pancreatic duct can also contribute to further dilatation [10]. These cysts are clinically important because of their potential complications such as cholangitis, choledocholithiasis, acute pancreatitis and, mainly, malignant transformation [11]. The risk of cholangiocarcinoma is up to 26% in some studies and the rate of occurrence increases with age [12-14]. The classic triad of symptoms (jaundice, right upper quadrant pain, abdominal mass) is not present in more than 20% of cases [15]. Complete cyst excision should be performed soon after diagnosis, since surgical intervention reduces or perhaps eliminates the risk of cholangiocarcinoma [16].
The clinical differential diagnosis between MS and CC is difficult, since there are no pathognomonic patterns of presentation. Endoscopic Retrograde Cholangiopancreatography (ERCP) and Magnetic Resonance Cholangiopancreatography (MRCP) have a high sensibility in the diagnosis of both conditions, but their ability in making a differential diagnosis is not reported in the literature, to our knowledge.
We describe an unusual case of a patient with a CC misdiagnosed as a MS based on ERCP and MRCP findings.

Case report

A 66 year old man was admitted to our Emergency Department with colic pain in the right upper abdominal quadrant. The pain had started 48h earlier and was associated with nausea, recent change in urine color (bright orange) and jaundice. The patient was apyretic. Physical examination revealed pronounced tenderness over the right hypochondrium and hypoactive bowel sounds. Lab tests showed: total bilirubin 9,7 mg/dl with direct fraction 5,1 mg/dl; aspartate aminotransferase 121 U/I; alanine aminotransferase 158 U/I; alkaline phosphatase 195 U/I; gamma-glutamyltransferase 871 U/I. All other lab parameters were within the normal range. Abdominal ultrasound showed dilatation of the intra- and extrahepatic bile ducts without an apparent obstructing lesion. An hyperechogenic image (with posterior shadow cone) was seen in the fundus of the gallbladder

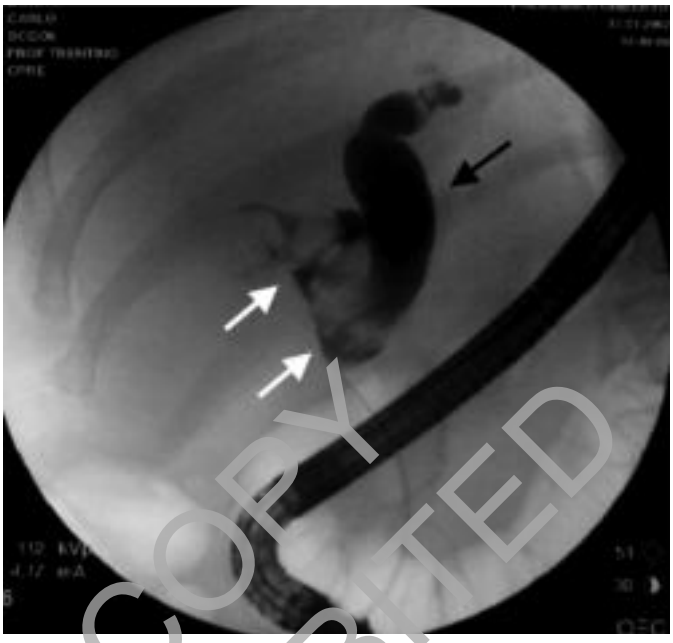

Fig. 1: ERCP image: it is clearly visible the dilatation of the common hepatic duct (black arrow). Multiple stones located in the gallbladder infundibulum looked to be the cause of a compression of the CBD (white arrows).

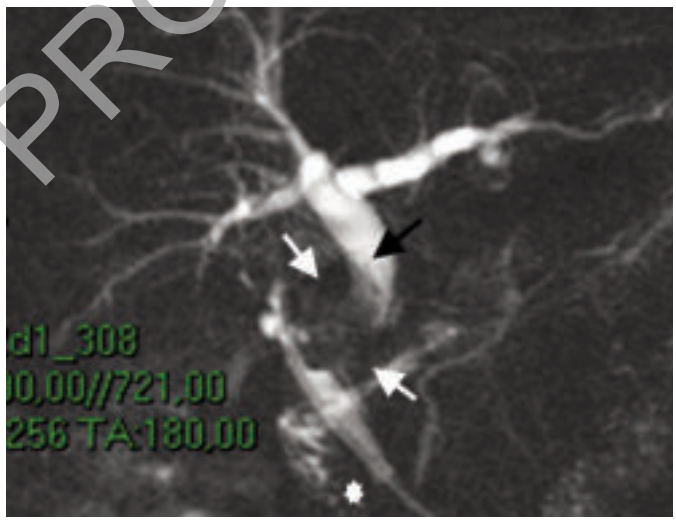

Fig. 2: MRCP image: it reveals dilatation of the common hepatic duct (black arrow) with some gallstones (white arrows) in the infundibolo-cystic duct and a plastic stent inside a non-dilated common bile duct (white star).

and the wall of the infundibular-cystic duct junction of the gallbladder showed diffuse thickening. On the following day an ERCP was performed for a more precise diagnosis. Cholangiography showed 15 mm dilatation of the common hepatic duct until the insertion of a very short cystic duct and then a normal-sized common bile duct (CBD). Some stones located in the gallbladder infundibulum looked to be the cause of a compression

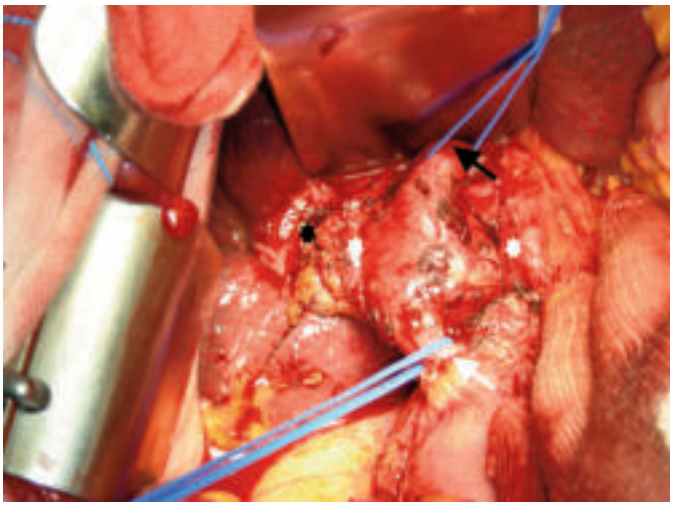

Fig. 3. Intraoperative picture: it becomes evident the choledochal cyst (between white stars), the scleroatrophic gallbladder (black star), the common hepatic duct (black arrow) and the choledocus (white arrow).

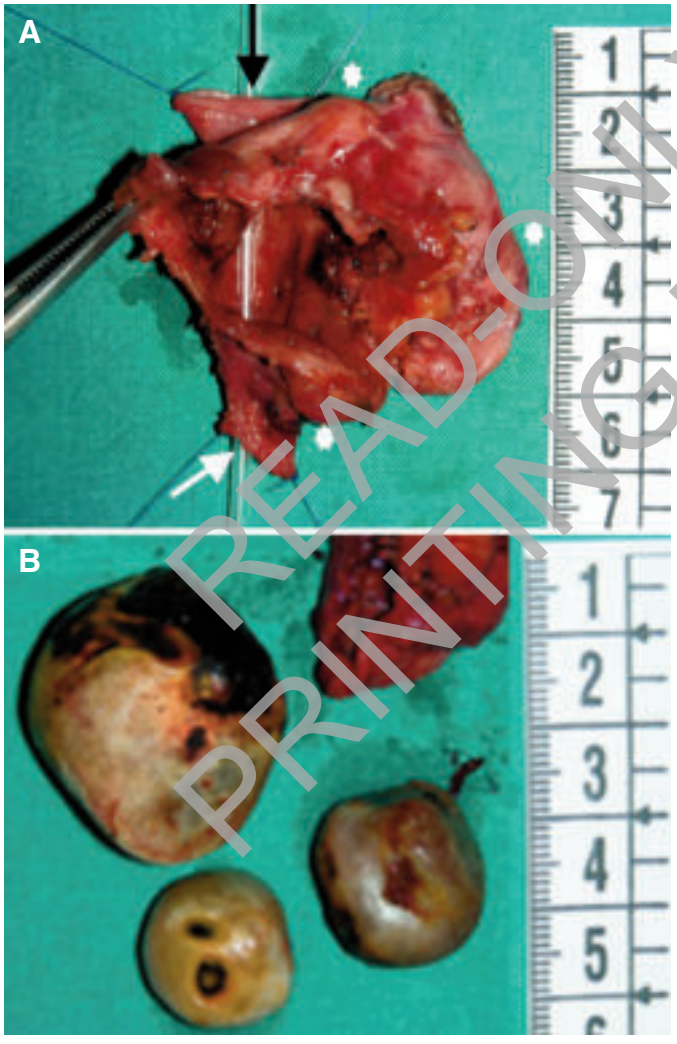

Fig. 4: (A) Surgical specimen: large type I choledochal cyst (white stars), contiguous to the common hepatic duct (black arrow) and the choledocus (white arrow). (B) Multiple stones contained into the choledochal cyst.

of the CBD (Fig. 1). Diagnosis of MS type I, according to McSherry classification [17], was made. A small sphincterotomy was performed and a plastic stent was left in place. Consequently a MRCP was done to confirm the diagnosis and to better define the type of MS and the anatomy of the biliary tract. The images revealed an extrinsic compression of the common hepatic duct, some gallstones in the infundibular-cystic duct and a normal CBD (Fig. 2). Diagnosis of MS type I was confirmed.
An elective surgery, in order to perform an open partial cholecystectomy leaving the neck of gallbladder in place was planned. At operation a scleroatrophic gallbladder and a cyst of the CBD approximately of 5 cm (Fig. 3) with some stones inside were found. Surgical diagnosis of a type I CC, according to the Todani classification system [18], was done and a cystectomy with reconstruction of the biliary tract using a Roux-en-Y hepatojejunostomy was performed. The postoperative course was uneventful, liver function tests returned to normal and the patient was discharged on post-operative day five. Pathologic examination revealed a small gallbladder measuring 3,5 cm with aspects consistent with chronic atrophic inflammation; this organ was contiguous with a cystic dilatation of the CBD of 5 cm fulfilled with stones and showing fibrotic hyperplasia with chronic inflammation (Fig 4a-b). Aganglionosis was observed on the distal resection margin. Postoperative 12-month follow-up showed no complication.

Discussion

MS and CC are pathological entities that should be ruled out in the differential diagnosis of obstructive jaundice. Correct diagnosis is necessary to avoid iatrogenic injury of the common hepatic duct during surgery due to long-standing inflammation and fibrosis. For both pathological conditions surgery is the gold standard treatment.
The most sensitive test for MS is ERCP, which allows correct preoperative diagnosis around 55% to 100% [19,20]. The features of ERCP in MS include a narrowing or curvilinear extrinsic compression involving the lateral portion of the common hepatic duct with proximal ductal dilatation and normal distal caliber.
In MS the diagnostic accuracy of MRCP is 50%[19]. MRCP can show some typical MS features such as the extrinsic narrowing of the common hepatic duct, a gallstone in the cystic duct, dilatation of the intrahepatic and common hepatic ducts, and a normal choledochus. Surgical treatment of MS depends on its type. Although laparoscopic cholecystectomy has almost completely replaced open cholecystectomy for the treatment of symptomatic gallstone disease, laparoscopic cholecystectomy is relatively hazardous in patients with MS, because safe dissection of Calot triangle is difficult due to severe local inflammation and adhesions [21]. Many procedures for MS type I are converted to open according to some

reports with a conversion rate of 74% with type I and of 100% with type II [22], therefore open cholecystectomy still remains the standard of care [23].
Traditionally considered a childhood disease, over the last two decades more CC cases are arising in adulthood [24-26]. The diagnosis can be sometimes incidental, such as a finding on CT scan, cholangiogram, or surgery [27].
ERCP has been reported to be the most sensitive imaging modality for CC, although in some cases a misdiagnosis is possible. Chronic inflammation and scarring may make difficult the ampulla cannulation or lead to lack of opacification of the biliary ducts. A high dye load may be necessary to visualize large cysts, obscuring, on the other hand, mucosal defects due to ulcers or malignancy [28]. Moreover, the sensitivity and the quality of the exam is operator-dependent [29].
MRCP is widely considered the gold standard for the diagnosis of CC [30], and the sensitivity has been reported as high as 90-100% [15]. Nevertheless, intraductal air, blood, debris, protein plugs or, like in the present case, stones can interfere with the correct visualization of the biliary system [31]. The most performed operation for CC is cyst removal and Roux-en-Y hepaticojejunostomy, but, depending on cyst type (Todani type IV, V), further intervention may be necessary [18,32,33].
The differential diagnosis between MS and CC is not easy. ERCP and CPRM do have a high sensibility, but their sensitivity is uncertain [34].
It is interesting that these two imaging methods could not make a correct diagnosis in the case we are presenting. This can be due to presence of multiple stones into the CC which, along with a scleroatrophic gallbladder, was mimicking impacted stones in the gallbladder infundibulum and misled to a diagnosis of MS.

Riassunto

Le cisti del coledoco sono dilatazioni cistiche delle vie biliari intra od extraepatiche con una incidenza di un caso su 150.000 nati vivi. La diagnosi di tale condizione patologica è più frequente in età infantile, ma può essere ritardata sino all'età adulta nel 20-50% dei casi. La manifestazione clinica più comune è il dolore addominale, con caratteristiche sintomatologiche ascrivibili a patologie biliari o pancreatiche. La sindrome di Mirizzi (SM) è una rara complicanza della inveterata litiasi della colecisti, ricorrendo in circa l'1% dei pazienti con colelitiasi e dovuta alla compressione ab estrinseco del dotto epatico comune da parte di calcolo/i contenuto/i all'interno della colecisti o del dotto cistico. I sintomi clinici includono ittero ostruttivo, colangiti ascendenti o, nelle fasi avanzate, fistola colecistocoledocica. Per entrambe tali condizioni patologiche la colangiopancreatografia retrograda endoscopica e la colangio-RM rappresentano gli strumenti diagnostici di scelta, con una sensitività ed una specificità fino al 100%.

Riportiamo il caso di un paziente di 66 anni ricoverato presso il dipartimento di emergenza ed accettazione del nostro ospedale per dolore addominale ed ittero, in cui sia l'esame endoscopico che quello radiologico posero diagnosi di SM, ma i cui reperti intraoperatori mostrarono una cisti del coledoco di tipo I.

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