

CASE REPORT

A Rare Case of Cystic Biliary Atresia

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ABSTRACT

Biliary atresia is one of the commonest causes of prolonged jaundice in neonate, whereas cystic biliary atresia is a rare form of biliary atresia with similar presentation, but a slightly different sonographic finding. Being able to differentiate them radiologically is important as the surgical management is different for these two diagnoses. An ultrasound examination was done for a 2-month-old baby girl with prolonged jaundice with the finding of a cyst along the portal triad, which raised the suspicion for cystic biliary atresia. The diagnosis was confirmed with intraoperative cholangiography (IOC) and Kasai procedure was done. The patient was well after the surgery with resolution of jaundice.

Malaysian Journal of Medicine and Health Sciences (2023) 19(3):380-382. doi:10.47836/mjmhs18.5.48

Keywords: Cystic biliary atresia, Choledochal cyst, Biliary atresia, Prolonged jaundice

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INTRODUCTION

Biliary atresia is a congenital disorder of the biliary tract characterized by progressive fibrosis of the biliary tree which eventually led to obstruction. It is often diagnosed with ultrasound before proceeding with IOC. Finding of a cystic lesion along the biliary tract may complicate the radiological diagnosis further by adding the possibility of choledochal cyst as a differential diagnosis (1) which will change the surgical management

CASE REPORT

A 2-month-old, term, baby girl was referred for prolonged jaundice. Antenatally her mother had a history of mild anemia in pregnancy, otherwise was uneventful. Other than that, she also had a history of passing pale colored stool. She was fully breastfed on demand since birth. Neither the patient nor her mother had a history of taking traditional medicine. Her blood investigations showed obstructive jaundice features. Other prolonged jaundice workup was unremarkable. She was referred for ultrasound abdomen scan to rule out biliary atresia.

Ultrasound examination demonstrated the typical findings of biliary atresia, including presence of a small atretic gallbladder (length of 18mm) despite adequate fasting with irregular gallbladder wall and thickened

anterior fibrotic cord (Fig. 1). The ultrasound also found an anechoic cystic lesion along the portal triad which did not show doppler signal (Fig. 2). The common bile duct was otherwise not well appreciated. No intrahepatic biliary dilatation or other cystic lesion seen elsewhere along the biliary tree.

The patient was arranged for operation by the pediatric surgical team. Intraoperatively, the gallbladder was found to be atretic (Fig. 3). There was cystic dilatation of the common bile duct which extended distally to pancreas. IOC was performed by injecting contrast into the atretic gallbladder, showing no flow of contrast into the intrahepatic system nor the small bowel (Fig.3). Injection of contrast into the cyst also showed no communication with the biliary tree or small bowel. Dissection of the common bile duct distal to the cyst showed no lumen. The whole portahepatic tract was sent for histopathological examination. Liver biopsy was performed over segment V during the same setting. The diagnosis of cystic biliary atresia was made during the surgery and. Subsequently, they proceeded with the Kasai procedure.

Post operatively, the patient was monitored in the ward. She recovered well with resolution of her jaundice. Serial blood investigations in the ward showed improvement of her transaminitis as well as bilirubin level. She was discharged well.

Histopathological examination of the common bile duct and the cystic duct show obliteration of the lumen by fibrosis. No patent lumen or epithelial lining noted.

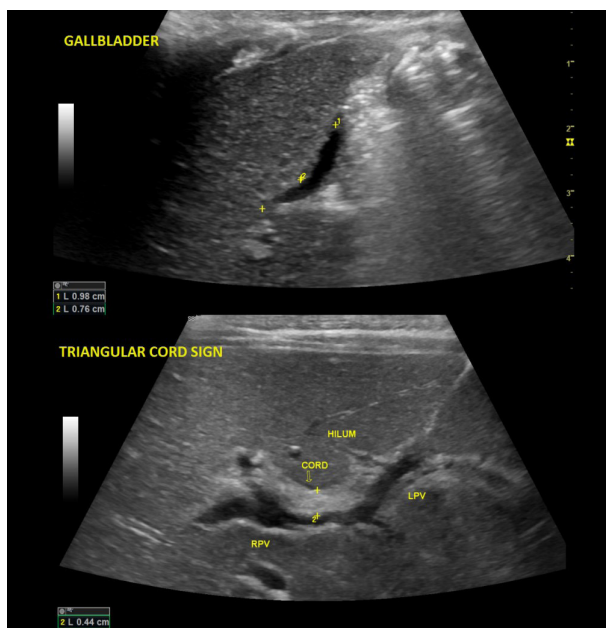


Figure 1: Ultrasound image on the top shows small gallbladder with irregular contour and loss of normal echogenic mucosa. Ultrasound images on the bottom shows thickened echogenic fibrous cord anterior to the right portal vein in keeping with a positive triangular cord sign.

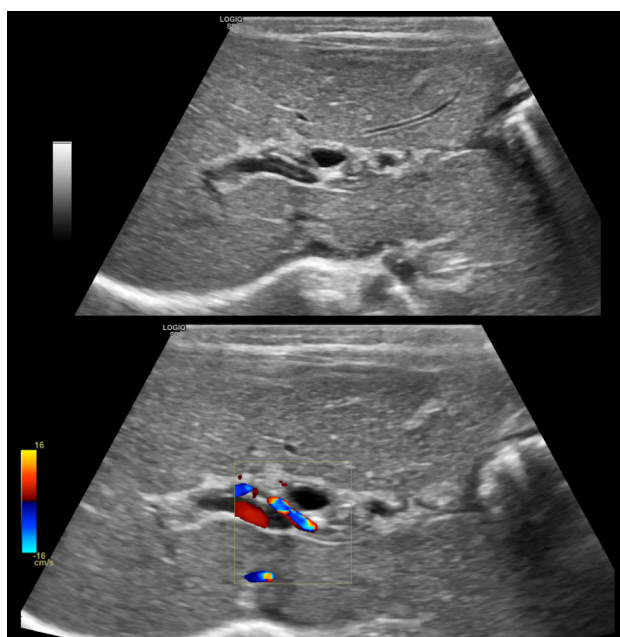


Figure 2: Ultrasound images showing the cystic structure at the portahepatis region with no color flow within.

The common hepatic duct shows very narrow lumina with the underlying stroma densely infiltrated by chronic inflammatory cells. The sections of the liver shows fibrosis and expanded portal tracts with portal-to-portal bridging forming hepatic nodules. Ductular proliferation resembling bile-duct plate is noted with associated bile plugging. There was mild-moderate portal lymphoplasmal infiltration admixed with some neutrophils. Panacinar cholestasis noted with the hepatocytes exhibiting ballooning, giant cell changes and multinucleation. Overall histopathological findings are consistent with biliary atresia with cirrhotic changes of the liver (Fig. 4).

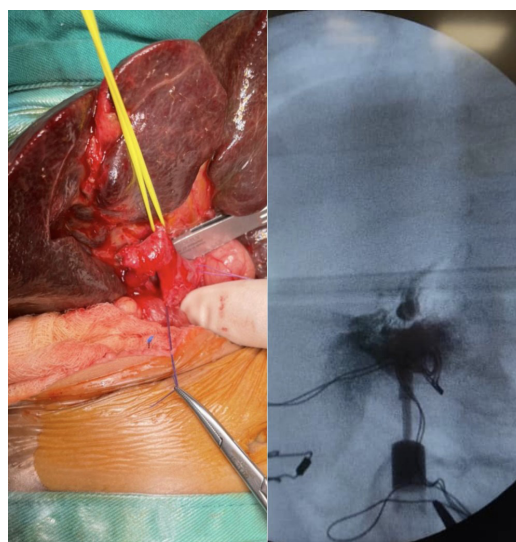


Figure 3: Image on the left showing intraoperative finding of a small atretic gallbladder, while image on the right showing Intraoperative Cholangiography (IOC) demonstrating no contrast flowing into the intrahepatic ducts proximally or into the common bile duct and small bowel distally.

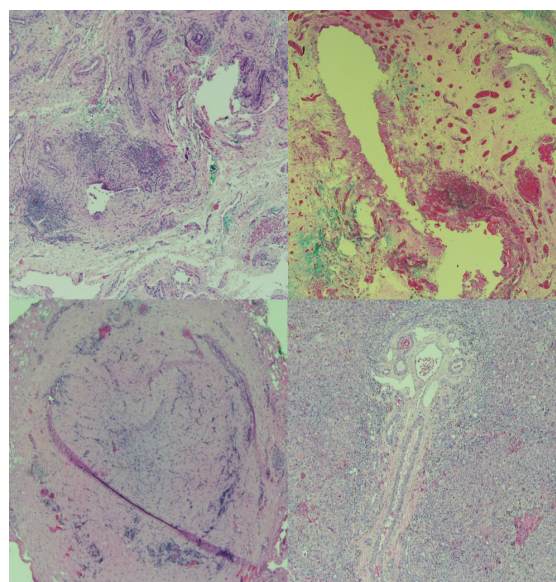


Figure 4: Images with 4x magnification of the common hepatic ducts (top left), common bile duct (top right) and cystic duct (bottom left) showing obliteration of the lumen by fibrosis. The underlying stroma of the common hepatic ducts is densely infiltrated by chronic inflammatory cells. 10x magnification images of the liver (bottom left) shows expanded and fibrotic portal tracts with hepatic nodules. There was evidence of ductular proliferation with associated bile plugging. Interface hepatitis as well as lobular inflammation with spotty necrosis are also seen. There is panacinar cholestasis with the hepatocytes exhibiting ballooning, giant cell changes and multinucleation.

DISCUSSION

Biliary atresia is a congenital disorder of the biliary tree. If left undiagnosed and untreated, the child may progress to liver cirrhosis. Cystic biliary atresia being a rare variant of biliary atresia, also manifests with similar presentation of prolonged jaundice with obstructive picture. While

the incidence of biliary atresia ranges from 1:5000 birth to 1:20,000 birth worldwide, only approximately 8% of them have the cystic variant (2).

Ultrasound is usually the first line investigation and is usually adequate to make the diagnosis and justify for operation. The typical findings of biliary atresia in ultrasound include an atretic gallbladder with irregular contour and the presence of triangular cord sign, which is an echogenic fibrous biliary duct remnant best seen anterior to the right portal vein. Mu SL et al. also suggested that hepatic artery diameter may be increased in the case of biliary atresia.

In this case, the patient had the typical findings of biliary atresia. However, this case was complicated further with the finding of an anechoic cystic structure along the porta hepatis. With the presence of cyst, choledochal cyst would be another diagnostic dilemma. The surgical management would slightly be different as biliary atresia will be managed with Kasai procedure, while choledochal cyst is managed by excision and hepaticoenterostomy.

However, the ultrasound diagnosis of cystic biliary atresia against choledochal cyst is made possible by understanding the pathophysiology, in which in the case of choledochal cyst, the obstructive jaundice is mainly caused by the compression of an otherwise patent biliary duct by the cyst, whereas for cystic biliary atresia, there should not be dilated duct as the biliary tree are fibrosed and obliterated (1). Therefore, in this case, the ultrasound diagnosis was cystic biliary atresia instead of choledochal cyst as there was no dilated biliary ducts. In fact, the common bile duct was hardly appreciable in the ultrasound.

The diagnosis was further confirmed intraoperatively by using IOC. In the case of cystic biliary atresia, injection of contrast into the cystic dilatation will not demonstrate any communication with the biliary tree, nor the small bowel loops. Whereas for choledochal cyst, injection of contrast into the cyst will demonstrate opacification of the biliary tree as well as flow of contrast into the intestinal lumen (1).

Other non-invasive imaging techniques that can be used to differentiate cystic biliary atresia and choledochal cyst include magnetic resonance cholangiopancreatography (MRCP) and hepatic scintigraphy (HIDA) scan (1). However, in comparison to ultrasound, both MRCP and HIDA are not as widely available, time consuming, and lack specificity as there has been cases reported as choledochal cyst on MRCP but was found to be cystic biliary atresia in HIDA (3).

Cystic biliary atresia is mainly a radiological and surgical diagnosis as the decision for surgery is based on the

radiological and intraoperative findings. Nevertheless, cystic biliary atresia and choledochal cyst are two distinct histopathological diagnoses. The histopathological findings of the biliary tract and cyst in the case of cystic biliary atresia typically lack of epithelium and inflammation with associated myofibroblastic changes, whereas choledochal cyst usually demonstrate normal epithelium with no subendothelial fibrosis (4). In our case, the cystic dilatation of the common bile duct was sent together with the rest of the extrahepatic biliary tract for histopathological examination and was found to have no epithelial lining with fibrotic and obliterated lumen, which were consistent with the finding of biliary atresia rather than choledochal cyst.

Ductular proliferation, bile plugging, hepatocytes ballooning with giant cell changes and portal lymphoplasmal infiltration are the possible histological changes of the liver in biliary atresia. Among these changes, ductular proliferation is considered highly sensitive and specific for biliary atresia (5), which is present in our case.

CONCLUSION

Cystic biliary atresia is a rare variant of biliary atresia presented as prolonged jaundice in neonates. The diagnosis of cystic biliary atresia should always be considered when a cystic lesion was detected sonographically without a patent biliary duct, especially in the presence of an atretic gallbladder and anterior cord sign.

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