

A Case of Cystic Biliary Atresia: Imaging Findings and Surgical Outcome, Makkah, Kingdom of Saudi Arabia

Ebtehal Althobaiti^{1*}, Ahmed Abouissa², Thanaa Ewis³, Tantawi Muhammad⁴

¹Department of Radiology, Alnoor Specialist Hospital, Makkah, Saudi Arabia

²Department of Interventional Radiology, Alnoor Specialist Hospital, Makkah, Saudi Arabia

³Department of Radiology, Maternity and Children Hospital, Makkah, Saudi Arabia

⁴Pediatric Surgery Department, Maternity and Children Hospital, Makkah, Saudi Arabia

*Corresponding author: Ebtehal Althobaiti, Department of Radiology, Alnoor Specialist Hospital, Makkah, Saudi Arabia, Tel: 966568159625; E-mail: ebtehal.althobaiti@outlook.com

Received date: April 29, 2021; Accepted date: September 6, 2021; Published date: September 16, 2021

Citation: Ebtehal A (2021) A Case of Cystic Biliary Atresia: Imaging Findings and Surgical Outcome, Makkah, Kingdom of Saudi Arabia. J Clin Radiol Case Rep Vol.5 No.4.

Abstract

Cystic biliary atresia (CBA) is a relatively uncommon cystic variant of type 1 Kasi classification of biliary atresia. It usually presented with cholestatic jaundice, elevated liver function tests, and ultrasound finding of a cyst at the hepatic hilum, which mimics the features of a choledochal cyst (CC). We present the first reported case of CBA in Saudi Arabia with the imaging findings and surgical outcome. A 53-day-old infant admitted to the neonatal intensive care unit (NICU) due to persistent jaundice since birth. Imaging findings and surgical exploration confirm the diagnosis of CBA. An excellent prognosis of CBA with early surgical intervention necessitates the radiologist to be familiar with this entity and differentiate it from CC.

Keywords: Cystic Biliary Atresia; Choledochal Cyst, Intraoperative Cholangiography, Kasi Portoenterostomy.

Abbreviations: CBA= Cystic Biliary Atresia, CC= Choledochal Cyst, MRCP= Magnetic Resonance Cholangiopancreatography, US= Ultrasound.

Introduction

Biliary atresia (BA) is defined as obstruction of all or part of the extra-hepatic bile ducts. The overall incidence is low, worldwide, it affects an estimated 1 in 8,000-18,000 live births (1). In Saudi Arabia, the data about BA is limited (2), (3). A retrospective study has been conducted in Riyadh by Holdar et al. between 2008 – 2015 and revealed that BA accounts for only 4.7% of all infantile cholestasis cases (2). Cystic biliary atresia (CBA) is a relatively uncommon cystic variant of type 1 Kasi classification of BA, which accounts for less than 10% (4).

It usually presented with a picture of cholestasis and typical ultrasound (US) finding of a cyst at the hepatic hilum, which also seen in an infant with choledochal cyst (CC) (1). Therefore, it is essential to differentiate between the two entities as evident

improved the surgical outcome of CBA with early intervention, unlike CC, where appropriate time selection is needed (5). We reported a case of CBA with the imaging findings and surgical outcome, which is the first described in Saudi Arabia.

Case Presentation

A 53-day-old full-term infant, admitted to the neonatal intensive care unit (NICU) prior two weeks due to persistent jaundice, pale stool, and dark urine since birth. Family history is irrelevant. Laboratory tests were performed and revealed high alkaline phosphatase 544 IU/L, high total and direct bilirubin 129, 102 µmol/L, respectively.

Ultrasound obtained (not shown) and revealed cystic lesion at the porta hepatis measured about 14 x 8 mm. Average gallbladder (GB) length with a prominent cystic duct measured 2 mm. The liver was enlarged without dilatation of the intrahepatic biliary radicles. The common bile duct (CBD) was not visualized. With careful searching, echogenic cord sign couldn't be seen.

After the inconclusive result of the Ultrasound, MRI was done to verify the communication of the cystic lesion with the intra- or extra-hepatic biliary radicles. MRI (Fig.1) revealed high T2 signal intensity hepatic hilar cyst. Magnetic resonance cholangiopancreatography (MRCP) with a 3D reformat (Fig.2) showed a prominent cystic duct that communicates with a 1.5-cm cyst at the porta hepatis. Both intra- and extra-hepatic biliary radicles are non-visualized.

These findings raise the possibility of cystic biliary atresia over the choledochal cyst as they both can have a hepatic hilar cyst. Still, they differ in communication with the extra-hepatic biliary radicles. So, to confirm the diagnosis, we had two options: either to go for Phenobarbital-enhanced hepatobiliary scintigraphy or cholangiography. Based on the infant age and after discussion with the primary team, it was decided to do percutaneous transhepatic cholangiography under general anesthesia in the operative theatre, and further decisions will be taken accordingly.

Under US-guidance, the cyst was accessed by a 22 gauge-needle, the catheter was placed. Aspiration revealed clear yellow fluid. During the injection of 2-3 ml of diluted (50%) iodinated contrast material, an anteroposterior fluoroscopic image was obtained (Fig.3), which confirmed the MRI findings. The contrast delineated the cyst and its communication with cystic duct and GB. The GB appeared small. Intra- and extra-hepatic biliary ducts were not opacified. Contrast started to leak into the peritoneal cavity, confirming the diagnosis of cystic biliary atresia (type 2 French classification).

Based on these findings, Surgeon decided to proceed for Kasai portoenterostomy procedure at the same session to prevent further delay in the management (Fig.4). One week later, bilirubin levels declined, and the stool became dark.

Discussion

Cystic biliary atresia (CBA) is an uncommon variant of type 1 biliary atresia but has a relatively favorable prognosis with early intervention. It is defined as a cystic dilatation at the porta hepatis accompanied with atresia of the common bile duct. On the other hand, a choledochal cyst is a condition of multiple degrees of abnormal cystic dilatation of the biliary tree (4). Both usually presented with persistent jaundice, clay-colored stool, elevated liver function tests, and ultrasound finding of a cyst at the hepatic hilum yet, they differ in the management approach and prognostic outcome (6).

In a retrospective study done by Wang X et al., they found the mean levels of total and direct bilirubin were 188 ± 34 , 147 ± 29 $\mu\text{mol/L}$, respectively, which almost applicable to our result. It considered being significantly higher in CBA compared to CC (6).

Imaging characteristic findings were investigated in prior studies. According to Lee SM et al. 46 patients were surgically confirmed cases of BA, a general US features that showed significant association with BA was abnormal gallbladder morphology (either not visualized or have an atretic lumen, length equal or less than 15 mm), triangular cord sign with a cut-off 3.4 mm resulted in a sensitivity of 78.2%, a specificity of 100%, and non-visualization of the CBD (7). Regarding the comparison between CBA and CC: Smaller cystic diameter, which stated differently in the literature, but with a maximum 2.5 cm favor CBA, our case was within this limit. Triangular cord sign was detected in CBA with variable sensitivities have been reported (range, 23–93%) but none in CC, this sign was not visualized in our patient which could be due to thin fibrous ductal remnant in the porta hepatis as suggested by Lee et al. Dilatation of intrahepatic bile ducts was seen in CC but none in CBA (7), (8). Recently, MRCP has been used as a non-invasive method before any intervention (6). It was helpful in our case as the US findings were not typical as mentioned previously.

All the previously mentioned methods to differentiate between CBA and CC are listed in the literature. However, the definitive diagnosis would be given by intraoperative cholangiography with a typical finding of a small gallbladder connected to a “non- communicating” cyst (9).

Type 1 BA, including its variant CBA, showed excellent outcomes comparing to the other types. Regardless of the followed surgical approach, the literature reported good results with early intervention of less than 70 days of life. Both hepaticojejunostomy and Kasi portoenterostomy have been reported with a satisfactory long-term outcome (4), (6), (10), (11). Kelay A et al. Published in 2017 that a sufficient restoration of bile flow and resolution of jaundice was seen in 40 – 45% infant with BA. However, potential morbidity due to cirrhosis, recurrent cholangitis, and portal hypertension is considerable. Therefore, life-long monitoring is required (12).

Conclusion

Excellent prognosis of CBA with an early surgical intervention promotes us as a radiologist to raise the suspicion of this entity whenever phasing an infant with cholestasis and US finding of a cystic lesion at the hepatic hilum and to be able to differentiate it from choledochal cyst.

Reference

1. Schooler GR, Mavis A (2018) Cystic biliary atresia: A distinct clinical entity that may mimic choledochal cyst. *Radiol Case Rep* 13: 415-418.
2. Holdar S, Alsalem B, Asery A, Al-Hussaini A (2019) Outcome of biliary atresia among Saudi children: A tertiary care center experience. *Saudi J Gastroenterol* 25: 176-80.
3. Kamal J (2012) Biliary Atresia: A Report from the King Abdulaziz University Hospital in Jeddah, Kingdom of Saudi Arabia. *Journal of King Abdulaziz University-Medical Sciences*.
4. Tang J, Zhang D, Liu W, Zeng JX, Yu JK, Gao Y, et al. (2018) Differentiation between cystic biliary atresia and choledochal cyst: A retrospective analysis. *Journal of paediatrics and child health* 54: 383-9.
5. Wang X, Qin Q, Lou Y (2014) A Retrospective Study Between Type I Cystic Biliary Atresia and Infantile Choledochal Cyst at a Tertiary Centre. *Hong Kong Journal of Paediatrics* 19: 175-80.
6. Lee SM, Cheon J-E, Choi YH, Kim WS, Cho H-H, Kim I-O, et al. (2015) Ultrasonographic diagnosis of biliary atresia based on a decision-making tree model. *Korean journal of radiology* 16: 1364-72.
7. Zhou L, Guan B, Li L, Xu Z, Dai C, Wang W, et al. (2012) Objective differential characteristics of cystic biliary atresia and choledochal cysts in neonates and young infants: sonographic findings. *Journal of ultrasound in medicine : official journal of the American Institute of Ultrasound in Medicine* 31: 833-841.
8. Parra D, Fecteau A, Daneman A (2016) Findings in percutaneous cholangiography in two cases of Type III cystic biliary atresia (with ultrasound correlation). *BJR | case reports* 1: 20150377.

9. Takahashi Y, Matsuura T, Saeki I, Zaizen Y, Taguchi T, et al. (2009) Excellent long-term outcome of hepaticojejunostomy for biliary atresia with a hilar cyst. *J Pediatr Surg.* 44: 2312-5.
10. Caponcelli E, Knisely AS, Davenport M (2008) Cystic biliary atresia: an etiologic and prognostic subgroup. *J Pediatr Surg* 43: 1619-24.
11. Kelay A, Davenport M (2017) Long-term outlook in biliary atresia. *Seminars in pediatric surgery*, Elsevier.