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Atypical Pediatric-Onset Mirizzi Syndrome in a Patient With Sickle Cell Disease: A Case Report

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Data Interpretation D
Manuscript Preparation E
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Patient: Female, 13-year-old
Final Diagnosis: Cholangitis • Mirizzi syndrome
Symptoms: Abdominal pain • dark color urine • fever
Clinical Procedure: Cholecystectomy • ERCP • MRCP
Specialty: Gastroenterology and Hepatology • Pediatrics and Neonatology • Surgery

Objective: Rare disease

Background: Mirizzi syndrome is a rare complication of gallstone disease, particularly rare in the pediatric population. It results from impaction of gallstones in the cystic duct or Hartmann pouch, causing external compression of the common hepatic duct. Most patients experience obstructive jaundice, right hypochondriac or epigastric pain, and fever. Sickle cell disease, a hemolytic condition common in Saudi Arabia, increases the risk of pigmented gallstone formation in affected children, leading to a higher likelihood of biliary complications at a younger age.

Case Report: The patient, a 13-year-old girl with known sickle cell disease, glucose-6-phosphate dehydrogenase deficiency, and cholelithiasis, presented with worsening right upper quadrant pain radiating to the back, dark urine, and pale stools, consistent with ascending cholangitis. Endoscopic retrograde cholangiopancreatography (ERCP) was performed to place a stent to decompress the biliary tree and achieve partial stone removal, stabilizing the acute condition and enabling control of sepsis and pancreatitis. After resolving the inflammation with antibiotics, supportive care, and hematological optimization, a subsequent laparoscopic cholecystectomy with bile duct exploration was performed, resulting in complete stone removal and an uncomplicated recovery.

Conclusions: We describe a complex pediatric sickle cell disease case presenting with Mirizzi syndrome, further complicated by obstructive jaundice, severe inflammation, and post-ERCP pancreatitis. This case underscores rare, early-onset Mirizzi syndrome in a child with sickle cell disease, highlighting the importance of high clinical suspicion when obstructive jaundice and fever appear in patients predisposed to biliary issues. Further reporting and research into its cause, especially its link to sickle cell disease, are crucial for better early detection and personalized treatment of Mirizzi syndrome.

Keywords: Case Reports • Gastroenterology • Mirizzi Syndrome • Saudi Arabia • Sickle Cell Disease

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Introduction

Stones lodged in the cystic duct or Hartmann pouch of the gallbladder that compress the adjacent bile duct represent a rare complication of cholelithiasis known as Mirizzi syndrome [1]. The impacted stone usually causes inflammation and, in severe cases, can progress to form a cholecystobiliary fistula or gallstone ileus [1]. Patients typically present with fever, right upper quadrant or epigastric colicky pain, and obstructive jaundice with stool discoloration and dark urine [1]. The prevalence of Mirizzi syndrome remains low worldwide, accounting for 0.5% to 5% of patients undergoing cholecystectomy [2]. The incidence tends to be lower in developed countries, usually less than 1%, while higher rates reaching 4.7% to 5.7% are reported in developing countries [2]. In children, gallstone disease is very uncommon, and most cases are attributed to chronic hemolysis [3]. Mirizzi syndrome in the pediatric age group is therefore an even greater rarity, with only a few cases documented [1]. In Saudi Arabia, the prevalence of Mirizzi syndrome is also limited, representing 0.7% of cholecystectomies performed [4]. Early recognition is essential for good prognosis; however, most of the definitive diagnosis of pediatric Mirizzi Syndrome occurs intraoperatively due to the difficulty of clinical diagnosis [5,6]

Case Report

A 13-year-old girl with a history of sickle cell disease, glucose-6-phosphate dehydrogenase deficiency, cholelithiasis, and Mirizzi syndrome presented to the emergency department with abdominal pain lasting 2 weeks and worsening 1 day before presentation. The pain was located in the right hypochondrium and radiated to the back. She reported fever and changes in the color of her urine and stool.

Upon physical examination, her height was 144 cm, weight was 30 kg, and body mass index was 14.46. The patient was lying in bed and exhibiting pain and distress, deep jaundice, chills, and rigors. Her vital signs showed a temperature of 39 °C, heart rate of 114 beats per minute, blood pressure of 100/80 mm Hg, and breathing on room air without supplemental oxygen. Her abdominal examination revealed right upper quadrant tenderness but no inspiratory arrest, indicating a negative Murphy sign.

Laboratory test results revealed an elevated white blood cell count of 35.1 k/mcL (reference range: 4.0-10.0 k/mcL) and C-reactive protein level of 20.18 mg/dL (reference range: 0.10-0.50 mg/dL), strongly suggesting inflammation. The patient had a low hemoglobin level of 9.4 g/dL (reference range: 11.5-15.5 g/dL); however, her baseline levels ranged from 8.0 to 9.0 g/dL. Elevated total and direct bilirubin levels are listed in **Table 1**. Other notable findings included low hematocrit (28.8%), increased red cell distribution width at 21% (reference range: 11.4%-13.5%), platelet clumping, an elevated reticulocyte count of 9.7%, and a mildly increased lactate dehydrogenase level of 468 U/L. Renal function and electrolyte levels were within the reference range. Urinalysis showed trace leukocytes.

The ultrasound revealed a distended gallbladder with multiple gallstones and sludge. A stone measuring 1.6 cm was lodged in the gallbladder neck, while the gallbladder wall was 0.16 cm thick.

A diagnosis of ascending cholangitis was made. Intravenous fluids, cefotaxime, and analgesics were initiated. Magnetic retrograde cholangiopancreatography (MRCP) revealed a stone measuring 0.7 × 0.7 cm causing extrinsic compression of the common hepatic duct with intrahepatic biliary radicle dilatation (Csendes type 1; **Figure 1A**). Endoscopic retrograde cholangiopancreatography (ERCP) was attempted but failed due to a technical issue and an inflamed major papilla, suggesting possible stone passage. A 5-Fr pancreatic stent, 3 cm in length, was temporarily inserted to prevent post-ERCP pancreatitis.

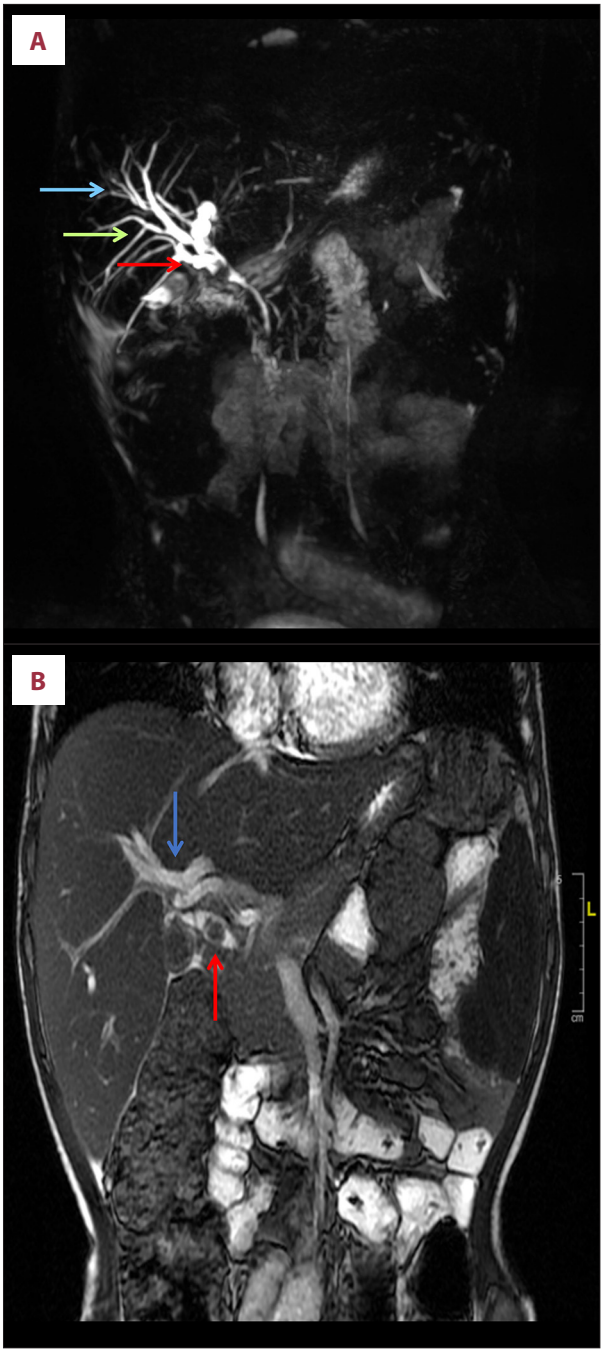
A second ERCP was performed after 48 hours using an adult duodenoscope, with smooth advancement to the second portion of the duodenum. The previously inserted stent was not visualized. A sphincterotome and 0.035-inch wire were used to cannulate the common bile duct (CBD). Cholangiogram showed a distal CBD diameter of 4 to 5 mm with a filling defect at the site of the cystic duct and significant bile duct dilation above the stone. An extraction balloon was used, and the stone was

Table 1. Laboratory test results over the course of management.

Laboratory test	Reference range	Before ERCP	After ERCP	Pre operatively
Total bilirubin	0.2-1.2 mg/dL	18.6	8.5	1.3
Direct bilirubin	0.10-0.50 mg/dL	12.57	5.42	0.5
Aspartate aminotransferase	11-34 U/L	504	99	18
Alanine aminotransferase	2-34 U/L	492	224	5
Alkaline phosphatase	64-359 U/L	593	469	248
Gamma-glutamyl transferase	9-36 U/L	297	206	11

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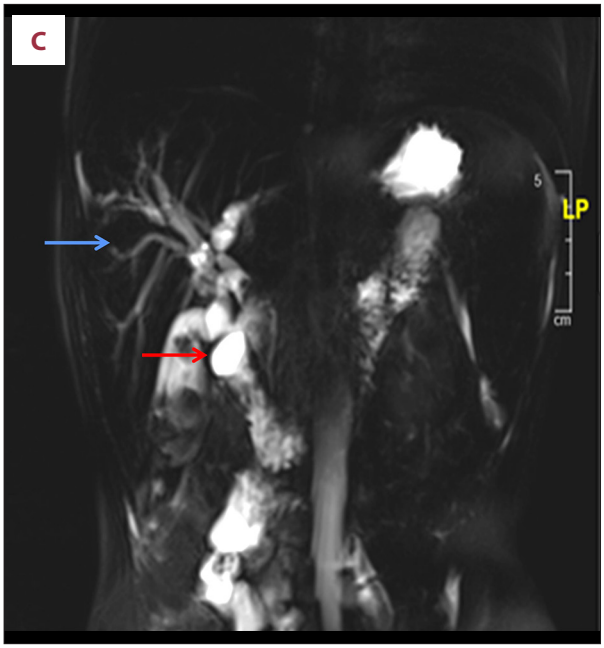


Figure 1. Magnetic resonance cholangiopancreatography (MRCP) findings. Initial MRCP demonstrated multiple variable-sized filling defects representing stones in the gallbladder, cystic duct, common hepatic duct (CHD), and common bile duct (CBD). A 0.7 × 0.7 cm stone (red arrow) caused extrinsic compression of the CHD with intrahepatic biliary ductal dilation (green arrow) (**A**). The green arrow also indicates the right anterior sectoral duct. Follow-up MRCP performed 6 weeks later, before surgery, demonstrated persistent cholelithiasis and biliary dilation with the CBD stent in situ (red and blue arrows, respectively) (**B, C**). Pneumobilia is present secondary to stent placement. Note the normal anatomic variant in which the right anterior hepatic duct drains into the left main hepatic duct. The CBD and pancreatic duct are intact.

mobile but could not be removed due to the very narrowed distal duct caused by the size of the stone. Basket lithotripsy was attempted, but the catheter could not pass the CBD. An 8.5-Fr, 7-cm straight plastic CBD stent was inserted and positioned above the stone (**Figure 2**). Laparoscopically intensive adhesions between the liver, omentum, and anterior and lateral abdominal walls were visualized (**Figure 3A**). The gallbladder was dissected from the gallbladder fossa in a retrograde fashion. The cystic artery was dissected and double-clipped with Hem-o-lok clips (**Figure 3B**).

Biliary decompression was achieved. The jaundice was relieved markedly and was evident clinically and on laboratory test results (**Table 1**). Subsequently, the patient developed post-ERCP pancreatitis and was treated conservatively. She needed a blood transfusion afterward due to peri-procedural hemolysis. Definitive cholecystectomy was postponed as an elective procedure until the inflammation was expected to resolve.

After 3 months, she was admitted electively for surgical treatment. She underwent follow-up MRCP, which showed the same findings (**Figure 1B, 1C**). An interval laparoscopic cholecystectomy with CBD exploration was performed.

Intraoperative findings revealed extensive adhesions between the gallbladder neck and the bile duct, confirming the progressive development of a type 2 cholecystocholedochal fistula (Csendes type 2). The gallbladder was inflamed with a

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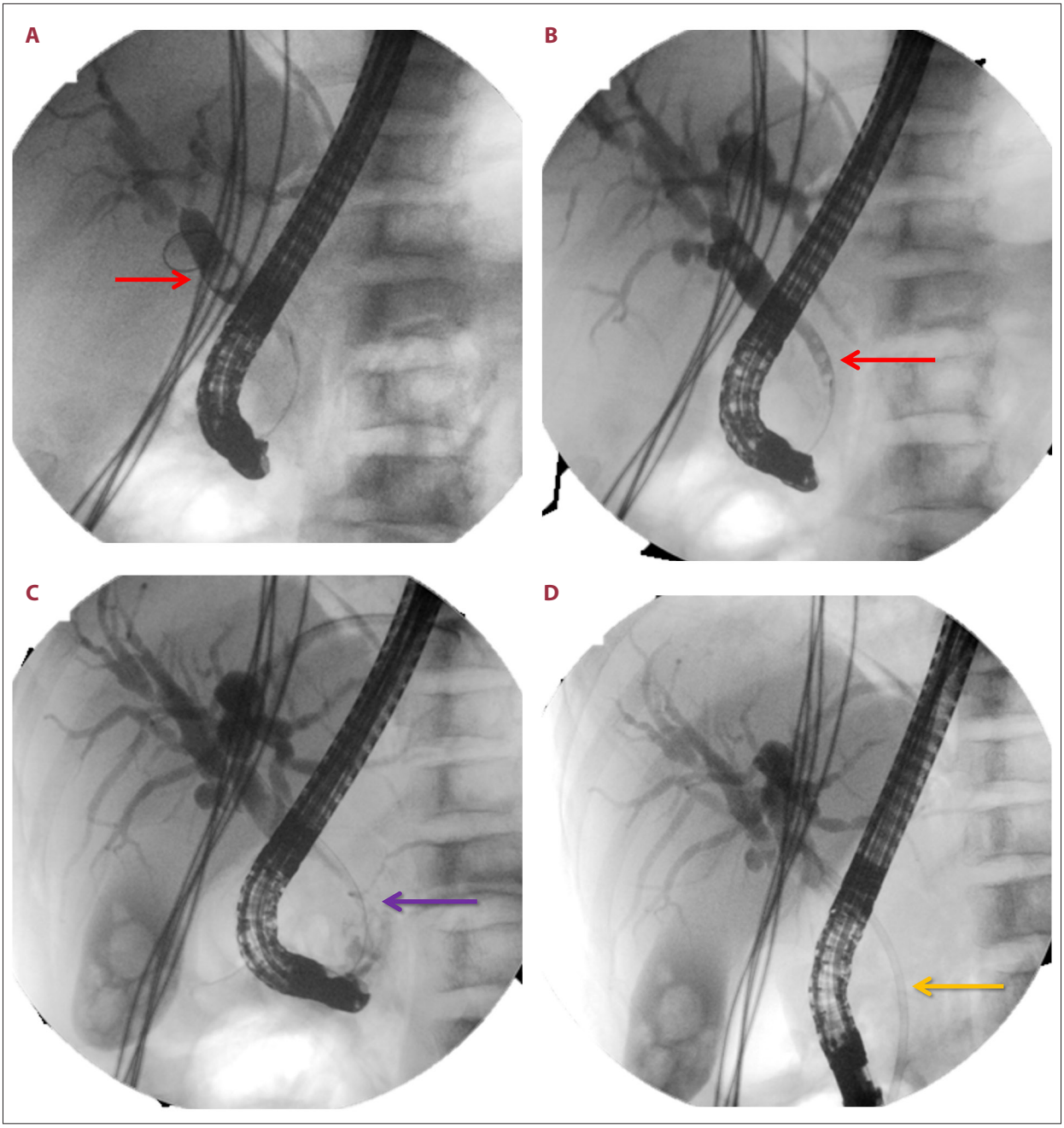


Figure 2. Endoscopic retrograde cholangiopancreatography (ERCP) findings and common bile duct (CBD) stent placement. A large filling defect representing a stone remains in the CBD (red arrow) (**A**). Multiple additional filling defects representing small stones are visible (red arrow) (**B**). Cholangiography demonstrated a distal CBD diameter of 4-5 mm with a filling defect at the cystic duct insertion and marked biliary dilatation proximal to the stone (purple arrow) (**C**). An extraction balloon and Dormia basket were used in an attempt to mobilize the stone, but the attempt was unsuccessful. An 8.5-Fr × 7-cm straight plastic CBD stent was inserted with its proximal end positioned above the stone (yellow arrow) (**D**). Excellent biliary drainage was achieved.

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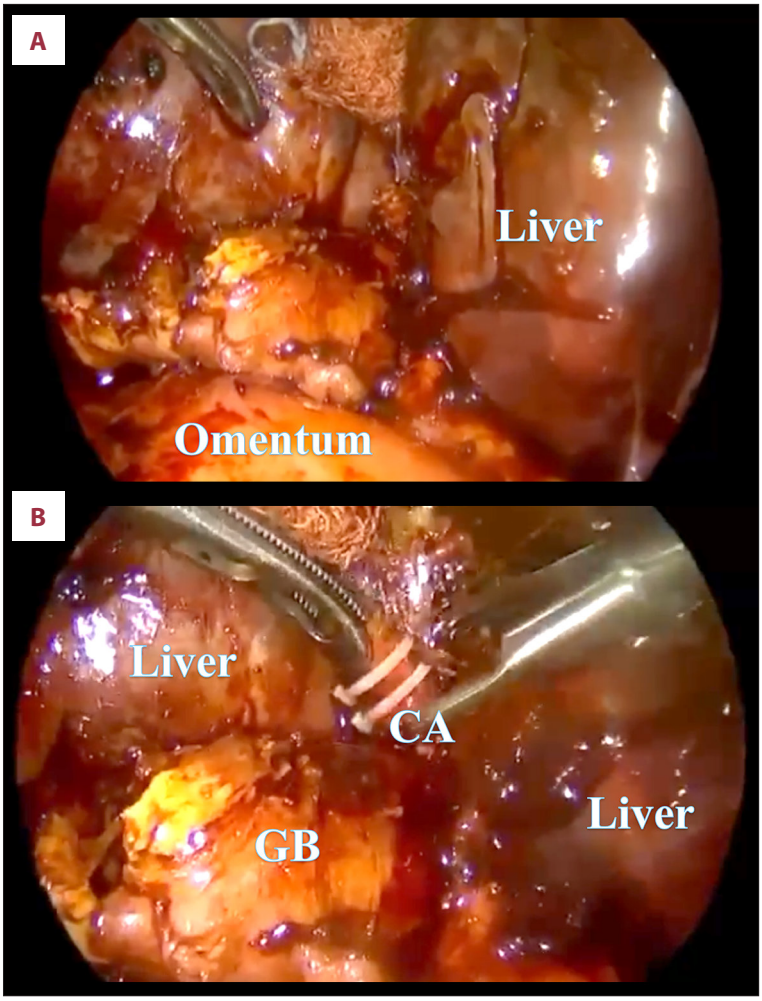


Figure 3. Intraoperative laparoscopic findings. Extensive adhesions were present between the liver, omentum, and the anterior and lateral abdominal walls. The gallbladder was not visualized (**A**). The gallbladder (GB) was dissected from the gallbladder fossa in a retrograde fashion (**B**). The cystic artery (CA) was dissected and double-clipped with Hem-o-lok clips (**B**).

perforated posterior wall, and the cystic and CBDs were dilated. The gallbladder was dissected retrograde from its bed. During dissection, frank pus and numerous small, pigmented stones were encountered from the posterior wall. Extensive irrigation and suctioning were performed.

Since sickle cell disease is usually associated with small stones, a decision was made to proceed with CBD exploration to prevent stone migration from the gallbladder to the bile duct through the cholecystocholedochal fistula. The exploration was performed by extending the CBD caudally to accommodate the choledochoscope. Intraoperative cholangiography revealed filling defects in the CBD. Multiple stones and sludge were removed from the CBD using suction irrigation and a Dormia basket. The stent did not interfere with the exploration and was left in place as an internal drain to manage a CBD leak after reconstruction. The CBD was primarily closed with a polydioxanone suture. Hemostasis was secured, and a drain was placed through the right anterior axillary port. She was stable and recovered steadily afterward.

The patient's postoperative recovery was smooth, and she was discharged on the seventh day of admission with scheduled follow-ups. Six weeks after surgery, she had an uneventful removal of the CBD stent in the endoscopy unit. Follow-up visits showed the patient to be pain-free with no further symptoms. The final pathology report showed acute on top of chronic calculous cholecystitis, with no evidence of dysplasia or malignancy.

Discussion

Pediatric Mirizzi syndrome is less common because gallstones take a considerable amount of time to grow large enough to block the cystic duct [1]. Since the stones are predominantly large, they are typically made of cholesterol. Cholesterol stones form when bile becomes supersaturated with cholesterol, leading to crystal formation [1]. It is also important to note that sickle cell disease is a significant risk factor for developing cholelithiasis [7]. However, the stones in sickle cell disease are usually calcium bilirubinate, known as pigment

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stones, resulting from hemolysis [8]. This contrasts with the cholesterol stones seen in Mirizzi syndrome. In our case, the patient had mixed stones, which contributed to the development of Mirizzi syndrome.

Several studies emphasize the importance of preoperative diagnosis, highlighting its role in reducing morbidity and improving prognosis [5]. However, to date, most Mirizzi syndrome diagnoses still occur intraoperatively. Mirizzi syndrome is classified by the Csendes system as type 1: compression of the common hepatic duct with no fistulization; type 2: fistulization into the common hepatic duct less than 33%; type 3: fistulization into the common hepatic duct from 33% to 66%; and type 4: fistulization into the CBD more than 66%. We were fortunate to have diagnosed Mirizzi syndrome early in our patient. ERCP has a higher accuracy rate of 92%, compared with MRCP and ultrasound [6]. In contrast, ERCP combined with computed tomography achieves an even higher accuracy of 94% [9].

The initial recommended therapy is ERCP; however, the complication rate of post-ERCP pancreatitis is high [10]. The best timing for performing laparoscopic cholecystectomy remains debated. Nevertheless, it is generally accepted that in cases of infection, it is wise to delay laparoscopic cholecystectomy for 6 weeks if ERCP fails [11]. We favored ERCP in our case due to the advanced technology available in our facility, compounded with the fact that initially our impression was ascending cholangitis. Laparoscopic and open cholecystectomy do not differ significantly in complication rates or avoidance, and the 2 procedures require a similar operative time. However, the most important prognostic factors are longer hospital stays and slower healing, which generally favor laparoscopic over open cholecystectomy [6].

In the literature, Marcos et al reported an unusual presentation of Mirizzi syndrome in a 3-year-old boy who was known to have esophageal atresia and tetralogy of Fallot, which was repaired at 4 months of age. An incidentally detected lamellated, calcified gallstone was found alongside mild dilation of the right intrahepatic biliary tree during non–contrast-enhanced computed tomography of the lungs. His clinical cholelithiasis was believed to be secondary to prolonged fasting, total parenteral nutrition, hemolysis due to bypass, and medications, including furosemide [12]. Further imaging with ultrasound and MRCP revealed a 1.5-cm gallstone in the gallbladder neck/cystic duct causing mild dilation of the right intrahepatic biliary tree. A laparoscopic cholecystectomy was performed.

Another case involved unusual obstructive jaundice in a 10-day-old male infant presenting with irritability and jaundice [13]. His laboratory test results showed direct hyperbilirubinemia and elevated gamma-glutamyl transpeptidase and alkaline phosphatase, with no other abnormalities. Ultrasonography revealed a cystic duct dilated by biliary sludge, with a normal gallbladder. Hepatobiliary scintigraphy showed no visualization of the intrahepatic and extrahepatic biliary tracts and no activity in the bowel for up to 1 hour after injection, indicating biliary atresia. Further imaging with MRCP demonstrated insertion of the cystic duct into the distal CBD near the ampulla of Vater, with a filling defect in the cystic duct causing ductal dilation and extrinsic compression of the CBD. The parents opted for medical management with ursodeoxycholic acid, which resulted in a good outcome [13].

Furthermore, Wilson et al reported a case of 13-year-old medically-free girl with similar presentation and Mirizzi syndrome confirmed by laboratory and radiological investigations. She had a difficult ERCP with no stenting and underwent laparoscopic converted to open cholecystectomy. CBD exploration was conducted to remove the lodged stone [14].

To our knowledge, this is the first pediatric case of Mirizzi syndrome associated with a hematological disorder reported in the English-language literature. Additionally, the type of gallstones this patient had, along with sickle cell disease and its sequelae of gallstone formation and associated complications, warrant further study.

Conclusions

Mirizzi syndrome in pediatric patients is very rare. Further investigation into its causes, especially its connection with sickle cell disease, is essential to enhance our understanding of this condition. In this report, due to maintaining high suspicion intraoperatively and close follow-up, the patient made a full recovery. However, we recommend that improving diagnostic strategies for preoperative detection is vital for better patient outcomes. It is also important to assess the long-term effects of different treatment approaches in pediatric patients. Finally, addressing the lack of reported cases of Mirizzi syndrome worldwide and in Saudi Arabia can contribute to a more complete understanding of this condition and support the creation of targeted prevention methods, ultimately enhancing the quality of life of affected individuals.

Department and Institution Where Work Was Done

Pediatric Surgery Department, King Fahad University Hospital, Dammam, Saudi Arabia.

Patient Consent Statement

In accordance with regional regulations and institutional guidelines, formal written consent was not required given the complete anonymization and de-identification of the patient data."

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