J Ped Surg Case Reports 10 (2016) 42–46

Contents lists available at ScienceDirect

Journal of Pediatric Surgery CASE REPORTS

journal homepage: www.jpscasereports.com

Management of a massive choledochal cyst in a 12 year-old girl: Which imaging modalities should be performed preoperatively?

Shabnam Hamidi [a], Michael H. Livingston [a,b], Amar Alnaqi [a,c], Yasmine Yousef [a], J. Mark Walton [a,c,*]

[a] McMaster Pediatric Surgery Research Collaborative, McMaster Children's Hospital, McMaster University, Hamilton, Ontario, Canada
[b] Clinician Investigator Program, McMaster Children's Hospital, McMaster University, Hamilton, Ontario, Canada
[c] Division of Pediatric Surgery, McMaster Children's Hospital, McMaster University, Hamilton, Ontario, Canada

ARTICLE INFO

Article history:
Received 18 January 2016
Received in revised form
27 April 2016
Accepted 30 April 2016

Key words:
Choledochal cyst
Cholangiography
Diagnostic imaging

ABSTRACT

A previously well 12 year-old girl presented to the emergency department with a three-day history of fatigue, pruritus, and epigastric discomfort. Abdominal ultrasound revealed a cystic mass in the right upper quadrant measuring 17.0 × 13.2 × 11.7 cm. Magnetic resonance imaging (MRI) confirmed the diagnosis of a choledochal cyst and hepatobiliary iminodiacetic acid (HIDA) scintigraphy demonstrated a communication between the cyst and biliary tree. Percutaneous cholangiography was also performed but did not provide additional diagnostic information. The patient was brought to the operating room a few days later for open resection of the choledochal cyst, cholecystectomy, and Roux-en-Y hepaticojejunostomy. Frozen sections were obtained to ensure complete excision of cyst mucosa. The patient continues to do well more than 18 months post-operatively with no signs of recurrent inflammation or malignancy. Multiple imaging modalities may be used preoperatively to assess the anatomy and subtype of choledochal cysts. The role of invasive options, such as percutaneous cholangiography or endoscopic retrograde cholangiopancreatograpy (ERCP), remains limited and unclear. Here, we present a potential imaging algorithm to assist with preoperative workup and avoid invasive diagnostic procedures whenever possible.

Choledochal cysts are dilatations of the biliary tree, which can involve the extrahepatic ducts, intrahepatic ducts, or both. This condition often presents with one or more components of the classic triad: abdominal pain, jaundice, and a palpable mass in right upper quadrant [1]. The Todani classification of choledochal cysts, an expansion of the system originally developed by Alonso-Lej et al., is used to classify the type of cyst based on the location and morphology of bile duct dilatation. Types range from I (true choledochal cyst) to V (Caroli's Disease) [2] (Fig. 1). If left untreated, choledochal cysts can cause significant morbidity and mortalitydueto recurrent cholangitis and the development of cholangiocarcinoma.

A variety of imaging modalities are available to diagnose the presence of a choledochal cyst and assess its anatomy preoperatively. These include ultrasound, computed tomography (CT), magnetic resonance cholangiopancreatography (MRCP), and biliary scintigraphy (i.e., hepatobiliary iminodiacetic acid (HIDA) scan) [3]. Some clinicians advocate for the use of percutaneous cholangiography and/or endoscopic retrograde cholangiopancreatography (ERCP) as adjuncts, since these procedures provide detailed images of the biliary tree, can rule out the presence of a pancreaticobiliary malformation, and provide thorough evaluation of the cyst itself to aid in classification [4–7]. Unfortunately, these procedures are invasive, require conscious sedation or general anesthesia, and with respect to ERCP, have a non-negligible risk of pancreatitis.

In this report, we present the case of an adolescent girl diagnosed with an unusually large choledochal cyst. She was assessed with a variety of imaging modalities preoperatively. While she experienced an excellent clinical outcome in hospital and in medium-term follow-up, we wonder whether all of these imaging techniques were helpful or necessary. We hope this case will encourage others to reflect on their treatment decisions with

Abbreviations: ERCP, endoscopic retrograde cholangiopancreatography; HIDA, hepatobiliary iminodiacetic acid; MRCP, magnetic resonance cholangiopancreatography; MRI, magnetic resonance imaging.

* Corresponding author. McMaster Children's Hospital, 1200 Main Street West, Room 4E3, Hamilton, Ontario, Canada, L8N 3Z5. Tel.: +1 905 521 2100x75244.
E-mail address: waltonj@mcmaster.ca (J.M. Walton).

http://dx.doi.org/10.1016/j.epsc.2016.04.028

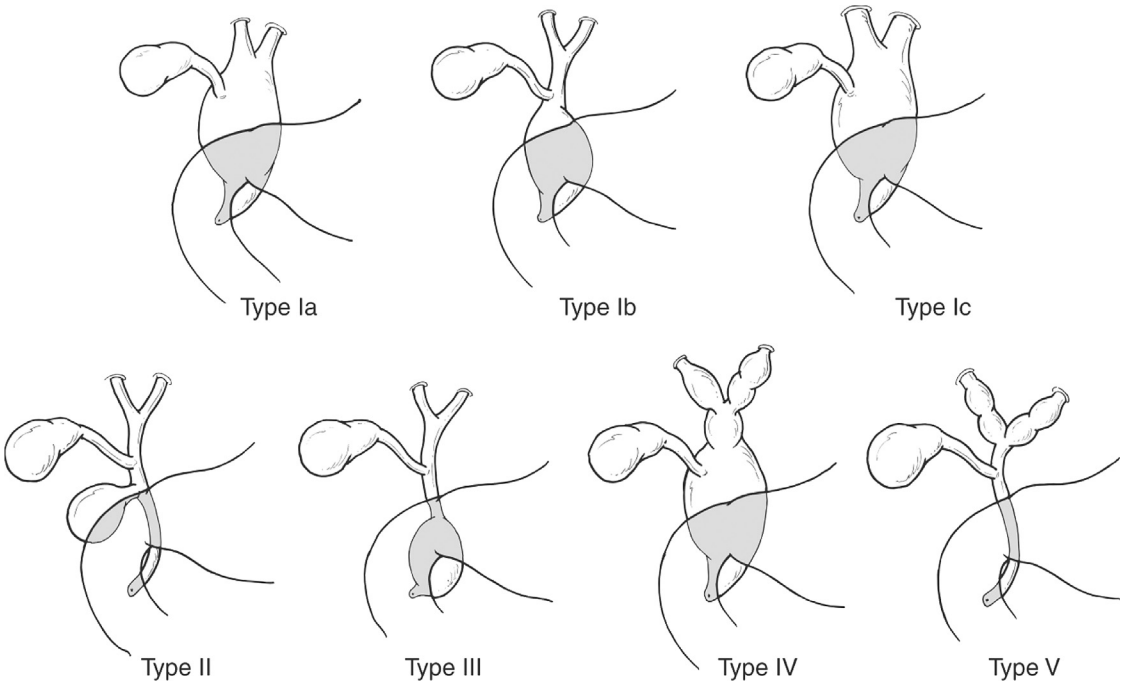

Fig. 1. Todani classification of choledochal cysts (reprinted with permission from Elsevier [2]): Ia (cystic dilatation), Ib (segmental dilatation), Ic (diffuse dilatation), II (diverticulum), III (choledochocele), IV (multiple intrahepatic and extrahepatic cysts), V (intrahepatic cysts only).

regards to preoperative imaging of choledochal cysts. We also present a possible algorithm to guide clinicians in selecting imaging modalities that maximize clinical utility, while minimizing risk to patients.

1. Case report

A 12 year-old girl of Asian descent presented to the emergency department in a community hospital with facial cellulitis. She was given a prescription for oral antibiotics and discharged home. Over the next few days, she developed dark urine, pruritus, fatigue, scleral icterus, and mild epigastric pain. There was no fever, nausea, emesis, or weight loss. She returned to the emergency department five days later with ongoing abdominal pain and was found to have elevated liver enzymes, bilirubin, and platelets. An abdominal ultrasound revealed a cystic mass in the right upper quadrant measuring 17.0 × 13.2 × 11.7 cm.

The patient was transferred to a tertiary children's hospital the following day for assessment and further investigations. On arrival, vital signs were within normal limits and the patient was mildly jaundiced without stigmata of liver disease. The abdomen was soft and non-distended, but examination was significant for a tender mass in the right upper quadrant. The facial cellulitis from a few days prior had resolved.

Initial laboratory investigations revealed markedly elevated liver enzymes, including alanine transaminase of 1702 units/L, aspartate transaminase of 695 units/L, alkaline phosphatase of 1087 units/L, and gamma-glutamyl transpeptidase of 657 units/L. Total bilirubin was elevated at 51 umol/L with a conjugated bilirubin of 39 umol/L. Lipase was normal at 19 units/L. White blood cell count was also normal at 4.0×10^9/L. Inflammatory markers were moderately elevated, including a platelet count of 639×10^9/L, C-reactive protein of 28.9 mg/L, and erythrocyte sedimentation rate of 58 mm/h.

The following day, the patient underwent magnetic resonance imaging (MRI) of the abdomen and MRCP, which confirmed the diagnosis of a choledochal cyst (Fig. 2). This examination also revealed dilatation of the cystic duct up to 1.7 cm, dilation of common hepatic duct up to 3 cm, and mild dilatation of the intrahepatic ducts. There was no lymphadenopathy, free fluid, or gallstones. The common bile duct could not be adequately visualized. HIDA scan confirmed the presence of a connection between the choledochal cyst and biliary tree (Fig. 3). Delayed

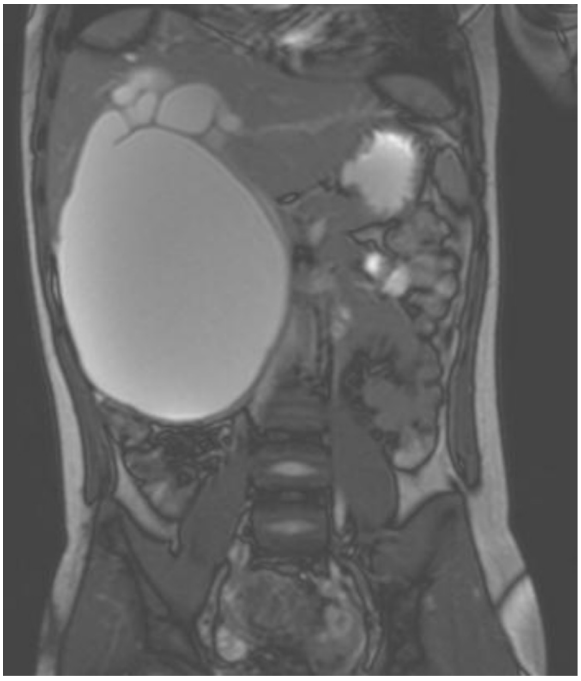

Fig. 2. Magnetic resonance images showing a massive choledochal cyst, dilated cystic and common bile ducts, and mildly dilated intrahepatic ducts.