A misdiagnosis of choledochal cyst type IB and recommendation to use gold-standard imaging techniques: A case report

Arsalan Hyder,[1] Nitik Sharma,[2] Farhan Ali,[3] Hina Sadiq,[4] Kashif Ali,[5] Hittik Kumar[6]

Abstract

A choledochal cyst (CC) is a rare congenital dilation of the biliary ductal system that can cause troublesome complications when left untreated. CC in children classically manifests as a rare triad of the right upper quadrant mass, jaundice, and abdominal pain. Here, we report the case of an eight-year-old boy seen in Paediatric Unit I of Dr Ruth K. M. Pfau Civil Hospital Karachi, Pakistan, on October 9, 2019, who was initially misdiagnosed for liver abscess due to the non-specific symptoms and managed appropriately with antibiotics, which failed to improve the symptoms. Moreover, a hydatid cyst was considered based on ultrasonography (USG) which was later ruled out by the negative serology. A definitive diagnosis of CC was established on magnetic resonance cholangiopancreatography (MRCP). In resource-limited countries, USG is always the first line screening tool for biliary duct abnormalities which in some cases is comprehensibly enough for the diagnosis of a rare entity like CC, thus requiring a highly specific imaging test like MRCP to be performed to devise an effective treatment and surgical plan.

Keywords: Choledochal Cyst, Pediatrics, Diagnostic Errors.

DOI: https://doi.org/10.47391/JPMA.4855

Introduction

A choledochal cyst (CC) is an infrequent congenital cystic dilatation of the intrahepatic and or extrahepatic biliary ductal system that can cause biliary obstruction and cirrhosis. Its occurrence is common in children but is detected in about one-fourth of adults with female predominance of 3-4:1.[1] The occurrence in the Western population is about 1 in 100,000-150,000 live births while it is much higher in Asian population with about 1/1,000, with Japan contributing two-thirds of the cases among Asians alone.[1] A widely accepted modified version of Alonso-Lej et al classification was put forward by Todani et al in 1977 which classifies these cysts into five different types.[2] Of these, type I and IV are the common forms.[1] Type 1 CC is characterised by dilatation of the extrahepatic biliary tree only and it is further divided into three subtypes: type IA, type IB, and type IC. Type IB, which was seen in this case, is the focal dilatation of a segment in the common bile duct excluding its proximal portion. CC is seen in about 80% of the cases during infancy and presents with a rare clinical triad of the right upper quadrant mass, jaundice, and abdominal pain.[3] We report a rare case of CC type IB that presented with eccentric features and was misdiagnosed for other cystic hepatic lesions because of the lack of technical quality of the examination. In the end, the patient was successfully managed and was referred for surgical intervention.

Case Report

An eight-year-old boy, presented to the Paediatric Unit I of Dr Ruth K. M. Pfau Civil Hospital Karachi, Pakistan, on October 9, 2019, with one-week history of abdominal pain and three-day history of vomiting and fever. The patient was unremarkable for jaundice, melena, or history of weight loss. On physical examination, there were no scar marks, palpable liver, or tenderness in the right hypochondrium on deep palpation. All other systemic examinations were unremarkable. Laboratory findings revealed increased levels of serum glutamic pyruvic transaminase (SGPT), alkaline phosphatase (ALP), C-reactive protein (CRP), and total bilirubin (T Bil), with decreased level of haemoglobin (Hb) and mean corpuscular volume (MCV) (Table-1), while other laboratory values were within normal limits.

Given the patient's initial presentation, age and examination, an ultrasound was performed, which revealed an enlarged liver of 12cm and a hypoechoic mass of 5.5×5.9cm in the right lobe of the liver. The findings were consistent with right liver abscess. Given this diagnosis, opinion was taken from paediatric surgery, and it was advised that for this size no surgery was needed, so ultrasound-guided aspiration and antibiotics can be given. The patient was given 0.9% dextrose and

[1,2,6]3rd Year MBBS Student, Dow Medical College, Dow University of Health Sciences, Karachi, [3]Department of Internal Medicine, [4,5]Department of Paediatric Medicine, Dr. Ruth K.M Pfau Civil Hospital, Dow University of Health Sciences, Karachi, Pakistan.

Correspondence: Arsalan Hyder. Email: arsalanhyder1211@gmail.com

Table: Laboratory investigations of the patient.

Parameters	Patient's results	Reference Range
Hb	11.5	13-17 g/dL
MCV	72.3	80-100 fL
TLC	11.4	4.5-11.0 × 109/L
Neutrophils	73.5	40%-75%
Lymphocytes	11.2	20%-45%
Platelets	395	150-450 103 /?L
CRP	105.7	<3 mg/dL
T-Bil	2.55	0.3-1.2 mg/dl
SGPT	110	7 to 56 U/L
ALP	622	40-112 U/L
PT	11.4	9-12 s
APTT	19.2	30-45 s
INR	1.09	0.7-1.2
Albumin	3.7	3.5-5 g/dL

[Hb, haemoglobin; MCV, mean corpuscular volume; TLC, total leukocyte count; CRP, C reactive protein; T-Bil, total bilirubin; SGPT, serum glutamic-pyruvic transaminase; ALP, alkaline phosphatase; PT, prothrombin time; APTT, activated partial thromboplastin time; INR, international normalised ratio].

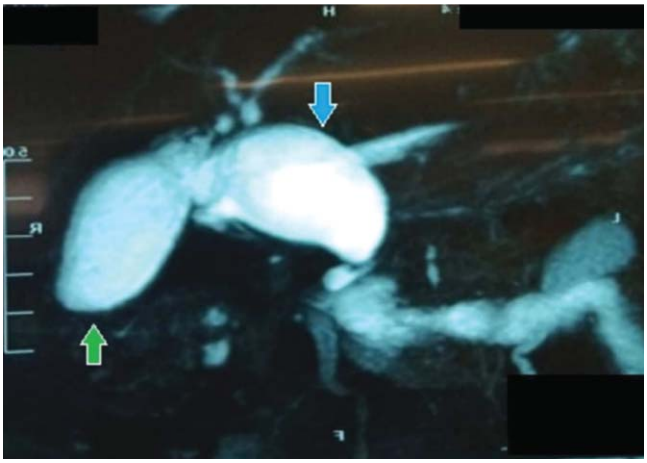

Figure: MRCP of the patient showing fusiform dilation of the CBD (blue arrow), eliciting a choledochal cyst and a normal gall bladder (green arrow).
MRCP: Magnetic resonance cholangiopancreatography; CBD: Common bile duct.

was started on a triple antibiotic regime consisting of injection Ceftriaxone (75mg/kg/OD), injection Metronidazole (10mg/kg/8 hourly) and injection Vancomycin (20mg/kg/8 hourly) but the patient complained of continuing pain. Repeat ultrasound showed persistent mass, suggestive of a hydatid cyst, for which echinococcus antibodies results came negative. Furthermore, computed tomography (CT) with contrast was performed which showed enlarged liver of 12.8cm with no mass. However, it showed dilation of proximal extrahepatic biliary duct, measuring 3cm and length of 4.5cm with no dilation of the intrahepatic biliary duct. A definitive diagnosis of CC type IB was established on magnetic resonance cholangiopancreatography (MRCP) (Figure-1). An informed verbal consent was acquired from the parents of the patient to publish this case in online literature.

Discussion

CC is a rare congenital abnormality in the biliary tree which is characterised by cystic dilation of bile duct. The aetiology of CC is equitably understood. As suggested by Wang et al,[4] 93.8% of the patients with the anomalous pancreaticobiliary ductal union (APBDU) were associated with CC. APBDU is an anomaly that causes union between the pancreatic duct and common bile duct (CBD) to join outside the duodenal wall; forming a long common channel that results in reflux of pancreatic contents into the CBD; thus, promoting cystic changes of the bile duct. Edil et al in their study revealed that the clinical features of CC in children differ from that of adults with jaundice being the most common feature in the paediatric group;[5] however, our patient presented with no sign of jaundice, which is unusual for the paediatric population, leading to a misdiagnosis of liver abscess. Ultrasonography (USG) was suggested, because of the coinciding symptoms of our patient with liver abscess and its space-occupying lesion nature. USG is the typical obstetric scanning test in the low-and-middle-income countries (LMIC),[6] such as Pakistan due to its cost-effectiveness and effortless technical handling. This may lead to- misdiagnosis for versatile complications and rare diseases of the biliary system as well as incomplete management of the patient. As no surgical intervention was needed for a 5.5×5.9cm mass for liver abscess, and the patient was started on an antibiotic regime. However, pain was not relieved by IV antibiotics, pointing towards another cause of complaints in our patient. Repeat USG showed a cyst of 5cm in the right lobe of the liver, suggestive of a hydatid cyst due to its overlapping clinical symptoms and high prevalence. Central Asia contributes millions of cases for echinococcus and is considered endemic for such infections. A review article from Khan et al[7] showed that Pakistan alone contributed 1,702 cases of echinococcus in the previous decades. It was denied of a hydatid cyst because of the absence of echinococcus immunoglobulins, leading to another misdiagnosis. A CT scan with contrast showed a focal dilatation of proximal extrahepatic biliary duct with a maximum diameter measuring 3.0cm and a length of 4.5cm, along with lab investigations that showed increased levels of certain specific markers such as ALP and SGPT (Table). The findings were suggestive of CC type IB, the diagnosis of which was confirmed after MRCP (Figure) which revealed dilatation of CBD with no evidence of intrahepatic duct dilatation. MRCP is the most sensitive and specific screening test for anomalies of the Biliary tree such as CC;

it not only helps physicians to correctly diagnose such infrequent diseases but also detects related complications as well as formulates an effective surgical plan.[8] So, it should be incorporated in a diagnostic algorithm in suspected CC cases. Because of the rarity of this disease and false interpretation of images by the examiner, USG can contribute to an increased risk of diagnostic errors in identifying various structures of liver and bile ducts.[9] Due to the high prevalence of hydatid cysts in Pakistan,[7] especially in Karachi, a rare case like CC can be misdiagnosed for cystic echinococcosis on routine USG, suggesting the need for another sensitive testing such as CT scan, MRI, and MRCP. The diagnosis of CC in our case was missed because the technical quality of the examination may not have allowed for recognising the anatomic pathology. Prenatal testing has proven to be compelling for diagnosis of CC so it should be considered in order to avoid further complications.[10] Despite the diagnostic dilemma, the patient was successfully diagnosed and managed, and, thereby sent for further surgical follow-up.

Conclusion

CC is a rare congenital anomaly of the hepatobiliary system. After being unable to correctly diagnose our patient's condition with USG and concomitantly having no response to antibiotics, we approached more reliable tests such as CT and MRCP that revealed a dilatation in the CBD. This shows that the use of definitive imaging modalities formulates the correct plan and prevents further delay in surgical intervention.

Disclaimer: None to declare.

Conflict of Interest: None to declare.

Funding Disclosure: None to declare.

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