CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 28 (2016) 42–44

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Ulcerated choledochocele: A case report

Dr. E. Ray-Offor[a,*], S.N. Elenwo[a], P.O. Igwe[a], C. Ngeribara[b]

[a] Department of Surgery, University of Port-Harcourt Teaching Hospital, Rivers State, Nigeria
[b] Total E & P Clinic, Port Harcourt, Rivers State, Nigeria

ARTICLE INFO

Article history:
Received 7 August 2016
Accepted 11 August 2016
Available online 13 September 2016

Keywords:
Gastrointestinal bleeding
Choledochocele
Case report

ABSTRACT

BACKGROUND: The cystic dilatation of the biliary tract is an uncommon anomaly. Choledochocele, a cystic dilatation of the distal common bile duct, rarely presents clinically as massive gastrointestinal bleeding.
AIM: This is to report a very rare disease condition and highlight minimal access options in surgical care.
CASE SUMMARY: A 13 year-old boy was referred with a day history of sudden onset of passage of bright red blood per rectum with a fainting episode. There was no anal protrusion, jaundice, recurrent epigastric pain nor bleeding from any other orifice. An initial endoscopic assessment of the upper digestive tract showed profuse bleeding from a sub-mucosal mass in the region of ampulla of Vater. Emergency laparotomy revealed small intestine filled with blood from duodenum to ileum. A duodenotomy showed a cystic mass with an ulcerated mucosa at the dome containing bilious fluid in the second part of the duodenum. The cyst was de-roofed and marsupialized. Post-operative recovery was complicated by features of adhesive small bowel obstruction on the 9th post op day and treated by laparoscopic adhesiolysis. He was discharged home in good clinical state.
CONCLUSION: Choledochocele is a differential diagnosis in the endoscopic finding of a submucosal mass in the second part of the duodenum. An initial oesophagogastroduodenoscopy endoscopy is necessary in the evaluation of massive lower gastrointestinal bleeding.

1. Introduction

The congenital dilatation of the biliary duct- choledochal cyst (CC) is a rare disease condition. Choledochocele is a cystic dilatation of the intra-duodenal portion of the distal common bile duct (CBD). It is type 3 choledochal cyst according to the Todani et al. classification and comprises 2% of CCs [1]. In this classification, type 1 is a fusiform dilatation of the CBD while type 2 is a diverticulum of the CBD.Dilation of intrahepatic ducts only or intra and extrahepatic ducts are classified as type V and IV respectively. CC is a benign condition that can be associated with serious complications including cholelithiasis, cholangitis, pancreatitis, and malignant transformation [2].

Two thirds of choledochal cysts are diagnosed before the patient is aged 10 years [3]. In the infantile age group, it commonly presents with hepatomegaly, obstructive jaundice and acholic stools, and is indistinguishable from biliary atresia in the absence of a palpable mass in the right side of the abdomen. Approximately 20% are diagnosed in adults, including elderly patients [4]. The presence of bleeding with choledochocele is rare and the radiological distinction of choledochocele from duodenal cysts is a diagnostic challenge [5]. To the best of our knowledge this is the first reported case of an ulcerated choledochocele in world literature.

It is with excitement we present this rare disease condition in a case report emphasizing the need for initial upper gastrointestinal tract assessment in a case of massive bleeding per rectum and to highlight the role of laparoscopy in the management of a surgical complication.

2. Case report

A 13 year old obese boy was referred for endoscopic assessment with a sudden onset of passage of bright red blood per rectum and a fainting episode of one day duration. There was no anal protrusion, jaundice, recurrent epigastric pain nor bleeding from any other orifice. He was transfused 2 pints of fresh blood before presentation on account of haemoglobin count of 7.1 g/dl. He had no previous bleeding episode.

On physical examination the boy was in no respiratory distress but pale, afebrile and anicteric. His pulse rate was 108 bpm but the other vital signs were normal. A physical examination of the body systems was unremarkable. The serum electrolyte urea and creatinine values were within the normal range. The liver function test showed: aspartate transaminase AST 15.70 IU/L, alanine transaminase ALT 17 IU/L, Gamma glutaryl transaminase GGT 20.20 IU/L and alkaline phosphatase 127.50 IU/L (normal range 50–128 IU/L). However the clotting profile revealed a prolonged prothrombin

* Corresponding author at: Department of Surgery, University of Port-Harcourt Teaching Hospital, P.M.B.6173, Port Harcourt, Rivers State, Nigeria.
E-mail addresses: erayoffor@yahoo.com, md@oakendoscopycentre.com (E. Ray-Offor).

http://dx.doi.org/10.1016/j.ijscr.2016.08.028

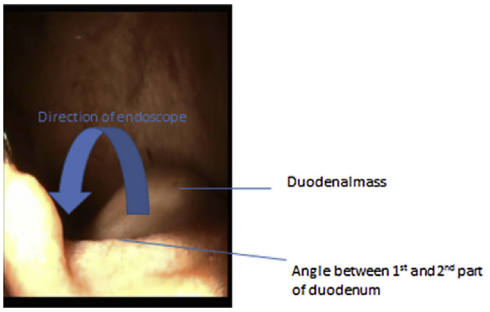

Fig. 1. Duodenoscopy showing duodenal mass.

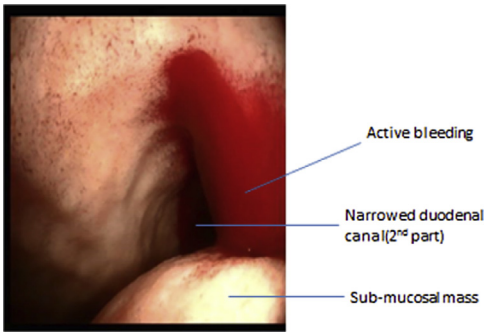

Fig. 2. Ulcerated choledochocele on duodenoscopy.

time with international normalized ratio INR of 1.75. He was given 10 mg of Vitamin K parenterally.

An emergency endoscopic assessment commenced with an oesophago-gastroduodenoscopy using a Karl Storz 13801PKS video-gastroscope (Germany). This showed profuse bleeding in the region of a sub-mucosal mass in the medial aspect of the duodenum (second part) narrowing the lumen (Figs. 1 and 2). The decision for an emergency laparotomy was made and colonoscopy deferred. At laparotomy the small bowel from the 2nd part of the duodenum progressing distally to the ileum was filled with blood. A duodenotomy was performed which revealed a medially located cystic mass with ulcerated mucosa at the dome containing bilous fluid in the region of the ampulla of Vater. The cyst was deroofed and marsupialized.

His immediate post-operative period was uneventful. Sutures were removed on the 7th day and patient was discharged home in stable clinical condition. On the 9th post op day patient came back with severe colicky abdominal pains and vomiting of recently ingested feed. Following relevant clinical and laboratory investigations an impression of adhesive small bowel obstruction was made. An urgent laparoscopy was performed with primary access in the mid clavicular line of the left subcostal region-Palmer's point. Capnoperitoneum was established and optical port inserted through this point. It revealed multiple small bowel and omental adhesions to the post-aspect of the anterior abdominal wall along the midline laparotomy incision. A secondary port was inserted under direct vision in the left lumbar area and adhesiolysis was successfully performed (Fig. 3).

The patient was discharged home on the second day post op in good clinical state. A follow-up visit a week post- surgery was unremarkable.

3. Discussion

The exact cause of CC remains unknown. Several theories have been postulated which include weakness of the bile duct wall, obstruction of distal choledochus, inadequate autonomic innervations, sphincter of Oddi dysfunction and reflux of pancreatic enzymes into the CBD secondary to anomaly of the pancreaticobiliary junction [6–10]. All of these theories are applicable to choledochal cyst type I, III (choledochocele), and IV anomalies, but they cannot be used to explain type II and V choledochal cysts in which the CBD is normal. Perhaps genetic factors play a role [11]. Choledochoceles exist in two main anatomic variations [12]. The more frequent form is that with the common bile duct and pancreatic duct opening separately into the duodenum and the other is a diverticulum arising from the terminal end of the common bile duct at the end of ampulla of Vater and pancreatic duct opens at its usual position.

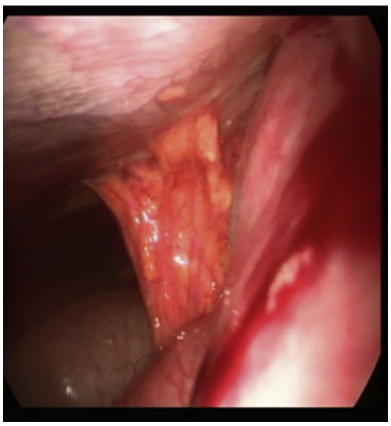

Fig. 3. Laparoscopy showing small bowel adhesions.

Two distinct clinical groups of patients are recognized with regard to age at presentation of CC [13]. The first group is the infantile group consisting of babies younger than 1 year, with or without obvious hepatomegaly, with obstructive jaundice and acholic stools. In contrast, infants older than 1 year, with the so-called adult form of CC, generally have one or more components of the classic triad: pain, jaundice, and a palpable mass. The entire triad is present in fewer than 30% of patients [14]. Jaundice is intermittent and often associated with vague abdominal pain. The index case was not associated with any of the symptoms of the triad. Portal hypertension is a rare complication of CC that may present with bleeding [15]. The various causes of portal hypertension include extrahepatic biliary obstruction leading to secondary biliary cirrhosis, recurrent inflammation leading to portal vein thrombosis and direct compression of the portal vein [16]. There was no clinical evidence of chronic liver disease nor febrile illness in our patient.

The laboratory studies that may be useful for the diagnosis and preoperative evaluation of a patient with a choledochal cyst include direct bilirubin, alkaline phosphatase, serum aspartate aminotransferase (AST), serum alanine aminotransferase (ALT), gamma-glutamyl transferase (GGT), and coagulation profiles. Our patient had normal liver enzymes but a deranged clotting profile. Imaging studies are the cornerstone of diagnosis of CC. Magnetic resonance cholangiopancreatography (MRCP) is the diagnostic test of choice because it offers high resolution detailed images of relevant anatomy, is noninvasive, and does not suffer from complications such as post-procedure pancreatitis [17]. Although ultrasound and CT are useful, MRCP detects most CC with sensitivities from 90 to 100% and specificities from 73 to 100%, with the exception of small choledochoceles and minor ductal anomalies [18].

The differentiation of a choledochocele from a duodenal cyst in imaging studies may prove difficult. This is often resolved by histopathologic examination of resected specimen [4,19]. A duodenoscopy showing a mass in the medial aspect of the second part of the duodenum in the region of the ampulla of Vater with normal

overlying mucosa that sinks softly and then elevates again when pressed with biopsy forceps (pillow sign) is suggestive of a choledochocele. In the index patient there was profuse bleeding from the dome of the cyst mass seen with an end-viewing endoscope. Endoscopic retrograde cholangiopancreatography (ERCP) a useful interventional procedure performed with a side viewing endoscope. It was not a viable option in this case due to poor visibility from the profuse bleeding state.

The management of choledochal cysts has evolved during the last 3 decades. Today, a complete excision of the cyst and biliary bypass has become the treatment of choice. This is necessitated by the risk of malignant transformation in biliary tract which is minimal with choledochocele [20]. Appropriate management of small choledochoceles consists of endoscopic sphincterotomy [21]. A transduodenal excision may be considered for large choledochoceles but may be associated with complications such as gastric outlet obstruction or pancreatitis [22]. In our patient a transduodenal excision with marsupialization was successfully done. It is our suspicion that there was mechanical mucosal injury to a large choledochocele cyst in this patient with deranged clotting profile leading to massive bleeding. An early laparoscopic intervention is a paradigm shift in management of adhesive small bowel obstruction with the benefit of reduced trauma and lower risk of new adhesion compared to laparotomy [23]. A laparoscopic adhesiolysis was safely performed for the small bowel obstruction with good post-operative outcome.

4. Conclusion

Choledochocele is rare to diagnose more so when there is associated bleeding from an ulceration. In the endoscopic evaluation of a case of massive bleeding per rectum an initial upper gastrointestinal endoscopy is essential.

Conflict of interest

The authors have no conflict of interest to declare.

Funding

Personal.

Ethical approval

N/A.

Author contribution

Dr. E. Ray-Offor: Concept/design, write up.
Dr. S.N. Elenwo: Editing.
Dr. P. Igwe: Literature search and write up.
Dr. C. Ngeribara: Data collection.

Consent

A written informed consent was obtained from the parent for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Guarantor

Dr. Emeka Ray-Offor.

References

[1] T. Todani, Y. Watanabe, M. Narusue, K. Tabuchi, K. Okajima, Congenital bile duct cysts: classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst, Am. J. Surg. 134 (2) (1977) 263–269.
[2] C.S. Huang, C.C. Huang, D.F. Chen, Choledochal cysts: differences between pediatric and adult patients, J. Gastrointest. Surg. 14 (2010) 1105–1110.
[3] J.A. O'Neill Jr., Choledochal cyst, Curr. Probl. Surg. 29 (6) (1992) 361–410.
[4] P.A. Lipsett, H.A. Pitt, P.M. Colombani, J.K. Boitnot, J.L. Cameron, Choledochal cyst disease: a changing pattern of presentation, Ann. Surg. 220 (5) (1994) 644–652.
[5] L. Zhu, Z. Lv, J. Liu, W. Xu, Choledochocele: a case report and discussion of diagnosis criteria, Eur. J. Pediatr. Surg. Rep. 3 (2) (2015) 85–89.
[6] H. Bismuth, J. Krissat, Choledochal cystic malignancies, Ann. Oncol. 10 (Suppl. 4) (1999) 94–98.
[7] T. Kato, T. Hebiguchi, K. Matsuda, H. Yoshino, Action of pancreatic juice on the bile duct: pathogenesis of congenital choledochal cyst, J. Pediatr. Surg. 16 (2) (1981) 146–151.
[8] R. Hill, C. Parsons, P. Farrant, M. Sellars, M. Davenport, Intrahepatic duct dilatation in type 4 choledochal malformation: pressure-related, postoperative resolution, J. Pediatr. Surg. 46 (2011) 299–303.
[9] T. Todani, Y. Watanabe, T. Fujii, S. Uemura, Anomalous arrangement of the pancreatobiliary ductal system in patients with a choledochal cyst, Am. J. Surg. 147 (5) (1984) 672–676.
[10] Y. Yamashiro, T. Miyano, K. Suruga, H. Shimomura, K. Suda, M. Matsumoto, H. Nittono, Experimental study of the pathogenesis of choledochal cyst and pancreatitis, with special reference to the role of bile acids and pancreatic enzymes in the anomalous choledocho-pancreatico ductal junction, J. Pediatr. Gastroenterol. Nutr. 3 (5) (1984) 721–727.
[11] T. Miyano, A. Yamataka, Y. Kato, O. Segawa, G. Lane, S. Takamizawa, S. Kohno, et al., Hepaticoenterostomy after excision of choledochal cyst in children: a 30-year experience with 180 cases, J. Pediatr. Surg. 31 (10) (1996) 1417–1421.
[12] P.B. Manning, T.Z. Polley, K.T. Oldham, Choledochocele: an unusual form of choledochal cyst, Pediatr. Surg. Int. 5 (1990) 22–26.
[13] S. Suita, K. Shono, Y. Kinugasa, M. Kubota, S. Matsuo, Influence of age on the presentation and outcome of choledochal cyst, J. Pediatr. Surg. 34 (12) (1999) 1765–1768.
[14] S.H. Kim, J.H. Lim, H.K. Yoon, B.K. Han, S.K. Lee, Y.I. Kim, Choledochal cyst: comparison of MR and conventional cholangiography, Clin. Radiol. 55 (5) (2000) 378–383.
[15] S.S. Saluja, P.K. Mishra, B.C. Sharma, P. Narang, Management of choledochal cyst with portal hypertension, Singapore Med. J. 52 (12) (2011) 239–243.
[16] L.W. Martin, G.A. Rowe, Portal hypertension secondary to choledochal cyst, Am. J. Surg. 190 (1979) 638–639.
[17] D.H. Park, M.H. Kim, S.K. Lee, S.S. Lee, J.S. Choi, Y.S. Lee, D.W. Seo, H.J. Won, M.Y. Kim, Can MRCP replace the diagnostic role of ERCP for patients with choledochal cysts? Gastrointest. Endosc. 62 (3) (2005) 360–366.
[18] V.Y. Sacher, J.S. Davis, D. Sleeman, J. Casillas, Role of magnetic resonance cholangiopancreatography in diagnosing choledochal cysts: case series and review, World J. Radiol. 5 (8) (2013) 304–312.
[19] K.W. You, S.W. Park, G.S. Lee, J.K. Kim, H.C. Moon, G.Y. Hong, A case of enteritis cystica profunda in the ampulla of Vater Mimicking Choledochocele, Clin. Endosc. 46 (2) (2013) 178–181.
[20] K.M. Ziegler, H.A. Pitt, N.J. Zyromski, A. Chauhan, S. Sherman, D. Moffatt, G. Lehman, J.L. Grosfeld, Choledochoceles: are they choledochal cysts, Ann. Surg. 252 (2010) 683–690.
[21] R. Law, M. Topazian, Diagnosis and treatment of choledochoceles, Clin. Gastroenterol. Hepatol. 12 (2014) 196–203.
[22] R. Masetti, A. Antinori, R. Coppola, C. Coco, C. Mattana, A. Crucitti, A. La Greca, G. Fadda, P. Magistrelli, A. Picciocchi, Choledochocele: changing trends in diagnosis and management, Surg. Today 26 (1996) 281–285.
[23] E. Ray-Offor, A.B. Allison, P.O. Igwe, Recent trend in management of adhesive small bowel obstruction, P.H. Med. J. 7 (2013) 239–242.