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Intraoperative endoscopic treatment of Mirizzi syndrome in a pediatric patient



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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.

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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

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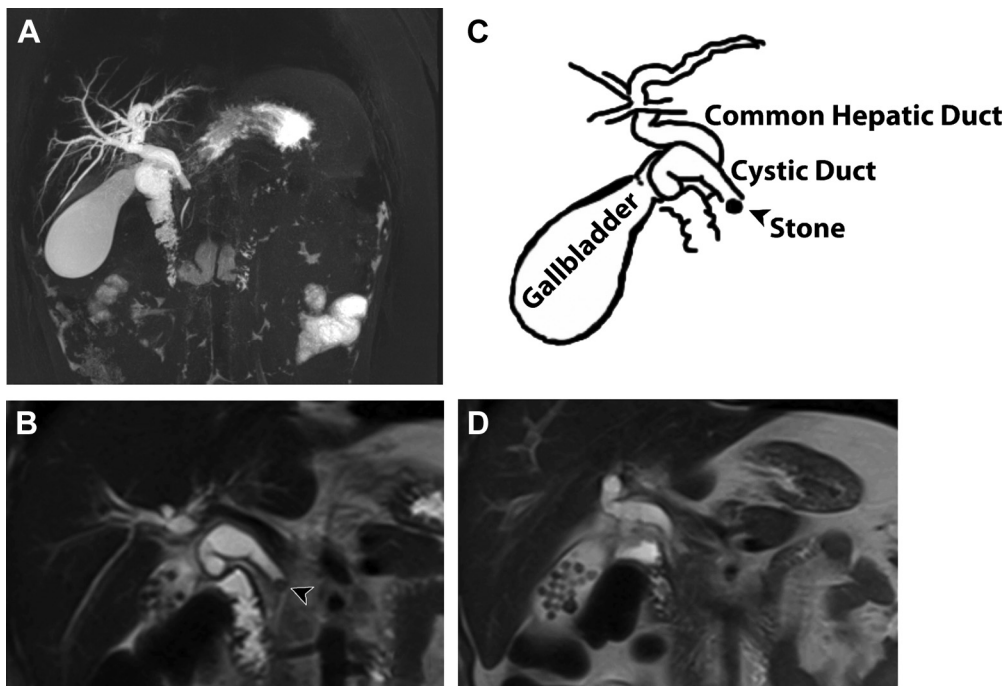


Fig. 1. (A) Three-dimensional rendering of MRCP images illustrating the elongated, tortuous, and dilated cystic duct. (B) MRCP image illustrating a 5–6 mm stone (arrowhead) at the confluence of the common hepatic duct and the cystic duct which is low just superior to the pancreatic head. (C) Line drawing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

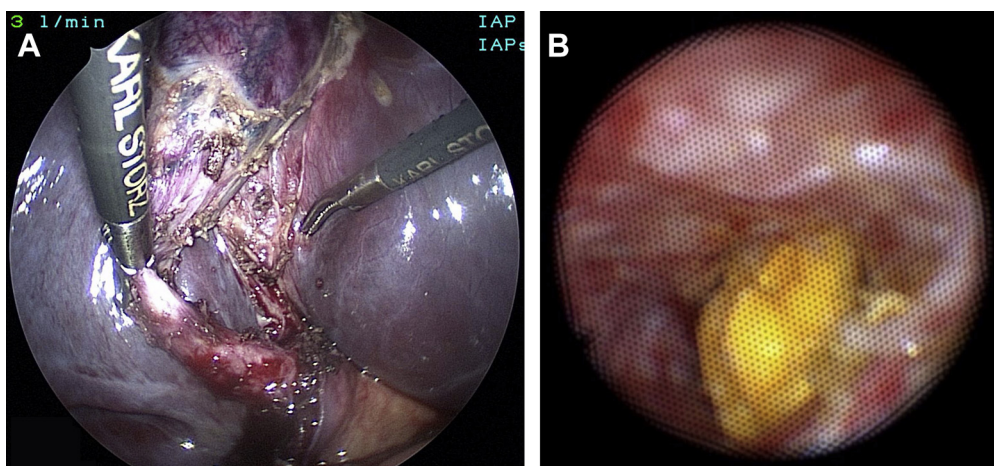


Fig. 2. (A) Intraoperative laparoscopic photo demonstrating the critical view of safety. (B) Intraoperative choledochoscopy illustrating an obstructing stone in the distal cystic duct, visualized using a 7-French flexible ureteroscope.

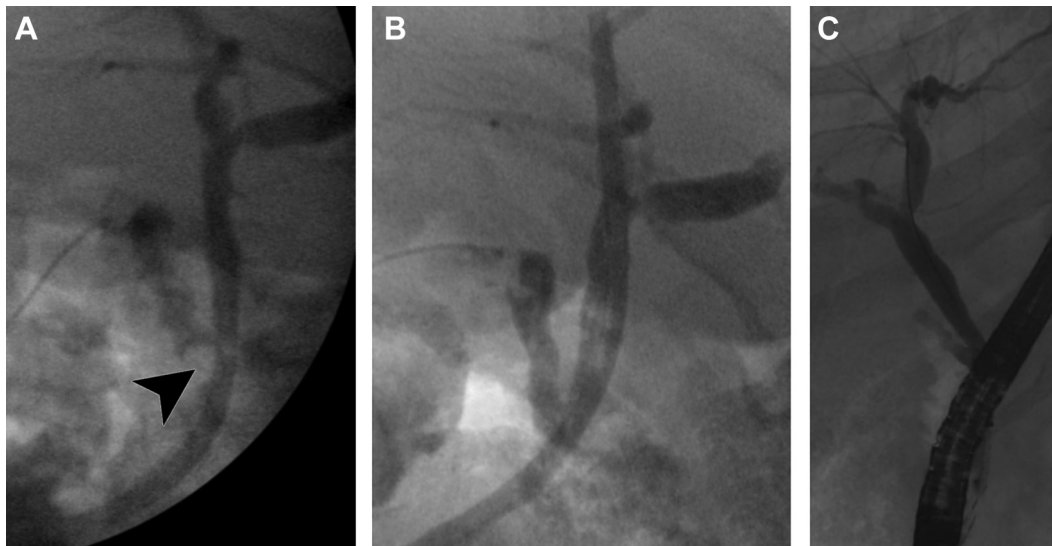


Fig. 3. (A) Intraoperative cholangiography demonstrating a filling defect (arrowhead) in the distal cystic duct. (B) Intraoperative cholangiography demonstrating resolution of the filling defect in the distal cystic duct. (C) Follow-up ERCP, performed approximately 10 weeks postoperatively, demonstrating no biliary ductal dilatation or intraluminal filling defects.

2. Discussion

In Mirizzi syndrome, a gallstone impacted in either the cystic duct or the neck of the gallbladder compresses the adjacent bile duct and results in complete or partial obstruction of the common hepatic duct or common bile duct [1,2]. The reported incidence of Mirizzi syndrome ranges from 0.7% to 2.8% of all cholecystectomies [4–7,9,12], but is rare in the pediatric population, with only a single case report identified in our literature review [10].

One population-based study estimated the prevalence of gallstones in children at 1.9% [12]. The etiology of the increasing incidence of cholelithiasis in children is likely multifactorial and related to both increased detection due to ultrasonography and the growing obesity epidemic [13]. For example, two recent studies have demonstrated an increasing incidence of pediatric gallbladder disease and have attributed some of the increase in incidence to obesity [13,14]. The incidence of Mirizzi syndrome in children may increase in parallel with the increasing incidence of cholelithiasis in the pediatric population.

Diagnosis of Mirizzi syndrome is challenging because standard imaging may fail to identify external compression of the bile duct [15,16]. Ultrasonography and computed tomography (CT) may demonstrate a stone impacted in the gallbladder neck, dilation of the biliary tree above the level of the gallbladder neck, and/or an abrupt change in the caliber (diameter) of the bile duct below the level of the stone [16]. However, as there is no constellation of pathognomonic findings either via imaging or laboratory analysis, preoperative recognition remains difficult in many cases.

Despite the challenges associated with definitive diagnosis, preoperative recognition of the syndrome is paramount due to the high risk of injury to the bile duct during surgical procedures [16]. Difficulty in identification of the gallbladder and bile ducts within the hepatoduodenal ligament in the presence of extensive adhesions may lead to injury or ligation of the common bile duct if it is mistaken for the cystic duct [16]. Furthermore, the severity, chronicity, and presence or absence of a cholecystocholedochal fistula may dictate the preferred operative approach [9].

3. Conclusion

This case of Mirizzi syndrome in a pediatric patient is presented in an effort to highlight that this rare complication of gallstone disease may occur in the pediatric population. Mirizzi syndrome should be included in the differential diagnosis for all patients with obstructive jaundice and/or cholangitis. 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.

Conflict of interest

None of the authors have conflicts of interest to disclose.

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