CASE REPORT

Case Report: Spontaneous perforation of choledochal cyst in an infant: Successful management in a centre with limited means [version 1; peer review: 1 approved, 1 approved with reservations]

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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 hepaticoduodenostomy remains the gold standard definitive treatment.

Keywords

Spontaneous Choledochal cyst perforation, bile duct perforation, T tube cholangiogram, hepatico-duodenostomy

Open Peer Review

Approval Status ? ✓

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1. Anand Pandey , King George's Medical University, Lucknow, India		
Gurmeet Singh , King George's Medical University, Lucknow, India		
2. Richard Kozarek , Virginia Mason Medical Center, Seattle, USA		
Any reports and responses or comments on the article can be found at the end of the article.		

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Introduction

Biliary peritonitis due to a ruptured choledochal cyst (CC) is a rare occurrence^{1,2}. 15–25% of the cases of CC present with a classical triad of pain, lump and jaundice¹. Cholangitis, pancreatitis, progressive biliary cirrhosis, stones in the cyst, portal hypertension and malignant transformation in the biliary tree are well known complications seen in CC³. 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 hepatico-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).

Figure 1. Intraoperative photograph showing perforated choledochal cyst.

Figure 2. T tube cholangiogram.

Figure 3. Intraoperative photograph revealing the anatomy during the second surgery.

We waited for another 2 weeks and built up the child nutritionally for the definitive surgery. In view of the absence of a trained radiologist and a magnetic resonance imaging (MRI) scanner at the institute, Magnetic Resonance Cholangiopancreatography (MRCP) could not be done. Hence, we decided to proceed with definitive surgery based on the “T Tube” cholangiogram imaging and previous operative findings. A laparotomy was performed. The dilated cyst was excised completely after separating it from the portal vein. The distal end of the cyst was blind ending and was not communicating with the duodenum. Distally the cyst was excised at its narrowest portion on the duodenum. Proximally, common hepatic duct was identified and then a hepatico-duodenostomy was performed. The child recovered well and started passing cholic stools from post-operative day 3. The child was started on oral feeds from post-operative day 5. The patient was discharged on post-operative day 8 on full oral feeds. The patient is currently in our follow up and doing well.

Discussion

CC is considered to be a congenital anomaly as it seen in foetuses and newborns. Its etiology is unknown, however many theories exist including abnormalities in the pancreato-biliary junction (PBJ)⁴, distal obstruction with congenital weakness of the duct⁵ and ischemia of the bile duct⁶. CC rupture and resulting peritonitis is a rare complication, reported in only 1.8% to 2.8% of cases².

Ultrasonography is a useful diagnostic investigation for CC when the cyst is intact, but in cases of perforation of the cyst, where the cyst decompresses, free fluid in the abdomen may be the only finding. Pre-operative hepato-biliary scintigraphy can demonstrate bile leak from the extra hepatic biliary tree in biliary channel perforation⁷. MRCP is a non-invasive method of evaluating the biliary tree, and owing to its excellent anatomical and contrast resolution, it can be used to diagnose bile duct perforation, perforated CC and PBM⁸. The cost and duration of MRCP imaging limits its use.

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 procedures⁹.

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 duct³. 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.

Consent

Written informed consent for publication of the patients' clinical details and clinical images was obtained from the parents of the patient.

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.

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Open Peer Review

Current Peer Review Status: ? ✓

Version 1

Reviewer Report 17 February 2020

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Richard Kozarek

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The case report by Rahman and Gupta describing an 8-month-old female with spontaneous perforation of a congenital choledochal cyst (CC) demonstrates the ability to provide appropriate patient care despite a paucity of diagnostic options. These providers had only an abdominal ultrasound which demonstrated intra-abdominal fluid following spontaneous perforation of a CC. Surgical exploration revealed bile peritonitis and a local bile duct perforation treated with a T-tube decompression. Diagnosis as a CC was only made at subsequent cholangiography leading appropriately to cyst resection and choledochal-enteric biliary anastomosis. All this was done without access to MRI/MRCP, apparently a CT scan, or access to an onsite radiologist. As a gastroenterologist who has provided ERCP support to pediatric patients for > 30 years, one who has been involved in the treatment of more than 100 adult patients with CC, and an individual who has been involved in education and training programs in the developing world for a comparable time period, I salute their diagnostic tenacity, appropriate treatment, and apparent outcomes.

In areas in which more resources are available, it is unlikely that this patient would have been explored without a CT scan, and if a grossly dilated common bile duct had been defined, a subsequent MRCP to define the extent of the cyst as used the pancreaticobiliary junction to define the type and extent of surgical resection. Although the authors suggest that the patient is doing well after CC resection and hepaticoduodenostomy, there is no report of the follow-up times. A follow-up 2 weeks after surgery is much less important than a 1 year follow-up with normal liver enzymes and growth parameters. The follow-up interval needs to be defined.

Is the background of the case's history and progression described in sufficient detail?

Yes

Are enough details provided of any physical examination and diagnostic tests, treatment given and outcomes?

Yes

Is sufficient discussion included of the importance of the findings and their relevance to

future understanding of disease processes, diagnosis or treatment?

No

Is the case presented with sufficient detail to be useful for other practitioners?

Yes

Competing Interests: No competing interests were disclosed.

Reviewer Expertise: Areas of expertise range from therapeutic endoscopy, inflammatory bowel diseases, and practice economics

I confirm that I have read this submission and believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard.

Reviewer Report 22 October 2019

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- It is an unusual report where patient was managed successfully.
- Attention is needed towards grammatical errors, which have crept in at places.
- Specific values of bilirubin are missing. Was there any history of clay coloured stools in retrospective assessment?
- There is repetition in second paragraph page 4 about MRCP. It may be deleted.
- Literature review may include other articles on ruptured choledochal cyst such as - Management of rupture of choledochal cyst. Ahmed I *et al.* Indian J Gastroenterol. 2011 Mar;30(2):94-6¹.

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Is the background of the case's history and progression described in sufficient detail?

Yes

Are enough details provided of any physical examination and diagnostic tests, treatment given and outcomes?

Partly

Is sufficient discussion included of the importance of the findings and their relevance to future understanding of disease processes, diagnosis or treatment?

Yes

Is the case presented with sufficient detail to be useful for other practitioners?

Partly

Competing Interests: No competing interests were disclosed.

Reviewer Expertise: Pediatric Surgery

We confirm that we have read this submission and believe that we have an appropriate level of expertise to confirm that it is of an acceptable scientific standard, however we have significant reservations, as outlined above.

Author Response 05 Nov 2019

umesh gupta, Uttar Pradesh University of Medical Sciences, Saifai, Etawah, India

Specific values of bilirubin are missing.

Specific values of bilirubin was total 0.8 mg %.

Was there any history of clay coloured stools in retrospective assessment?

No clay coloured stools was present in retrospective assessment .

Competing Interests: No competing interests were disclosed.

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