Laparoscopic single stage procedure for perforated choledochal cyst

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A B S T R A C T
Spontaneous perforation of choledochal cyst is a rare presentation. Complicated choledochal cysts are routinely managed by staged open surgical procedures for excision and reconstruction [1, 2]. Complex laparoscopic surgery in infants is an evolving field but no reports have been published describing a minimal invasive approach to the management of spontaneous perforation of choledochal cyst. We report a case of perforated choledochal cyst in a 12 week old, female which was managed by single stage laparoscopic resection of the cyst with biliary reconstruction. She presented with fever and bilious vomiting for 4 days. She had chemical pancreatitis and further work up showed her to have a perforated choledochal cyst. After conservative treatment of her pancreatitis, she underwent laparoscopic resection of the choledochal cyst with reconstruction. She had an uneventful postoperative recovery. This is the first ever reported case of laparoscopic management of a perforated choledochal cyst in infants which was managed by single stage laparoscopic surgery with immediate reconstruction.

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Choledochal cyst is a rare congenital disorder characterized by abnormal dilatation of the biliary system. Classic presentation includes the triad of abdominal pain, jaundice and palpable mass in children younger than 10 years of age [1–4]. Spontaneous perforation of choledochal cyst is a rare presentation with or without peritonitis [5]. There are several reports of the management of complex pancreatico-biliary issues using minimally invasive techniques. These include reports of successful laparoscopic resection and reconstruction of symptomatic but non perforated choledochal cysts even in neonates [6]. Perforated cysts are generally managed by two-stage or single-stage open procedures, with recent trend toward single-stage open procedure [7,8].

1. Case report

We present a case of a three month old female diagnosed with a perforated choledochal cyst, successfully managed by laparoscopic single stage resection and reconstruction with Roux-en-Y hepaticojejunostomy.

She was born at 36 weeks by cesarean section and did not have any other previous significant medical, surgical or birth history. She presented to the emergency room secondary to fever, intermittent cry and non bilious vomiting of 4 days duration. Her blood tests at presentation were significant for a leukocytosis of 24.4 × 10^9/L and elevated lipase at 1089 U/L with a normal amylase level. Her bilirubin was slightly elevated at 2.1 mg/dL and her Gamma Glutamyl Transpeptidase level was raised to 412 U/L. After initial resuscitation and hydration, imaging studies were obtained.

Abdominal ultrasound showed ascites and prominent distal common bile duct (CBD). Magnetic resonance cholangiopancreatography (MRCP) showed fusiform dilatation of the CBD with the dilatation extending up to the head of the pancreas. The distal part of CBD appeared normal consistent with a Type 1A choledochal cyst. The cyst measured 1.3 × 1 × 0.7 cm in dimension (Fig. 1). The final diagnosis based on the history, clinical examination and investigations was perforated type I A choledochal cyst.

She was treated with intravenous fluids, bowel rest and empiric antibiotics. After 3 days of conservative management, the child improved dramatically and a follow up USG showed improving ascites and the lab values improved as well. At that point the child was scheduled for a diagnostic laparoscopy, resection of choledochal cyst and biliary reconstruction.
Laparoscopy was performed using 4 ports (Fig. 2). The choledochal cyst was identified with its site of perforation in the anterior wall with surrounding inflammation. The cyst extended distal to the insertion of the cystic duct up to duodenum where its caliber normalized (Fig. 3). The gallbladder was left attached to the liver bed and was helpful for cephalad retraction of liver. The hepatic artery and portal vein were separated from the posterior wall of the cyst. The cyst was transected proximally at the level of insertion of the cystic duct and distally an endoclip was applied to the common bile duct where its diameter normalized and the choledochal cyst transected.

We chose to perform Roux-en-Y hepaticojejunostomy for biliary drainage, and exteriorized the proximal jejunum through an enlarged umbilical incision. Jejunum was transected at 15 cm from the ligament of Treitz. A 25 cm Roux-en-Y limb was measured and a stapled jejuno-jejunostomy was constructed extracorporeally. The roux limb was delivered back and pneumoperitoneum was established again. An ante-colic hepaticojejunostomy using 5.0 interrupted vicryl intracorporeal sutures was fashioned (Fig. 4). Cholecystectomy was completed and a Jackson–Pratt drain was left at the site of hepaticojejunostomy. EBL was 15 mL. Operating time was 280 min.

Postoperatively, the child was transferred to the intensive care unit overnight for observation. She remained on intravenous fluids postoperatively. She had stooling on postoperative day 2 and diet was started the same day. The Jackson-Pratt drain was removed on the fourth postoperative day. By day 5, she was able to tolerate up to 90 mL of feeds every 3 h and was discharged home.

She was followed up as an outpatient 6 weeks after surgery and her postoperative recovery has been uneventful.

2. Discussion

Choledochal cyst is a dilatation or diverticulum of all or part of the common bile duct. The incidence varies with geographical location ranging from 1:2,000,000 to 1:13,000. It is seen more commonly in Asians and there is a female preponderance up to 3:1 [9–11].

Morphological classification of choledochal cysts by Alonso-Lej et al. was revised by Todani et al. in 1977 to include five different types including two different subtypes for type I cyst based on the extent of involvement of the duct [11]. Type I cyst which includes fusiform dilatation of the extra hepatic duct is the most common type and accounts for up to 85% of cases [12].

The majority of choledochal cysts present before the age of 10 and clinical presentation includes recurrent abdominal pain with RUQ mass (30–60%), conjugated hyperbilirubinemia (80%) and or failure to thrive [3,4]. The classic triad of jaundice, right upper quadrant mass and pain is known to occur only in a minority of patients [13,14]. Non-specific symptoms include fever, nausea, vomiting, pruritus or loss of weight. Biliary cysts may also be diagnosed incidentally during prenatal ultrasound or asymptomatic patients undergoing imaging for unrelated causes.

If left untreated, choledochal cysts may become symptomatic with abdominal pain and may be complicated by cholangitis likely from biliary stasis. Biliary stasis may also predispose to
choledocholithiasis. Untreated cysts carry significant risk of cholangiocarcinoma in the long term with risks reaching up to 2.5%–26% which also carries a very poor prognosis due to delayed presentation and advanced disease status [15,16]. Spontaneous perforation of a choledochal cyst is a rare event which may or may not present with features of pancreatitis or peritonitis and has been reported in 1–2% of cases [5,17–19]. Some case series have reported higher perforation rates between 8.3 and 18% [20–22]. Etiologies considered to cause perforation include mural embryopathy/weakness of the wall of the common bile duct, congenital anomalies of the pancreatico-biliary junction, pancreatitis, trauma, parasitic infections etc [5]. Sudden rise in intracystic pressure from outflow obstruction secondary to obstruction of the common channel by a protein plug and weakness of the cyst wall from recurrent episodes of cholangitis are considered as likely cause of spontaneous perforation [25,26]. These patients present with acute biliary peritonitis. Contained or sealed perforations may present less acutely with bilioma or pseudocyst. Absence of significant peritoneal signs in some patients could be attributed to the sterile nature of bile leading to non-specific symptoms. This may potentially delay diagnosis and surgical intervention. There are reports of missed perforations identified intra operatively during definitive surgery [8,20]. Chiang et al. case series included six patients with perforated choledochal cyst and none of them had clinical evidence of peritonitis at the time of presentation [27].

Traditionally, a two-stage approach is recommended in the management of a perforated choledochal cyst. During the first stage, exploratory laparotomy, peritoneal lavage and external biliary drainage preferably by a T-tube were performed [28]. The second stage of the procedure includes excision of the cyst with biliary reconstruction 6–12 weeks later. Reconstructive options include hepaticoduodenostomy or hepaticojejunostomy, although the later is preferred more often. The principle of resection, regardless of approach, should be complete excision of the cyst to reduce the risk of malignancy. Occasionally, a proximal cuff is retained when met with small sized ducts to aid in the construction of hepaticojejunostomy [29].

Laparoscopic assisted of choledochal cyst was first described by Farello et al. in 1995 [29]. Since then it has been accepted into clinical practice as an alternative to open surgery. Better visualization and accuracy of dissection around the duct and vital structures around the porta hepatis with laparoscopy has been attributed to the less blood loss reported with laparoscopic surgery [30]. Laparoscopic approach has also been reported with early institution of diet and shorter hospital stay. Longer operative time associated with the learning curve for this advanced laparoscopic approach has been shown to improve after the first few cases. Meta analysis by Shen et al. (2015) involved 1016 patients from seven retrospective studies including 408 cases of laparoscopic resection of choledochal cyst. They recommended laparoscopy as feasible and

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**Fig. 2.** Laparoscopic trocar placement for perforated choledochal cyst excision.

**Fig. 3.** Anatomy of perforated choledochal cyst with the arrow showing the site of perforation.
safe treatment for choledochal cyst and opined it to become the gold standard with further refinement in techniques [31,32]. In this case report we utilized the laparoscopic single stage approach in the management of perforated choledochal cyst. To our knowledge, this is the first reported case in the literature. In our experience the inflammation associated with perforation of the choledochal cyst made the dissection more challenging and we believe it did attribute to the long operative time.

3. Conclusion

We anticipate that with the continued improvement of the minimally invasive techniques, this approach will become the preferred method when approaching this disease.

Conflicts of interest
None.

References