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A giant choledochal cyst: A case reported from Tanzania

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ABSTRACT

INTRODUCTION: Choledochal cyst is a rare common bile duct malformation that usually presents in the pediatric group with abdominal pain, distension, and jaundice. The pathophysiology remains unclear for the cause, and surgery aims to restore biliary enteric drainage.
CASE PRESENTATION: We present a six-year-old female who presented with gradual abdominal distention associated with jaundice. Abdominal ultrasound was suggestive of choledochal cyst, and CT-scan confirmed the diagnosis. She was operated on successfully and fared well.
CONCLUSION: Choledochal cysts are a rare entity of common bile duct malformations and should be considered as a differential diagnosis in the pediatric age group. Diagnosis can be easily made by non-invasive and in-expensive radiologic modalities like ultrasonography in resource-limited settings.

1. Background

Choledochal cysts (CC) were initially reported by Vader and Ezler in 1723 and later classified by Todani et al. in 1977 [1]. These are rare congenital cystic dilatations of the biliary tract, mostly developing in children under ten years and are four times more common in females [2,3]. Its pathogenesis is unclear, although, in 1969, Babbit first described that an abnormal pancreaticobiliary junction (APBDJ) outside the Ampulla of Vater allows mixing of the pancreatic and biliary juices activating the pancreatic enzymes. The active enzyme causes inflammation and deterioration of the biliary duct wall leading to dilatation and cyst formation. Further pressures in the pancreatic duct can further dilate the weak walled cyst [4,5].

The clinical presentation is variable depending on the age. Common presentations in pediatric patients include abdominal pain, jaundice, and right upper quadrant mass [6]. Cholangitis, pancreatitis, portal hypertension, and liver function test abnormalities are common complications and are thought to be a result of ABPDJ or stone obstruction [7].

CC is typically diagnosed using multimodality imaging, including ultrasound, computed tomography (CT), and magnetic resonance cholangiopancreatography (MRCP). Ultrasound is the most frequently used, given its low cost and accessibility, and has been shown to be reliable and cost effective as a single modality imaging in the pediatric population [8]. Here we report a CC presented in early childhood.

This work has been reported in line with the SCARE 2020 criteria [9].

2. Case presentation

A six-year-old-female, second born in the family, presented to a referral hospital in Tanzania, with abdominal distension for one month, progressively worsening, associated with pain and intermittent fever. She also presented with yellow discolouration of the eyes for the duration. One week prior to admission, the patient was admitted at a primary health facility and treated for malaria and severe anaemia, where she was transfused and received artesunate 50 mg intravenously. Ascitic tapping was done due to massive ascites; amount and colour not specified. She was referred to us with a preliminary diagnosis of a liver cyst.

Abbreviations: APBDJ, abnormal pancreaticobiliary junction; aPTT, partial thromboplastin time; ALT, alanine transaminase; AST, aspartate transaminase; CC, choledochal cyst; CT, computed tomography; ERCP, endoscopic retrograde cholangiopancreatography; INR, international normalised ratio; MIS, minimally invasive surgery; MRCP, magnetic resonance cholangiopancreatography; MRI, magnetic resonance imaging; PT, prothrombin time; PTC, percutaneous transhepatic cholangiography.

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On her admission, she was clinically ill-looking, moderately pale, with a tinge of jaundice, wasted with a weight for height of -2 standard deviation, had mild lower limb oedema, but did not have any peripheral lymph nodes palpable. Her vitals were within normal range. Physical examination revealed a grossly distended abdomen (Fig. 1) with positive shifting dullness and tender hepatomegaly of approximately 10 cm below the costal margin. Abdominal ultrasound revealed a huge cystic lesion with irregular margins, with some separations and unmeasurable internal echoes around the right upper quadrant. Abdominal CT-scan showed a giant type IA CC measuring approximately 10.2 cm (AP) × 6.7 cm (T) × 14.0 cm (CC) arising from the entire extrahepatic duct. There was mild biliary obstruction secondary to the compression effect of the giant cyst, and gross ascites (Fig. 2).

Other lab investigations revealed a lymphocytosis of 19.87 × 10^9/l with predominant neutrophils (63%), haemoglobin of 7 g/dl, raised liver enzymes, AST-88.49 mmol/l, ALT-45.09 mmol/l, raised total and conjugated bilirubin of 29.51 mmol/l and 28.06 mmol/l respectively, unaltered bleeding functions (INR-1.00, PT-12.5 s, Aptt-30.5 s). Serum albumin was ordered but not performed due to lack of reagents at the hospital. The child was scheduled for an elective surgery, performed by a pediatric surgeon at the centre. The abdomen was opened through a right extended subcostal incision, and a huge type 1A CC was found (Fig. 3). Two litres of amber colored ascites with no sediments was encountered and drained. The periportal veins were not dilated and no splenomegaly seen. The gall bladder was mobilized, the cyst was dissected from the confluence of the hepatic ducts and down towards the duodenum and was submitted for histopathologic analysis (Fig. 4). The distal end was closed, jejunum mobilized, and end-to-side hepaticojejunostomy was done in a Roux-en-Y manner. She was transfused pre and

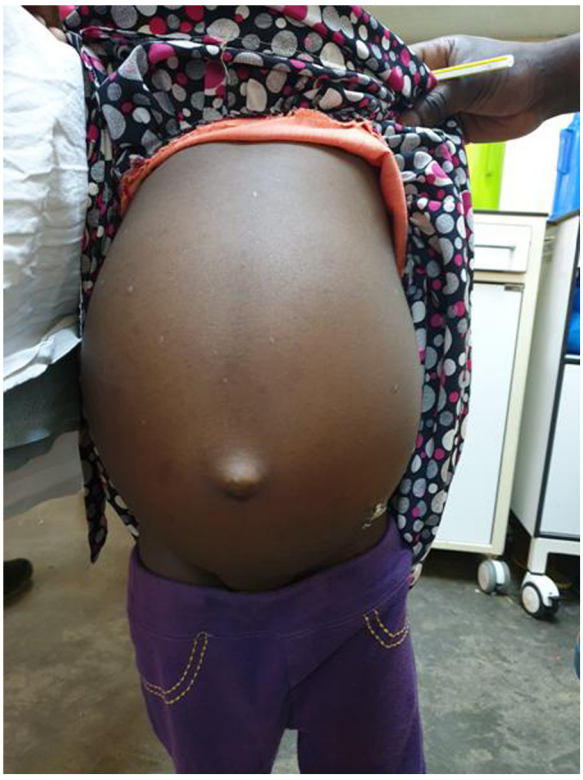

Fig. 1. Grossly distended abdomen due to ascites.

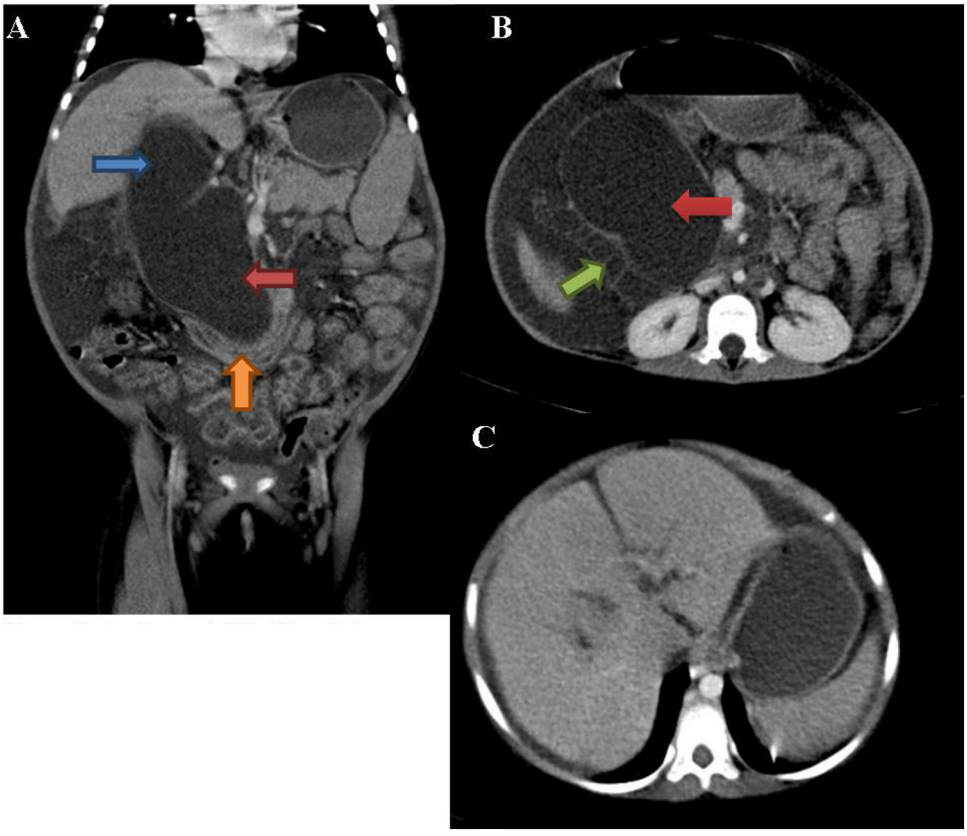

Fig. 2. A: Coronal CT of the abdomen shows cystic dilatation of the extrahepatic duct in keeping with type 1a giant choledochal cyst. Blue arrows – common hepatic duct. Red arrow – common bile duct. Orange arrow – 2nd portion of the duodenum. B: Axial CT of the abdomen shows dilated cystic duct running alongside the common hepatic duct. Green arrow – cystic duct. Red arrow – common hepatic duct. C: Axial CT of the abdomen shows mildly dilated intrahepatic biliary radicles.

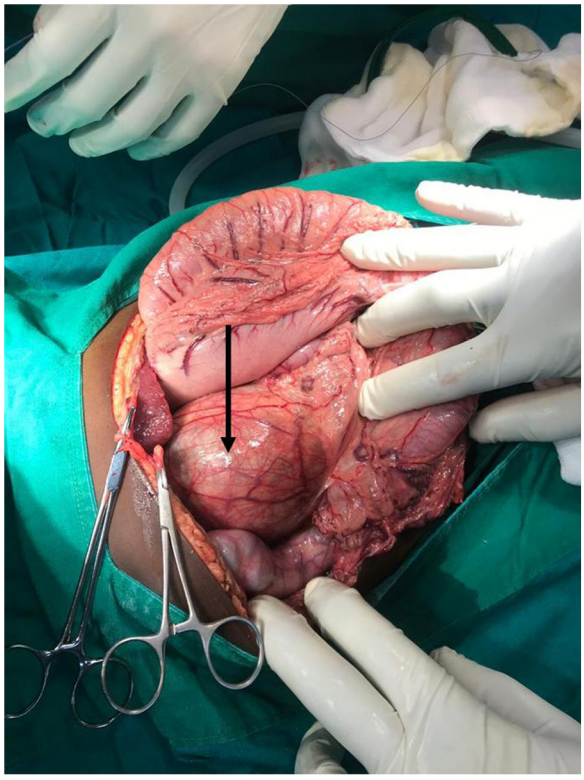

Fig. 3. Choledochal cyst (Arrow).

post operatively, raising the haemoglobin to 13.1 g/dl. Serum bilirubin dropped drastically to 18.12 mmol/l, the drain was removed on the fifth-day post-surgery, and the patient was discharged on the tenth-day post-surgery without any complaints.

Histopathology report for the specimen microscopically showed a cystic lesion with a thick fibromuscular wall. The wall showed embedded benign ducts in clusters. The luminal side was infiltrated by inflammatory cells, including foamy macrophages with

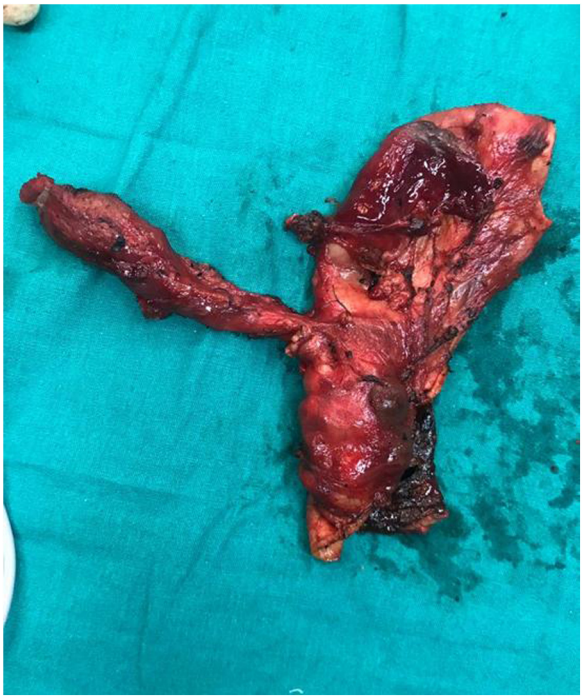

Fig. 4. Dissected cyst from common hepatic duct towards the duodenum.

bile pigment. There was extensive submucosal haemorrhage and lining epithelial attenuation (Fig. 5).

3. Discussion

Only 20% of patients with CC present with the classic triad of abdominal pain, jaundice and a palpable mass, predominantly in the pediatric population [2], where jaundice mainly occurs in type I and IV CC [10,11]. Rare findings associated with CC include cholangitis, pancreatitis, portal hypertension, liver function abnormalities and coagulopathy [8,12]. It was evident in the index case that the child had a history of abdominal pain and presented with jaundice

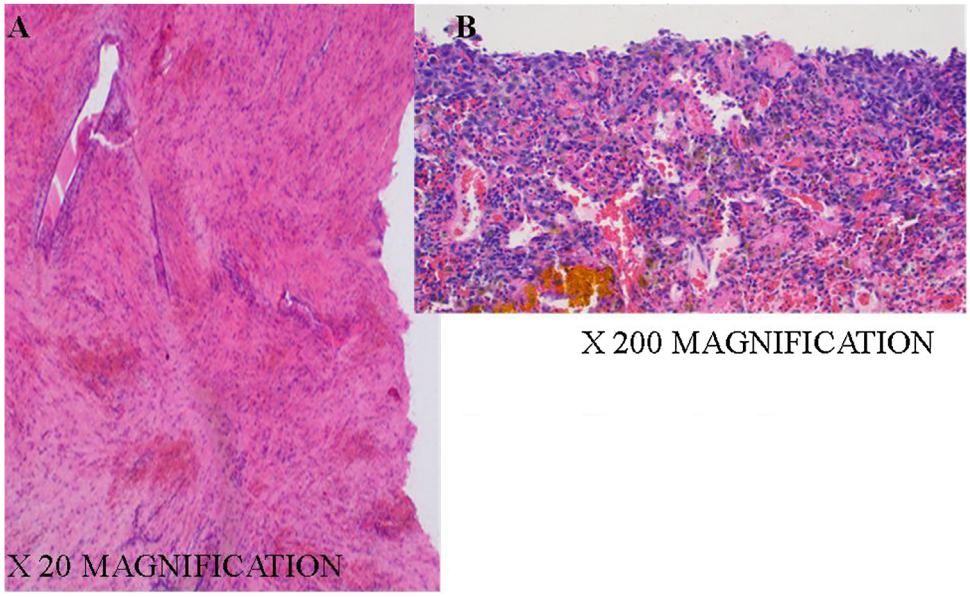

Fig. 5. A: Fibromuscular wall. B: H&E stained sections show a cystic lesion with thick fibromuscular wall. The wall showed embedded benign clusters of ducts. The luminal side of the cystic lined by attenuated glandular epithelium. The submucosal layer is infiltrated by mixed inflammatory cells accompanied by hemorrhage and foamy bile pigmented histocytes.

and a palpable mass. In a study by Soares et al. in 2014, Type 1 CCs were predominant in the pediatric age group [8]. According to literature, CCs remain relatively uncommon, and less documented in a lot of parts of the world including Africa, Western Europe and the United States. However, in Asia, incidence reaches up to 1 in 1000 [13].

The diagnosis of CC is commonly first made using abdominal ultrasound. Other Imaging techniques such as CT, magnetic resonance imaging (MRI), and endoscopic retrograde cholangiopancreatography (ERCP) are often used to define the anatomy of the biliary tree [14]. MRCP is considered as the best option in preoperative diagnosis to define the biliary tract, not only due to its high specificity (90–100%) and sensitivity (70–100%), but it also has the advantage of effortlessly delineating the pancreatic and biliary ducts and therefore aiding in the classification of the cyst according to Todani's classification [15,16].

Percutaneous transhepatic cholangiography (PTC) or ERCP are highly sensitive but are utilized less frequently given their invasiveness and associated risks including cholangitis, bleeding, pancreatitis and perforation [17]. In our case, we used abdominal ultrasound and CT-scan to identify the origin of the abdominal mass and to describe the mass in terms of size. MRCP was not used due to financial constraints. CC can effectively be identified via CT-scan without the need for further imaging modalities, especially in the pediatric age group and aided us on the characteristics of the cyst in relation to other abdominal organs.

The classification of CC was first laid out by Alanso-Lej, Revor and Pessagno in 1959 [12]. It was later modified by Todani et al. describing five main types [18]. Type I involves the dilatation of the entire common hepatic or common bile duct or segments of each, and it is the most commonly encountered (80–90% of all CC). Type I CC can be further sub-classified into Ia, which is a cystic dilatation of the common bile duct as in the presented case. Ib is a focal segmental dilatation of the common bile duct and Ic is represented by a fusiform dilatation of both common hepatic and common bile duct. Type II is a diverticular dilatation from the common bile duct. Type III is a choledochocele, located within the duodenal wall at the pancreaticobiliary junction. Type IV CC has multiple cysts which can involve both the intrahepatic and extrahepatic biliary tree. Type IV CC can be further subdivided into Type IVa and IVb cysts depending on intrahepatic involvement. Type IVb refers to multiple extrahepatic biliary cysts without intrahepatic involvement. Type V CC, or Caroli disease, appears as an intrahepatic cystic dilatation without evidence of extrahepatic dilatation [14]. Type I CC, along with type IV cysts, have the highest risk of malignancy [12].

Surgical management via cyst excision is the definitive treatment for CC. It aims to fully excise the cyst and restore biliary enteric drainage into the duodenum or via Roux-en-Y hepaticojejunostomy [19]. Laparoscopic approach is currently the gold standard of minimally invasive surgery (MIS) technique in managing CC in children [20]. It was first reported in 1995 by Farello [21], and has been used since for type I and type IVa choledochal cyst excision. The comparison studies show leak rates, morbidity and mortality in laparoscopic approach are comparable with the open technique [22], with a metaanalysis by Shen et al. in 2015 reporting an added advantage of less blood loss, better cosmesis and a shorter hospital stay [23], although it is technically demanding [20]. This approach was not used in our case due to less expertise in laparoscopic surgery and the rare nature of the disease. Other MIS techniques including robotic surgery and single incision laparoscopic surgery have been reported recently, with no clear clinical benefit of both as compared to conventional laparoscopic technique [20].

Although a few cases of CC have been reported in Egypt [24] and South Africa [25], there is a paucity of data from Africa. We were not able to find any cases from East Africa in our search through pub med and google scholar.

4. Conclusion

Choledochal cyst is an uncommon pathology, and the management is surgical. However, the technique for biliary tract reconstruction post-removal of the cyst is not clearly established. Since there are no prospective randomized clinical studies, judgment and experience of the surgeon will determine the technique for the biliary tract reconstruction. On the other hand, common bile duct malformations should be in the differential lists, especially in the pediatric age group who present with jaundice, abdominal pain and a palpable mass. Ultrasonography studies are cheap and readily available hence aid in making the diagnosis in resource-limited settings. Higher-resolution modalities can be used to further describe the pathology in relation to other abdominal structures. Laparoscopic excision of CC and hepaticojejunostomy remains safe and feasible in children, and should be used in well equipped centres with greater experience and expertise in laparoscopic surgery. Although it consumes more time, the surgical outcome appears comparable to open procedure.

Declaration of Competing Interest

The authors report no declarations of interest.

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Ethical approval

Ethical approval was obtained from the department of General surgery, KCMC Hospital.

Consent

Written informed consent was obtained from the patient 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.

Author contribution

- Jamil Suleiman – Reviewed medical records and writing of the script.
- Jay Lodhia – conceptualization and writing of the script.
- David Msuya – performed the surgery and reviewed medical records.
- Adnan Sadiq – radiology analysis and reporting.
- Pactrick Amsi – histology analysis and reporting.
- Rune Philemon – reviewed the records and conceptualizing of script.

Registration of research studies

Not applicable.

Guarantor

N/A.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Availability of data and materials

All data used in this study are available from the corresponding author upon request.

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