Cystic biliary atresia: An unforeseen experience

AH Abu Bakar[1] PK Bal[2] MF Madzlan[2] JE Gilbert Fernandez[2] N Mohd Badaruddin[2] MT Md Nor[3]

[1] Paediatric Surgery, Universiti Kebangsaan Malaysia Medical Centre (UKMMC)

[2] Paediatric Surgery, Hospital Raja Permaisuri Bainun, Ipoh

[3] Paediatric Surgery, Hospital Raja Perempuan Zainab II, Kota Bharu

Introduction

Cystic biliary atresia (CBA) is rare but it is an important entity. There are few radiological features which are suggestive of CBA. This report represents a case on diagnosis of CBA.

Case Summary

Clinical history

A term baby girl whose antenatal history was unremarkable was workup in the primary care for prolonged jaundice. Workup was normal and ultrasound was not done at this stage as the stool was pigmented. In view of progressive hyperbilirubinemia and stool subsequently become pale, she was referred to us at day 69 of life. During our examination, abdomen was distended with hepatomegaly.

Investigation

Liver function test showed hyperbilirubinemia with prominent direct bilirubin. Liver enzymes were all raised. Ultrasound revealed small gallbladder measuring 0.3 x 0.4x 1.8cm with presence of triangular cord sign and cystic lesion size 0.4 x 0.6cm at porta hepatis

Surgery

Surgery was done at day 78 of life and revealed coarse appearance of the liver with neovascularization and atretic gallbladder. No ascites was noted. On table cholangiogram (OTC) showed cystic appearance with no proximal and distal flow of contrast (figure 1) and intra-operatively noted cystic dilatation of common bile duct measuring 3cm extending distally to the pancreas with obliterated lumen (figure 2). Surgery was proceeded with Kasai portoenterostomy.

Outcome

Post operatively she recovered well and was discharged at post op day 7 with post Kasai protocol. During follow up, her stool was pigmented and bilirubin is coming down. Liver biopsy showed ductular proliferation with bile plug. Common bile duct and cystic duct show obliteration of lumen by fibrosis and mildly infiltrated by lymphocytes.

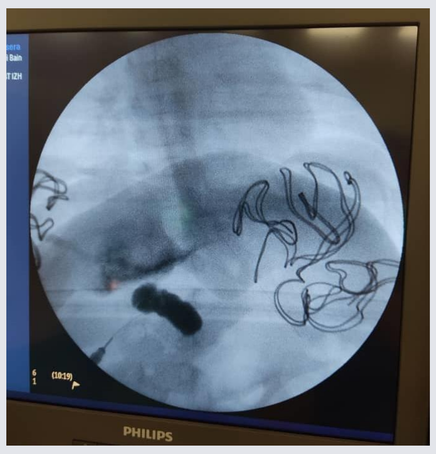

Figure 1. OTC showed cystic appearance with no contrast flow proximally and distally.

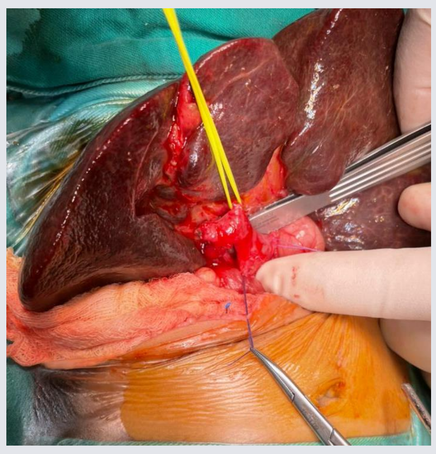

Figure 2. Intra-op finding revealed cystic dilatation of common bile duct.

Discussion

Neonate with prolong jaundice can be due to many causes either medical or surgical causes. Pale colour stool is the most worrying symptom. It is a significant symptom in surgical causes which warrants urgent intervention. Cystic BA will have the same clinical presentation of biliary atresia (BA).

A thorough work up should be done to define the cause for jaundice. This works up should include biochemical and radiological investigations. Ultrasound of hepatobiliary system is adequate in diagnosing BA. Triangular cord sign is the diagnostic feature for BA. In cases of cystic BA, additional feature in ultrasound is cystic lesion at porta hepatis.

Prognosis in cystic variant of biliary atresia appears to correlate with age at surgery, which is similar to classical biliary atresia; thus, earlier surgical intervention results in an improved outcome. CBA should be considered in neonate who suspected to have choledochal cyst.

Conclusion

Cystic BA is a rare subtype of BA. The clinical and laboratory investigations are similar to the classical BA. Presence of triangular cord sign with cystic lesion should raise suspicion of cystic BA. This would assist the operating surgeon in the preparation for surgery.

References

1. Gary et al. 2018. Cystic biliary atresia: A distinct clinical entity that may mimic choledochal cyst. Radiology Case Reports.
2. Jue Tang et al. 2017. Differentiation between cystic biliary atresia and choledochal cyst: A retrospective analysis. Journal of Paediatrics & Child Health.
3. Koshinaga et al. 2018 Obliterative cholangiopathy in acquired cystic biliary atresia type III after cyst perforation: a case report. BMC Paediatrics
4. Ankur et el. 2012. Cystic Biliary atresia: Confounding and intriguing. Journal of Paediatrics.