Exceptional Mirizzi syndrome in a young child: A laparoscopic approach

Marcos Prada-Arias, José Luis Vázquez-Castelo, Patricia Blanco-Lobato, Javier Gómez-Veiras, Margarita Montero-Sánchez, Juan Manuel Vieito-Fuentes

Department of Pediatric Surgery, University Hospital of Vigo, Spain
Department of Radiology, University Hospital of Vigo, Spain

Article info

Article history:
Received 15 July 2014
Received in revised form 29 August 2014
Accepted 3 September 2014

Key words:
Mirizzi syndrome
Child
Laparoscopy

Abstract

Mirizzi syndrome is defined as an obstruction of the hepatic duct by an impacted biliary stone in the Hartmann’s pouch of the gallbladder or in the cystic duct (Mirizzi type I). The gallstone may erode the bile duct causing a cholecystobiliary fistula (Mirizzi type II). This very rare complication of long standing cholelithiasis is mainly reported in adults. We report an exceptional case of a type I Mirizzi syndrome in a 3-year-old boy, incidentally discovered during a computed tomography study. Ultrasonography and cholangioresonance confirmed the diagnosis. At laparoscopy, partial fusion between the Hartmann’s pouch and the hepatic duct was found. Despite difficult dissection, a total laparoscopic cholecystectomy was performed. Preoperative diagnosis of this syndrome is difficult owing to unspecific clinical presentation and low sensitivity of the standard radiological studies. To detect and correctly identify the type of Mirizzi syndrome during surgery is very important in order to avoid serious complications and to perform the most adequate surgical treatment. Open subtotal cholecystectomy is the recommended procedure in Mirizzi type I, laparoscopic total cholecystectomy being possible in some cases. Although this entity is exceptional in children, it must be known and considered by the Pediatric Surgeon because its development is possible.

1. Clinical case

We present a 3-year-old boy with past medical history of esophageal atresia and tetralogy of Fallot, for which he underwent cardiac surgery at the age of 4 months. The patient had an enhanced chest computed tomography (CT) for the study of the lungs; in the basal images a lamellated, calcified gallstone was incidentally detected associated to mild dilatation of the right intrahepatic biliary tree (Fig. 1). This cholelithiasis could be due to a combination of known lithogenic factors such as total parenteral nutrition (status post esophageal atresia repair and cardiac surgery), hemolysis by bypass (cardiac surgery) and medication (furosemide). By ultrasonography (US) and CT we confirmed the presence of a 1.5 cm gallstone in the gallbladder neck/cystic duct causing mild dilatation of the right intrahepatic biliary tree (ranging between 1.5 and 1.7 mm in diameter) (Figs. 2 and 3). The left intrahepatic biliary tree was not dilated (normal intrahepatic bile ducts are not routinely seen at US in children). The common bile duct (CBD) was found to be normal in size (1.9 mm in diameter). At the time, the patient was asymptomatic without clinical evidence of cholangitis, abdominal pain or jaundice. Serum
hepatic enzymes levels were elevated, aspartate aminotransferase 72 UI/L (4.0–50.0), alanine aminotransferase 84 UI/L (4.0–38.0), gamma-glutamyl transpeptidase 424 UI/L (4.0–50.0) and alkaline phosphatase 1156 UI/L (80.0–900.0) with normal serum bilirubin, 0.30 mg/dL (0.2–1.2). These values are indicative of dissociated cholestasis that is a typical finding of early cholestasis secondary to partial obstruction of the biliary tract. A diagnosis of type I MS was made and a laparoscopic approach was performed. We began the gallbladder dissection from the fundus toward the neck. Dissection of the cystic artery and cystic duct were easy and uneventful. As a result of the impacted gallstone, we found dense adherences and partial fusion between the gallbladder neck and the right hepatic duct, without a plane of dissection. Total cholecystectomy was completed with difficulty and a minimal perforation (1 mm in diameter) of the hepatic duct was made (Fig. 4) and subsequently closed with a monofilament suture. At the end of the procedure, we did not observe bile leak and decision was made not to perform an intraoperative cholangiography. The postoperative course was uneventful, without abdominal pain or jaundice and with normal levels of serum bilirubin. On the fifth postoperative day, US performed to evaluate biliary dilatation prior to discharge, showed a small biloma (2 x 3 cm) that spontaneously resolved at 3 months. Currently, a year after the procedure, our patient is doing well and is symptom-free with normal serum bilirubin and liver function tests. Minimal right intrahepatic biliary tree dilatation is still present; therefore periodic US follow-ups are performed.

2. Discussion

MS is a chronic complication of gallstone disease, encountered in 0.7–5.7% of patients with symptomatic cholelithiasis [6–9]. According to a recent serial study, mean age at presentation is 62 years (range 29–86 years) [10]. There is a female preponderance, with 70% frequency of all cases, probably a reflection of gallstone prevalence in this gender [1,7]. The pathogenesis of MS includes various components: a long cystic duct parallel to the HD, an impacted gallstone in Hartmann's pouch, gallbladder neck or in the cystic duct, and a partial extrinsic mechanical obstruction of the HD by the stone itself and/or from the resulting inflammation [6].

MS presents in both acute and chronic forms with similar frequency. Although the clinical presentation of MS is not specific, the most common clinical signs are obstructive jaundice (50–100%) and abdominal pain usually referred to the right upper quadrant (50–100%) [6,8,11,12]. The most common laboratory findings include hyperbilirubinemia and elevated transaminase enzymes [6,10]. Elevated levels of CA19-9 tumor marker have been described in patients with type II MS without associated malignity [1]. MS is currently considered a risk factor for gallbladder cancer, and it has been reported that 6–28% of patients with preoperative diagnosis of MS actually had gallbladder cancer [11].

Preoperative diagnosis of MS is difficult and is only made in 50% of the patients [13]. Confirmed preoperative diagnosis is important, because the incidence of bile duct injuries in MS operations without preoperative diagnosis could be as high as 17% [11]. If preoperative diagnosis is not made, intraoperative recognition of MS is essential for proper management.

The diagnosis of MS is based on the clinical findings, radiological images and endoscopic procedures and a high index of suspicion [1]. US and CT reportedly have a low sensitivity in detecting MS of approximately 40% [10,13]. US findings include: visualization of a small contracted gallbladder usually with thick walls; detection of a large calculus or multiple smaller stones entrapped in the Hartmann’s pouch, gallbladder neck or cystic duct; mild to moderate dilatation of the proximal biliary ducts and a normal size bile duct distal to the calculus [9,12]. The main utility of CT is the exclusion of malignancy in the porta hepatis area or in the liver [6,11]. CR is a useful tool to demonstrate MS. Typical features of MS shown by CR are the extrinsic narrowing of the HD, a gallstone in the cystic duct, dilatation of the intrahepatic and HD, a cholecystobiliary fistula and a normal CBD. CR has the advantage of avoiding the complications associated to endoscopic retrograde cholangiopancreatography (ERCP). However, the diagnostic accuracy for CR is approximately 50% [13]. ERCP remains the gold standard for the confirmation of MS with a diagnostic accuracy reaching 90% [1,10]. The features of ERCP in MS include the
visualization of a focal narrowing or curvilinear extrinsic compression of the lateral portion of the distal HD with proximal ductal dilatation and normal CBD [11]. Intraoperative cholangiography may be useful and can help confirm the diagnosis and determine the presence, location and size of a cholecystobiliary fistula. Intraoperative US has been reported to be a useful tool to identify the anatomy of the biliary tree and help with an accurate dissection of the bile duct in an inflamed area [11].

In 1982, McSherry et al. classified MS into two types based on ERCP findings: type I involves the external compression of the bile duct by an impacted stone in the cystic duct or in the Hartmann’s pouch; type II consist of a cholecystobiliary fistula caused by a stone that has eroded into the bile duct [14].

In 1989, Csendes et al. modified McSherry classification and divided MS into four types, according to the extent of bile duct destruction. Type I corresponds to McSherry’s type I. Type II consists of a cholecystobiliary fistula that involve less than one-third of the circumference of the bile duct. Type III relates to a cholecystobiliary fistula involving up to two-thirds of the bile duct circumference. Type IV is a cholecystobiliary fistula with complete destruction of the bile duct wall and a gallbladder completely fused to the bile duct, forming a single structure with no recognizable dissection plane between both structures [15]. In 2007, Csendes added a type V, which includes the presence of a cholecystoenteric fistula in combination with any other type of MS [1]. MS type I is fairly common (10.5–51%) and MS type IV is rather uncommon (1–4%). MS type V could be present in up to 29% patients with any other type of MS [8].

Open surgical approach remains the standard treatment for MS because of dense adhesions, anatomic distortion and the frequent need for bilioenteric drainage procedures [10,16]. Subtotal cholecystectomy, spearing the gallbladder neck or infundibulum, and closure of the stump with absorbable monofilament sutures over the bile duct, is the recommended surgical treatment for MS type I and most cases of MS types II and III. However, in some cases of MS type I a classic total cholecystectomy could be performed. In MS types II and III, simple repair of the cholecystobiliary fistula is not recommended for the risk of long-term stricture formation, which could cause cholestasis and cholangitis. In these cases a primary closure of the fistula over a T-tube drainage is feasible. If local inflammation precludes closure of the duct over a T-tube, continuity must be established by a bilioenteric anastomosis [11,13,15].

Laparoscopic cholecystectomy (LC) is considered controversial, technically challenging and dangerous, placing the patient at a probably unnecessary risk of bile duct injuries [16]. However, LC could be carefully undertaken in selected patients with MS type I [11,10,17]. A serial study of LC in MS reported a complication rate of 0–60%, bile duct injury of 0–22%, with a conversion rate as high as 31–100%, although most conversions were due to MS type II [1,10]. Preoperative diagnosis of MS decreases the rate of conversion and complications [16].

The degree of periductal inflammation, size of the impacted gallstone, and length of fusion between the infundibulum and cystic duct with the extrahepatic duct directly affect operative dissection. The major technical challenges for the laparoscopic surgeon in MS are: to grasp the infundibulum containing the impacted stone to allow dissection within the triangle of Calot, to separate the impacted stone from the HD and to distally identify the cystic duct for ligation [10]. Different authors have proposed several strategies to facilitate a laparoscopic approach and to decrease the risk of injury to the bile duct and conversion rate. These strategies include: subtotal LC, initial dissection from the fundus toward the infundibulum, to milk out the stone to the infundibulum in order to make the dissection of the gallbladder much easier, to open the gallbladder to remove the impacted stone in order to identify the infundibulum and cystic duct from within the gallbladder, and to perform intraoperative cholangiography to assess whether the bile duct compression has or not resolved after the dissection and to detect an unsuspected bile duct injury [10,18,19].

3. Conclusions

Preoperative diagnosis is difficult, being very important to establish it through clinical suspicion, imaging tests and endoscopic procedures. If MS is encountered during surgery every effort should

Fig. 3. Axial (left) and sagittal (right) T2 weighted MR images show the same findings: the large biliary stone in the gallbladder neck (arrowhead) and the dilated right biliary tree (arrow).

Fig. 4. Intraoperative picture showing iatrogenic HD perforation (arrow) made during dissection of dense adhesions between Hartmann’s pouch and HD.
be made to perform an accurate and cautious surgery trying to identify the type of MS to perform the most adequate surgical procedure. Although this entity is exceptional in children, it must be known and considered by Pediatric Surgeon because its development is possible.

**Conflict of interest**

We declare no conflict of interest.

**References**


