CASE REPORT

Case Report: Spontaneous perforation of choledochal cyst in an infant: Successful management in a centre with limited means

Rafey Abdul Rahman, Umesh Kumar Gupta
Department of Pediatric Surgery, Uttar Pradesh University of Medical Sciences, Saifai, Etawah, Uttar Pradesh, 206130, India

Abstract

Background: Biliary peritonitis due to a ruptured choledochal cyst (CC) is a rare occurrence. The difference between bile duct perforation (BDP) and ruptured choledochal cysts continues to be a matter of debate. Simple drainage, T tube placement and cholecystostomy have been proposed as the initial treatment of choice. Definitive surgery in the form excision of the CC and hepatico-enterostomy has been described as the ideal treatment option. We report a successful management of a unique case of perforated choledochal cyst in an infant who presented with biliary peritonitis.

Case report: An 8 months old female child presented with biliary peritonitis as result of spontaneous perforation of a choledochal cyst. The patient was successfully managed initially by placement of T tube in the perforated cyst followed by a T tube cholangiogram. Definitive surgery was performed 5 weeks after the initial surgery in which cyst was excised and hepatico-duodenostomy was performed. The child is currently in follow up and doing well.

Conclusion: Perforated CC can present as acute abdomen sometimes having only subtle signs. In absence of any previous established diagnosis of CC and trained radiological support the condition becomes challenging to diagnose preoperatively. External T-tube drainage followed by T-tube cholangiogram can help in delineating the anatomy. Cyst excision along with hepaticoportoentersomy remains the gold standard definitive treatment.

Keywords
Spontaneous Choledochal cyst perforation, bile duct perforation, T tube cholangiogram, hepatico-duodenostomy
Introduction
Biliary peritonitis due to a ruptured choledochal cyst (CC) is a rare occurrence \[^1\], 15–25% of the cases of CC present with a classical triad of pain, lump and jaundice \[^1\]. Cholangitis, pancreatitis, progressive biliary cirrhosis, stones in the cyst, portal hypertension and malignant transformation in the biliary tree are well known complications seen in CC \[^3\]. Bile duct perforation and choledochal cysts are considered as inter-related pathology having a common cause in the form of pancreato-biliary maljunction (PBM) \[^4\]. The difference between bile duct perforation (BDP) and ruptured choledochal cysts continues to be a matter of debate. Simple drainage, T tube placement and cholecystostomy have been proposed as the initial treatment of choice. Definitive surgery in the form excision of the CC and hepatoco-enterostomy has been described as the ideal treatment option.

In this case report we describe the successful management of a case of a ruptured choledochal cyst in a female infant at a centre with limited means.

Case report
An eight months old female child of South Asian ethnicity was admitted in the department of Paediatric medicine in April 2019 with complaints of progressive abdominal distension, non-passage of stools and mild generalised pain in abdomen for 3 days. There was no history of vomiting, fever or jaundice. On clinical examination features of peritonitis were observed. A rectal examination was performed and was found to be insignificant. An X Ray of the abdomen in the erect position was performed, with results suggestive of ascites with paucity of air in the pelvis. An ultrasound of the abdomen revealed free fluid in the abdomen, along with mildly dilated intrahepatic biliary radicles. The patient’s complete blood work up revealed low levels of haemoglobin at 9.8 grams (Normal range: 12–14 gm/dl), while the rest of the parameters were within the normal limits. In the liver function tests (LFT), serum glutamic oxaloacetic transaminase (SGOT) and SGPT were mildly elevated with values of 140 and 143 units/litre respectively. The remaining LFT parameters were within the normal range. As peritonitis was suspected an emergency laparotomy was performed. Biliary peritonitis secondary to a perforation in the common bile duct (CBD) was observed and the CBD was grossly dilated. The wall of the small and large bowel was oedematous but otherwise appeared normal. Bile stained ascitic fluid was drained and a “T tube” was placed through the perforation site in the CBD. (Figure 1). Post operatively bile output through the “T tube” was 150–200 ml per day which was high. We expected it to be around 50 ml per day thinking that some amount of bile may pass into the gut through the “T tube”. However, as the system was obstructed the output through the “T tube” remained persistently high. The child was transfused with packed red blood cells and fresh frozen plasma post-operatively. Other supportive treatment in the form of intravenous (IV) fluid, antibiotics and analgesics were administered. IV antibiotics included Cefotaxime (90 mg/kg/day in 3 divided doses), Amikacin (15mg/kg/day once a day) and Metronidazole (20mg/kg/day in 3 divided doses) covering gram positive, gram negative and anaerobic bacteria. The child was started on oral feeds from post-operative day 5.

The child was discharged on postoperative day 8 with the “T tube” but was readmitted again with features of dehydration. The patient was stabilised again and a “T Tube” cholangiogram was performed in the third week post-surgery (Figure 2) which revealed a dilated CC along with dilated intrahepatic biliary radicles with no contrast flow into the duodenum (Figure 3).
Our case presented with features of peritonitis and USG demonstrated free fluid in the abdomen. In the absence of a previous diagnosis and no past history suggestive of choledochal cyst, it was difficult for us to accurately diagnose the condition preoperatively. Since the centre had no facility for hepatobiliary scintigraphy and MRCP, or a trained radiologist, we went ahead with laparotomy and were able to diagnose and treat the patient with external biliary diversion. Since there was no precipitating factor for perforation of CC like trauma, we presumed the rupture was spontaneous.

Through our intraoperative findings and the “T tube” cholangiogram we were able to identify the CC and perforation site. Absence of a trained radiologist and with no close alternative radiological centre, as our institution is situated in a rural area, we were unable to perform additional diagnostic imaging like MRCP. In the post-operative week 5 we went ahead with definitive surgery. We performed complete cyst excision and a hepatico-duodenostomy. We chose hepatico-duodenostomy over hepatico-jejunostomy for various reasons. Firstly, it reduced the duration of surgery; secondly, the duodenum was mobile reaching easily up to the common hepatic duct; and thirdly the literature shows similar success rates and complications for both procedures.

It was also important for us to distinguish BDP from a ruptured CC as management of both differs. BDP occurs exclusively in infants less than 20 weeks of age and perforation mostly occurs at the junction of cystic duct and common hepatic duct. Simple surgical drainage often leads to closure of BDP but a perforated CC requires cyst excision and hepatico-enterostomy.

**Conclusion**

Perforated CC can present as biliary peritonitis sometimes having only subtle signs. In absence of any previous established diagnosis of CC and trained radiological support becomes challenging to diagnose this preoperatively. External T-tube drainage followed by T-tube cholangiogram can help in revealing the anatomy of the hepatobiliary tree. Cyst excision and hepatico-enterostomy remains the gold standard definitive treatment.

**Data availability**

**Underlying data**

All data underlying the results are available as part of the article and no additional source data are required.

**Grant information**

The author(s) declared that no grants were involved in supporting this work.
References

The benefits of publishing with F1000Research:

- Your article is published within days, with no editorial bias
- You can publish traditional articles, null/negative results, case reports, data notes and more
- The peer review process is transparent and collaborative
- Your article is indexed in PubMed after passing peer review
- Dedicated customer support at every stage

For pre-submission enquiries, contact research@f1000.com