Stone removal in a five-year-old child with extrahepatic biliary obstruction using ERCP: A case report and a mini-review

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Abstract

We describe a 5-year-old child with extrahepatic biliary stone who successfully underwent endoscopic retrograde cholangiopancreatography for stone removal. He suffered from persistent colicky abdominal pain accompanied by fever that biliary stone confirmed him. ERCP along with other methods, can be considered a safe procedure for managing BD in children.

Introduction

Biliary obstruction is defined as the blockage of the extrahepatic biliary system (1). The etiology of the benign or malignant extrahepatic biliary system blockage involves various reasons, including gall stones in the cystic duct causing pressure on the bile duct (Mirizzi syndrome), choledochal cysts, and choledocholithiasis. Benign blockage of the extrahepatic biliary system can be due to stricture diseases such as fibrotic strictures from gall stone passage, PSC, and iatrogenic strictures from bile duct cannulation. Neoplastic cases are presented with stricture diseases causing biliary obstruction, including pancreatic head cancer (causing distal CBD stricture), ampullary carcinoma or adenoma, and cholangiocarcinoma (2). The most common cause of biliary obstruction in developed countries is choledocholithiasis due to cholesterol stones. Pigmented stones due to hemolysis and infectious diseases, recurrent pyogenic cholangiohepatitis with increased risk for cholangiocarcinoma, and calculi in intrahepatic bile ducts are common etiologies in the Asian population which are rare in western countries. Recurrent pyogenic cholangiohepatitis is characterized by recurrent bacterial cholangitis, stricture, and dilatation of the biliary system. Gallbladder malignancy is more common in East Asia, Central and South America, Central and Eastern Europe, and the north of India (2, 3).

In general, choledocholithiasis is uncommon in children. Since the extrahepatic biliary obstruction in children is very rare, most reports describe the condition's etiology in adult patients. The prevalence of cholelithiasis in pediatrics has been reported to be 0.13% - 0.3%. However, the incidence is higher in obese children and adolescents and is estimated at 2% - 6.1% (4).

More than 80% to 90% of all patients with CBD stones can be treated by non-surgical methods through sphincterotomy and stone extraction in combination with Dormia baskets or balloon catheters. In case of non-extractable stones >1 cm, additional procedures such as mechanical lithotripsy, including balloon dilatation, extracorporeal shock-wave lithotripsy, electrohydraulic probe lithotripsy, laser lithotripsy, stenting for immediate and definitive stone treatment are applied (4). Endoscopic retrograde cholangiopancreatography is a diagnostic and therapeutic technique routinely used for adults (5). Relative to the published studies regarding adult ERCP, the articles on pediatric ERCP remain limited for several reasons (6). Firstly, it is technically more challenging to be used for children. Secondly, pancreaticobiliary pathology in the pediatric population is rare, so the study cannot have an adequate sample size. Additionally, in children weighing

more than 10 kg, pediatric ERCP duodenoscopes and accessories have limited application (8). Moreover, the advancement of MRCP has limited the use of ERCP for diagnosis.

This case report presents a CBD stone case with a left-sided gall bladder treated with ERCP in an otherwise healthy 5-year-old male.

Left-sided gallbladder refers to a gallbladder located on the left side of the ligamentum teres. It is a rare anomaly usually related to the absence of segment IV, portal vein anomalies, or biliary system anomalies. Diagnosis of the associated anomalies is essential for managing liver transplantation, liver resection, and complicated hepatolithiasis. Preoperative diagnosis of the left-sided gallbladder with associated anomalies is required to reduce the risks of operative complications (7).

Case report

A 5-year-old male was admitted to Namazi hospital (Shiraz, Iran), suffering from a persistent colicky abdominal pain for the last 30 days. Except for his premature birth, he had no history or family history of the following conditions; previous medical problems, taking any medications, past surgeries, abdominal trauma, weight loss, obesity, metabolic syndrome, hemolytic diseases, severe skin itching (Bayler disease), recurrent icterus, anemia, splenectomy, symptoms of chronic liver disease or liver dysfunction, liver disorders (e.g., Wilson's disease), steatorrhea, chronic diarrhea, as well as any underlying causes of gallstone formation. The pain was localized in the epigastric and periumbilical areas exacerbating after consuming dairy and high-fat foods. The pain was accompanied by fever, vomiting, and constipation. The patient was born with GA = 28W and a weight of 900 g. His current body weight and height were 16 kg (10–25th percentile) and 112 cm (50–75th percentile), respectively. Vital signs were stable; blood pressure 100/70 mm Hg, heart rate 104 beats/min, respiratory rate 28 breaths/min, and body temperature of 36.5°C. He had hepatomegaly and abdominal tenderness in the epigastric and periumbilical areas in the physical examination.

The hemoglobin, white blood cell, and platelet counts were 12.9 g/dL, 3700/mm3, and 192×106/mm3, respectively. Hemoglobin electrophoresis was normal. Hemolysis was not noted. Blood chemistries were as following: cholesterol 98 mg/dL (reference range, 120–200 mg/dL), total protein 6 mg/dL (6.1–7.9 mg/dL), albumin 4 mg/dL (3.5–5.6 mg/dL), alkaline phosphatase 962 U/L , AST 104 U/L (15–40 U/L), ALT 180 U/L (5–45 U/L), GGT 160U/L (5–32 U/L), total bilirubin 1.7 mg/dL (<2.0 mg/dL), amylase 44 U/L (16–91 U/L), and lipase 90 mg/dL (4–29 mg/dL) (Table 1).

Abdominal ultrasound imaging revealed a distended gallbladder by diffuse wall thickening with a maximum thickness of 3.5 mm, a CBD 10.8 mm in diameter, and an 8.5-mm-sized stone in the distal CBD, suggestive of acute cholecystitis.

Non per oral diet, intravenous hydration, and administration of analgesics and antibiotics (cefotaxime and metronidazole) were started after the patients' admission. The medical care team decided to perform MRCP and ERCP to evaluate and manage the CBD stone. In MRCP evaluation, mild dilation of central intrahepatic bile ducts and CBD (6 mm) was apparent, associated with a dark signal of a 5 mm stone within the pancreatic portion of CBD located at a 15 mm distance to the major papilla. The gall bladder's position was on the left side of the subhepatic area, concurrent with portal vein abnormality. The main portal vein was trifurcated, and ascending portion (umbilical portion) of the left portal vein was hypoplastic. Additionally, small teres ligament differentiation of segment IV was suboptimal (Figure 1).

Since our center is well-equipped, ERCP was performed under general anesthesia. During the procedure, dilatation of CBD with a stone in the middle was investigated (Figure 2). After sphincterotomy, a 10*15 mm pigmented stone was removed using a stone retrieval balloon (Figure 3). After the stone removal, all signs and symptoms were alleviated. There were neither any complications during ERCP nor after discharge. The patient was discharged, and Ursodeoxycholic acid (10 mg/kg/dose) and Polyethyenglycol syrup (PEG) (1 cc/kg/day) were prescribed for 20 days. No complications, signs, or symptoms were observed in the follow-up visit, and all lab results and abdominal sonography were within the normal range. Finally, the patient was referred for elective cholecystectomy.

Table 1: Laboratory data during hospitalization

4 months later	12 Days After Admission	3 Day After Admission	On Admission	Lab Data
6490		7600	3700	**WBC, /mm^3**
13/9		12/1	12/9	**Hb, gm/dL**
299		467000	192000	**Platelet, /mm^3**
			14	**PT, sec**
			1/1	**INR, index**
			34	**PTT, sec**
			12	**B.U.N, mg/dL**
			0/37	**Cr, mg/dL**
			135	**Na, mEq/L**
			3/7	**K, mEq/L**
	25	64	104	**AST, IU/L**
12	56	131	180	**ALT, IU/L**
406	665	1037	962	**ALK.P, IU/L**
9/4	114	225	160	**GGT**
		1/2	1/7	**Total bilirubin, mg/dL**
		0/5	0/5	**Direct bilirubin, mg/dL**
		6/4	6	**Total protein, g/dL**
		4/2	4	**Albumin, g/dL**
			Neg	**Coombs**
			44	**Amylase**
			90	**lipase**
			12	**ESR, mm/h**
3/6			2	**CRP, mg/L**
			98	**cholesterol**
			122	triglyceride
			Neg	Viral marker (HBS Ag, HAV Ig M, HCV Ab)
		437	135	**Urine COPPER**
			Neg	**IgG**
			Neg	**Anti-LKM**
			Neg	**ANA**
363				**LDH**
3/1				**Uric acid**
			No growth	**Blood culture**
			No growth	**Urine culture**

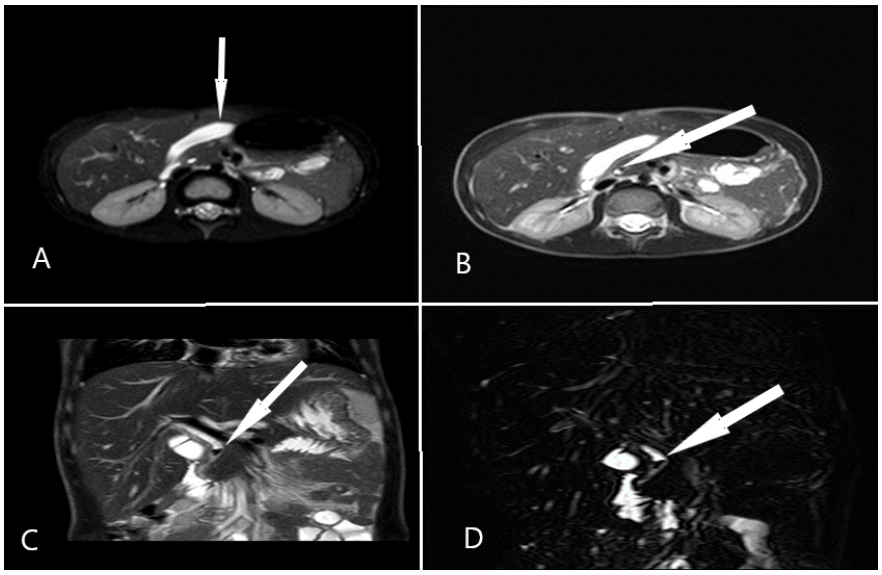

Figure 1: A) axial T2 MRCP sequence: gall bladder in left subhepatic area. B) axial T2 MRCP sequence: tiny signal void (dark) stone in the pancreatic portion of CBD. C) coronal T2 MRCP sequence: signal void tiny stone in the distal part of the CBD. D) coronal 3D reconstruction MRCP image presenting tiny stone in CBD.

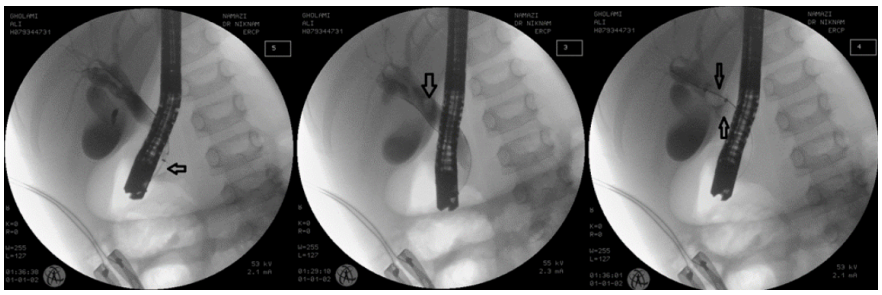

Figure 2: ERCP: dilatation of the CBD with a filling defect (left image) in the middle part and stone removal using a stone retrieval balloon (middle and right images).

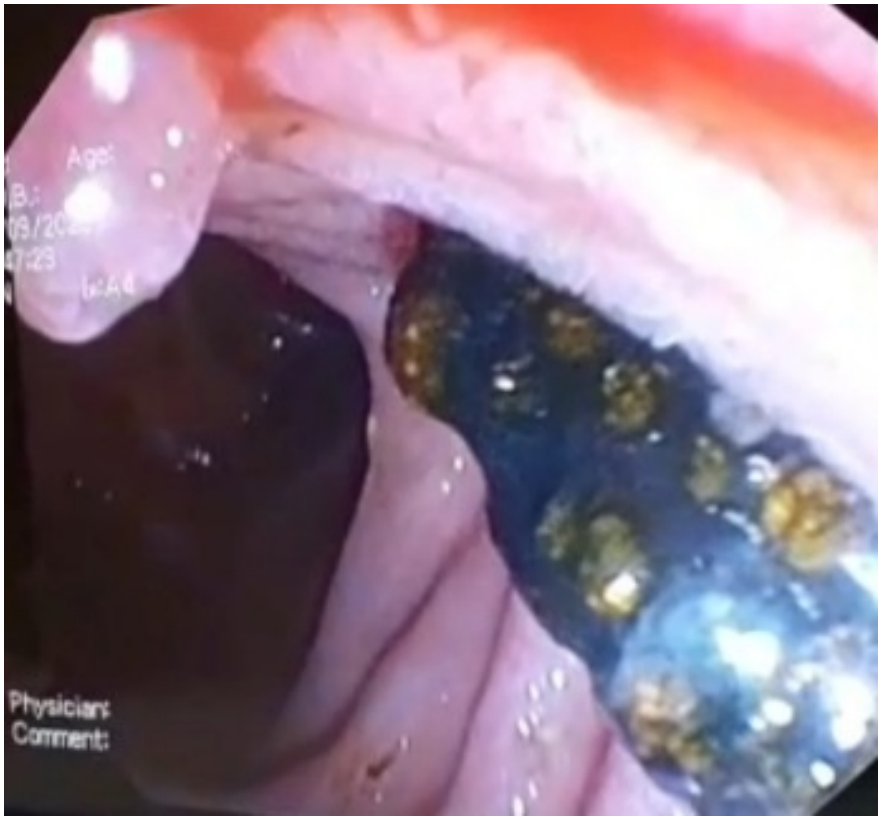

Figure 3: Endoscopic view of pigmented stone after removal using a stone retrieval balloon

Discussion and conclusion

CBD stone is a rare condition in children, although it has become more common recently (8). Unlike adults, it is mostly symptomatic in children depending on several factors such as age, ethnicity, geographical localization, medical facilities, and referral status (9). The most common cause of cholelithiasis in children is a hemolytic disease (20–30%). Other pathologic situations such as congenital hepatobiliary diseases, obesity, receiving total parenteral nutrition, ileal disease or resection, use of ceftriaxone, metabolic syndrome, choledochal cyst, PFIC, NEC, biliary cirrhosis, prematurity, Wilson disease, cystic fibrosis, congenital heart diseases, and idiopathic cholelithiasis should also be considered as the causes of cholelithiasis in pediatrics. (10). Houman et al. reported a 12-year-old boy with a hemolytic uremic syndrome, established by renal biopsy, who developed cholestatic jaundice. It was discovered by ERCP and extracted by sphincterotomy (11). It has been proven that for treating pancreaticobiliary disease, endoscopic sphincterotomy or CBD explorations is a safe method, even in pediatric population (12, 13). The case presented in this report was symptomatic. The patient had no history or family history of any medical conditions except prematurity. One of the important findings in the MRI and MRCP was the left-sided gallbladder. Despite stone removal, he is still at risk for CBD stone formation because of having an anomaly in the biliary system. So, the patient was advised to seek cholecystectomy in a soon future.

The left-sided gall bladder refers to a gallbladder lying on the left side of the falciform ligament (14). The reported incidence of this anomaly is estimated to be between 0.1% and 1.2% (13). It is very rare and includes three anatomic abnormalities: a right-sided ligamentum teres, an ectopic left-sided gallbladder, and a situs inversus (9). The possible associated abnormalities with the left-sided gallbladder are portal vein anomalies, biliary system anomalies, and left lobe hypoplasia (15). It has been reported that the likelihood of intraoperative bile duct injuries in individuals with left-sided gall bladder is higher than the average

population (up to 7.3%) due to anomalies of the bile duct, portal vein, and other anatomical structures in the hepatobiliary system (16). The patient discussed in our report has trifurcated main portal vein with a hypoplastic ascending portion (umbilical portion) of the left portal vein. Moreover, small teres ligament differentiation of the fourth segment was suboptimal.

As mentioned before, a left-sided gall bladder is a rare condition. Nevertheless, it is possible to accurately diagnose a left-sided gall bladder before surgery and perform laparoscopic cholecystectomy by adjusting the port position. Increased size of the left portal vein and distribution of the left portal vein crossing over to the right side of the liver is the crucial common features of the left-sided gallbladder. These variations probably have considerable clinical implications in managing hepatic resection, including donor hepatectomy (17).

In recent years, the treatment approaches for managing choledocholithiasis in children have become more specific; however, no gold standard procedure is available yet. The endoscopic approach for managing biliary tract obstruction in medical centers performing ERCP is usually the first choice. Similarly, laparoscopic evaluation of CBD has also proven safe and effective. In the absence of ERCP, laparoscopic investigation can be an appropriate alternative. Laparoscopic cholecystectomy for biliary stone disease in the pediatric population has been well proven as the standard of care, similar to adult patients (18). ERCP was commonly applied from 1970 to 1979 to diagnose and treat hepatobiliary diseases in children. However, its use has been more restricted in recent years due to being an invasive procedure (19). For instance, Felux et al. (19) and Lou et al. (20) have reported that ERCP in children accounts for almost 3.3 and 4% of all ERCP procedures in their centers, respectively. In Asian countries, only a few studies with small sample sizes regarding pediatric ERCP have been performed (21). The success rate of endoscopic procedures, especially in children, requires a complete evaluation of the condition before ERCP, and it highly depends on the specialist's skill. It is essential to understand that the ERCP indications should not be extended blindly because they may cause unnecessary complications (22).

To the best of our knowledge, no report on bile duct stone removal by ERCP in children has been published previously in Iran. This case is the first report of a successful pediatric ERCP for treating a bile duct obstruction due to a stone. The CBD stone was endoscopically removed. ERCP, along with other methods, can be considered a safe procedure for pediatric biliary diseases in well-equipped centers. The patient is still at risk for CBD stone formation because of having an anomaly in the biliary system. Laparoscopic cholecystectomy is necessary in this patient.

Availability of data and materials

The data supporting this case report's findings is available from the corresponding author upon request.

Abbreviations

PSC: primary sclerosing cholangitis, CBD: common bile duct, ERCP: Endoscopic retrograde cholangiopancreatography, GA: gestational age, AST: aspartate aminotransferase, ALT: alanine aminotransferase, GGT: gamma-glutamyl transpeptidase, MRCP: magnetic resonance cholangiopancreatography, MRI: magnetic resonance imaging, PEG: polyethylene glycol, PFIC: (progressive familial intrahepatic cholestasis), NEC: necrotizing enterocolitis

Declarations

Ethics approval and consent to participate

This research was approved by the ethics committee of shiraz university of medical sciences with IR.SUMS.REC.1401.665 code.

Consent for publication

Written informed consent was obtained from the patient's mother to publish this case report and any accompanying images. All authors have viewed and agreed to the submission.

Competing interests

All authors declare that this manuscript has no conflict of interest.

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Authors' contributions

RN and SMMM performed the endoscopy and cared for the patient. MS, MD and RM collected the clinical data and wrote the manuscript. MGHJ helped with the imaging diagnosis. MA revised the manuscript. All authors have read and approved the final manuscript.

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