J Ped Surg Case Reports 5 (2016) 8–11

Contents lists available at ScienceDirect

Journal of Pediatric Surgery CASE REPORTS

journal homepage: www.jpscasereports.com

Intraoperative endoscopic treatment of Mirizzi syndrome in a pediatric patient

Nicole A. Wilson [a,b], Derek Wakeman [a,b], Elizabeth C. Utterson [a,b], Adam M. Vogel [a,b,*]

[a] St. Louis Children's Hospital, One Children's Place, Suite 5S40, St. Louis, MO 63110, USA
[b] Washington University in St. Louis, Department of Surgery, Wohl Hospital Room 9901, 660 South Euclid Avenue, St. Louis, MO 63110-1010, USA

ARTICLE INFO

Article history:
Received 7 December 2015
Received in revised form
14 December 2015
Accepted 18 December 2015

Key words:
Mirizzi syndrome
Cholecystectomy
Biliary
Choledochoscopy
Cholelithiasis
Choledocholithiasis

ABSTRACT

Mirizzi syndrome occurs when an impacted gallstone, together with an associated inflammatory response, causes external obstruction of the common hepatic duct or common bile duct. Patients classically present with obstructive jaundice, right upper quadrant pain, and sometimes with fever. Mirizzi syndrome is a rare presentation of complicated gallstone disease and is even more rare in the pediatric population. However, as the number of obese pediatric patients increases, so does the incidence of gallstone-related disease. We present a case of Mirizzi syndrome treated by open cholecystectomy and cystic duct stone extraction in a pediatric patient. Recognition and awareness of Mirizzi syndrome is important, even in the pediatric population, to aid in safe operative intervention and to avoid intraoperative bile duct injury.

Mirizzi syndrome is thought to arise as a rare chronic complication of symptomatic gallstone disease [1–3] in which an impacted gallstone at the gallbladder infundibulum or neck, together with an associated inflammatory response, causes external obstruction of the common hepatic duct or common bile duct. Patients with Mirizzi syndrome can develop obstructive jaundice and repeated attacks of cholecystitis. Eventually, the stone may erode into the common hepatic duct, resulting in a cholecystocholedochal or cholecystohepatic fistula [3].

Mirizzi syndrome occurs in 0.7%–2.8% of patients undergoing cholecystectomy [4–9]. This presentation is even less common in the pediatric population, where only a single case report was identified on literature review [10]. Furthermore, Mirizzi syndrome represents a diagnostic challenge because standard imaging may fail to demonstrate external compression of the bile duct and no findings are pathognomonic for the syndrome. For these reasons, Mirizzi syndrome is often omitted or overlooked from the differential diagnosis in pediatric patients with obstructive jaundice and cholangitis. However, awareness and recognition of this syndrome is essential for safe operative intervention.

1. Case report

A 13-year-old obese female (BMI 31.5) initially presented to a referring facility's emergency department with a 4-week history of nausea, vomiting, and intermittent epigastric pain that radiated to her back. An abdominal ultrasound demonstrated cholelithiasis with no evidence of cholecystitis. Initial laboratory analysis included: hemoglobin 13.1 g/dL, white blood cell (WBC) count 5800 cells/mcL (55.3% neutrophils), and serum total bilirubin 0.80 mg/dL. She was discharged home, but re-presented to the same facility two days later reporting worsening pain and emesis. Repeat laboratory analysis at that time revealed: hemoglobin 14.3 g/dL, WBC count 15,200 cells/mcL (81.4% neutrophils), serum total bilirubin 5.2 mg/dL, alkaline phosphatase 270 IU/L, and lipase 808 U/L. She was given piperacillin/tazobactam and transferred to our pediatric hospital for further management. Upon arrival her temperature was 38.1 °C. Repeat abdominal ultrasound demonstrated cholelithiasis without findings of acute cholecystitis and a moderately dilated bile duct (9 mm). Subsequent magnetic resonance cholangiopancreatography (MRCP) confirmed the presence of an obstructing stone in the distal cystic duct and secondary obstruction of the common hepatic duct (Fig. 1). Endoscopic retrograde cholangiopancreatography (ERCP) was performed with sphincterotomy and placement of a

* Corresponding author. Pediatric Surgery, St. Louis Children's Hospital, One Children's Place, Suite 5S40, St. Louis, MO 63110, USA. Tel.: +1 314 454 6022.
E-mail address: vogelam@wudosis.wustl.edu (A.M. Vogel).

http://dx.doi.org/10.1016/j.epsc.2015.12.006

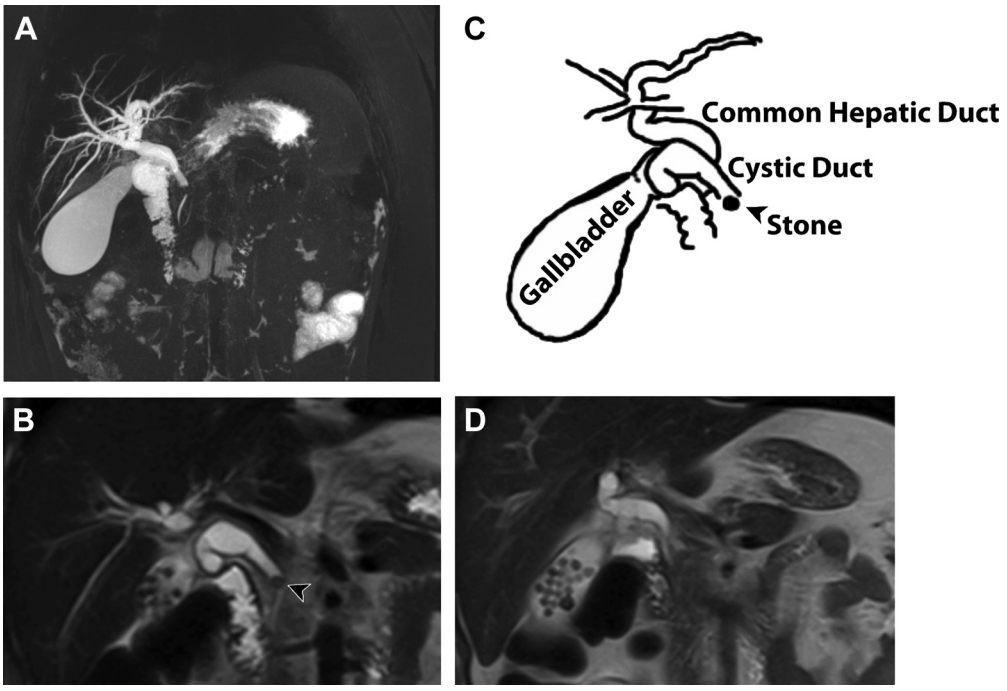

Fig. 1. (A) Three-dimensional rendering of MRCP images illustrating the elongated, tortuous, and dilated cystic duct. (B) MRCP image illustrating a 5–6 mm stone (arrowhead) at the confluence of the common hepatic duct and the cystic duct which is low just superior to the pancreatic head. (C) Line drawing illustrating gallbladder and biliary ductal anatomy. (D) MRCP image illustrating the common hepatic duct above the confluence with the cystic duct.

common bile duct stent. The stone in the distal cystic duct was found to be mobile on ERCP, but was unable to be retrieved at that time.

After three additional days of intravenous antibiotics (4-day course total), her symptoms resolved, her bilirubin normalized, and she was taken to the operating room for cholecystectomy. Moderate-severe inflammation was encountered around the cystic duct, particularly distal toward the common bile duct. The critical view of safety [11] was obtained laparoscopically (Fig. 2A) and intraoperative cholangiography showed a filling defect in the distal cystic duct (Fig. 3A). Despite the ERCP findings of a mobile stone, the stone was surprisingly immobile intra-operatively. Multiple attempts were made to clear the duct laparoscopically, including attempts to milk the stone retrograde into the gallbladder and stone retrieval with a Fogarty catheter. However, laparoscopic attempts to clear the duct were unsuccessful and a subcostal incision was performed. The obstructing stone was visualized using a 7-French flexible ureteroscope inserted into the cystic duct (Fig. 2B). The stone was successfully crushed and retrieved with an endoscopic nitrile basket. Subsequent cholangiography confirmed no residual stones in the biliary tree (Fig. 3B). Postoperatively, the patient did well and was discharged home on the third post-operative day without known complication. She was seen in follow-up and an ERCP was performed approximately 10 weeks postoperatively for stent removal (Fig. 3C).

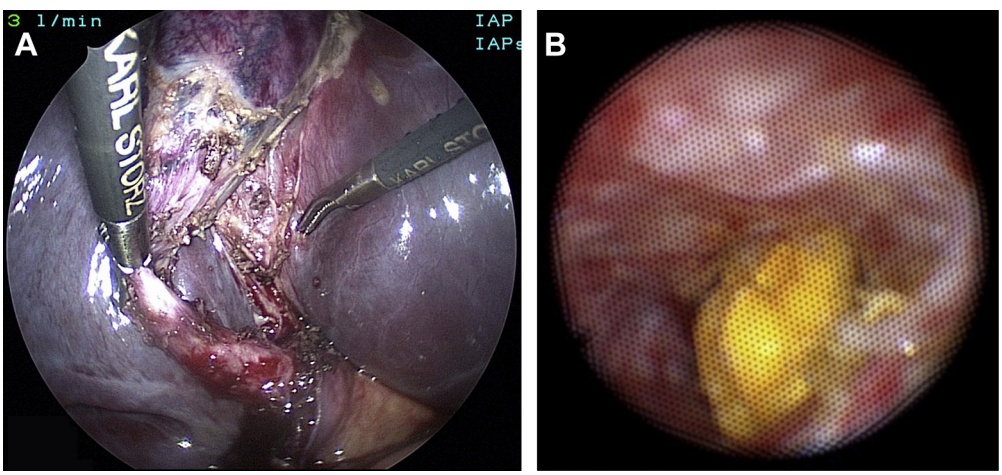

Fig. 2. (A) Intraoperative laparoscopic photo demonstrating the critical view of safety. (B) Intraoperative choledochoscopy illustrating an obstructing stone in the distal cystic duct, visualized using a 7-French flexible ureteroscope.