Simultaneous gallbladder-preserving cholecystolithotomy and laparoscopic splenectomy as a surgical option for hereditary spherocytosis in a child: A case report

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ABSTRACT

For hereditary spherocytosis complicated by cholelithiasis, cholecystectomy is simultaneously performed with splenectomy. However, jaundice promptly disappears after removing the spleen, and the risk for recurrent cholelithiasis decreases in majority of cases; gallbladder-preserving cholecystolithotomy can be an option for such patients. We report a case of a 6-year-old boy with hereditary spherocytosis who was referred by a previous physician for the chief complaint of anemia (hemoglobin level, 7–8 g/dl). Abdominal ultrasound and CT revealed numerous small calculi in the spleen and gallbladder. The patient developed calculous cholecystitis twice. Hence, laparoscopic splenectomy and gallbladder-preserving cholecystolithotomy were simultaneously performed. The operating time was 2 h and 58 min, and the blood loss was 45 ml. No postoperative complication developed, and the patient was discharged 5 days after the operation. To date, i.e., a year after the operation, no recurrent cholelithiasis has been observed in the patient.

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Hereditary spherocytosis (HS) is a congenital disease wherein red blood cells become round shaped because of abnormal membrane permeability to sodium, which is induced by a congenital defect of spectrin, a major component of red blood cells. The round-shaped red blood cells are fragile and prone to hemolysis and are destroyed in the spleen because of their loss of deformability, thus inducing chronic anemia. Subsequently, patients develop jaundice and splenomegaly, and cholelithiasis occur in approximately 50% of patients [1]. In HS cases complicated by cholelithiasis, cholecystectomy is often simultaneously performed with splenectomy. However, we report a case wherein cholecystolithotomy and splenectomy were simultaneously performed to preserve the gallbladder in a child.

1. Case report

The patient was a 6-year-old boy with a history of neonatal jaundice that was treated with phototherapy. In terms of family history, few relatives from the maternal family developed cholelithiasis in the past. The patient developed hemolytic crisis induced by parvovirus infection and received a diagnosis of HS from his previous physician. During the course of observation, the fatigue and anemia persisted. In addition, gallbladder stones were observed using ultrasound (US) in the previous hospital. Therefore, the patient was referred to our hospital for splenectomy. While waiting for an operation, calculous cholecystitis occurred twice and antibiotics were administered for a month. Blood tests revealed the following results: white blood cell count, 6300/μl; Hb level, 8.3 g/dl; T-bil level, 8.3 g/dl; T-bil level, 2.8 mg/dl; D-bil level, 0.7 mg/dl, suggesting anemia and jaundice associated with a higher unconjugated bilirubin level. No other abnormalities were detected in the biochemical examination or coagulation test. Abdominal US revealed a contracted gallbladder that contained multiple small stones of 2–3 mm (Fig. 1).

Abdominal CT revealed an enlarged spleen (91 × 51 mm) and accumulated small stones inside the gallbladder. Dilatation of the common bile duct or pancreatic duct was not observed (Fig. 2).

MRCP revealed small stones in the gallbladder but not in the common bile duct or intrahepatic bile duct. No other anatomical abnormality was observed in the biliary system. Hence, a diagnosis
of HS complicated by gallstones was made, and laparoscopic splenectomy and gallbladder-preserving cholecystolithotomy were simultaneously performed after obtaining consent from the patient’s family.

During the operation, a wound retractor was first set through a vertical incision in the umbilical region. A 12-mm port was inserted on the right hypochondrium and was guided toward outside the gallbladder fundus through the incision site of the port (Fig. 3A). While holding the bottom of the gallbladder with two forceps, a small hole was made in the gallbladder and bile was aspirated. Then, we made a small window of approximately 1 cm and fixed the gallbladder wall and skin at four points (upper/lower and left/right) using a 3-0 absorbable suture to prevent any leakage of bile into the peritoneal cavity. From the same window, six black gallstones (2–3 mm) were removed using a curette (Fig. 3B). A choledochofiberscope (OLYMPUS CHF-P20Q Choledochoscope) was inserted through the window, confirming that there were no residual stones inside the gallbladder (Fig. 3C).

Subsequently, a tube (6 Fr.) was inserted in the gallbladder, and direct cholangiography was performed to confirm the absence of stone shadow in the common bile duct. The incision of the gallbladder was closed with 3-0 absorbable sutures, and the organ was returned to the peritoneal cavity. The 12-mm port was reinserted into the same site, and laparoscopic splenectomy was performed as a routine procedure. The operating time was 2 h and 58 min, and the amount of blood loss was 45 ml. The patient did not develop any postoperative complication. Anemia improved, and the patient was discharged 5 days after the operation. To date, i.e., a year after the operation, no recurrent cholelithiasis or gallstone reformation has been observed. Using the ellipsoid method, abdominal US revealed 64% shrinkage in the gallbladder size [2], suggesting that a successful contractile function of the gallbladder has been maintained (Fig. 4).

2. Discussion

The standard treatment for HS cases complicated by symptomatic gallstones is simultaneous splenectomy and cholecystectomy. However, surgical management has recently changed [3]. The disappearance rate of jaundice is generally high with splenectomy for treating HS; therefore, preventive cholecystectomy is not usually performed [4]. In other words, the risk for gallstone reformation would be remarkably reduced, and the gallbladder could be preserved if gallstones inside the gallbladder were removed with concomitant splenectomy.

Gallbladder-preserving cholecystolithotomy was first reported by Kelett et al. in Japan in 1989 [5]. In this procedure, percutaneous transhepatic gallbladder drainage was performed under US guidance with a dilator that dilates the duct up to approximately 16 Fr so that a choledochofiberscope can pass through. However, the drain had to be placed for an extended period of time, which was a problem. Therefore, Qian et al. reported a method for cholecystolithotomy by percutaneously lifting the fundus of the gallbladder to the abdominal wall and fixing it using forceps [6]. In our case, we fixed the fundus of the gallbladder using one of the incision sites of the port necessary for laparoscopic splenectomy. It is considered to be a safe method because the gallbladder can be held and lifted while observing the organ with a laparoscope.

The greatest advantage of this method is the lack of injury to the bile duct. Bile duct injury has been one of the most severe complications of laparoscopic cholecystectomy, with its occurrence rate ranging from 0.4% to 4% [7]. It can also prolong the hospital stay. Another disadvantage of laparoscopic cholecystectomy is postcholecystectomy syndrome, which comprises symptoms of the upper abdomen such as discomfort, dyspepsia, and diarrhea that coincide with symptoms experienced by patients before laparoscopic cholecystectomy. The incidence of postcholecystectomy syndrome reportedly ranges from 5% to 40% [8,9]. Cholecystolithotomy can potentially avoid postcholecystectomy syndrome.

Yi-Ping et al. reported that of 439 cases that underwent percutaneous cholecystolithotomy during the past 10 years, 182 (41.6%) had developed recurrent stone formation [10]. Li et al. reported that a high BMI and a family history of cholelithiasis were risk factors for recurrent stone formation [11]. However, both studies were performed in adult patients, and there are no large-scale investigations currently undertaken for pediatric patients with HS.

This case demonstrates that in HS complicated by gallstones, the risk for recurrent cholelithiasis is low after removing gallstones with percutaneous cholecystolithotomy and simultaneous...
splenectomy. Therefore, this approach should be considered as a useful method in this patient group, which could possibly preserve the gallbladder.

3. Conclusion

In a patient with HS and cholelithiasis, it was possible to perform gallbladder-preserving cholecystolithotomy and splenectomy at the same time with no complications. Hence, further case accumulation and investigations should be conducted, considering that the preservation of gallbladder can be an option for HS complicated by gallstones, in which the risk for recurrent cholelithiasis disappears with splenectomy. There is no unnecessary organ in a human body; it may be significant to review for cholecystectomy, particularly in children.

Conflicts of interest

The authors declare no conflicts of interest in association with the present study.

References