CASE REPORT

CHOLEDOCHAL CYST AND BILIARY STONE

Anjum Saeed, Asaad Assiri*

Department of Paediatrics, Division of Gastroenterology, Hepatology & Nutrition, The Children's Hospital & Institute of Child Health, Lahore-Pakistan, *Department of Paediatrics, College of Medicine, King Saud University Medical City, Riyadh-Saudi Arabia

Choledochal cyst is not an uncommonly seen entity in children. Presentation may be varied and at different age groups. Abnormal pancreaticobiliary junction is usually a phenomenon in acquired type of choledochal cyst. A four years old child presented with symptoms of acute viral hepatitis A but persistence of symptoms beyond the usual time necessitated other conditions to be considered. Choledochal cyst was the final diagnosis complicating with biliary stone which was managed by surgical excision.

Keywords: Choledochal cyst; Biliary stones; Children.

Citation: Saeed A, Assiri A. Choledochal cyst and biliary stone. J Ayub Med Coll Abbottabad 2018;30(1):127–9.

INTRODUCTION

Choledochal cyst's (CC's) are cystic dilatation of the biliary system and may be congenital or acquired. Traditionally CC's are classified into five types depending upon the position of the cyst.[1] Pathogenesis is mainly unknown but few theories and mechanisms are proposed for the development of CC's. Acquired type has strong association with abnormal pancreaticobiliary junction (APBJ) which is approximately present in 70% of these cases.[2,3] CC's may remain asymptomatic or may present with biliary cirrhosis. Chronic or intermittent abdominal pain is the most common presentation in children and adolescent. CC's may present with complication like recurrent jaundice, cholangitis, biliary stones, pancreatitis, perforation of common bile duct and malignant transformation of the cyst.[4] Super added infection with hepatotropic viruses may have serious outcome in these children. Normal or mildly elevated liver enzymes with raised gamma glutamyl transferase (GGT) and alkaline phosphatase may be the first clue towards diagnosis of CC's. Ultrasound abdomen, Computed tomography (CT), Magnetic resonance cholangiopancreatography (MRCP), endoscopic retrograde cholangiopancreatography (ERCP) and Endoscopic ultrasound are helpful modalities in diagnosing and classification of CC's. Management is surgical excision of the cyst with biliary diversion and has good prognosis if intervened earlier.[5]

CASE REPORT

Four years old girl was referred to Paediatric Gastroenterology unit for admission in January 2015 because of right upper quadrant pain and vomiting from the emergency department. She was developmentally normal child with incomplete immunization. Three days prior to admission, her abdominal pain worsened and started intractable vomiting which was non-bilious and without blood. She was running moderate to high grade intermittent fever with these symptoms. There was no history of jaundice, pale stool, dark urine, deranged conscious level or bleeding from anywhere. She had a contact with jaundiced child 4 weeks prior to presentation. On physical examination, she was found dehydrated without any evidence of jaundice, pallor, clubbing and skin rashes or bleed. Abdominal examination revealed clear tenderness in the right hypochondrium with palpable liver 4 cm below the right costal margin (total span of 9 cm), soft in consistency and smooth surface. There was no evidence of splenomegaly and ascites. Other system examination was unremarkable.

Investigations done at the time of admission showed total bilirubin of 25 micmol/L, direct fraction of 22 micmol/L, ALT 218, AST 199, GGT 244, Alkaline Phosphatase 546 (U/L) and Albumin 31 g/L. CBC, amylase, lipase, BUN & electrolytes, urine analysis were all normal. Viral screening for Hepatitis A, B, CMV and EBV was sent and awaited. Ultrasound abdomen done in the emergency room showed mild hepatomegaly with normal echo-texture.

She was hydrated with intravenous fluids, started with antibiotics and her fever subsided in three days. The other symptoms (vomiting and abdominal pain) which improved initially reoccurred after three days with frequency and intensity both were less. Follow up of LFT's at one week was almost same but with normal bilirubin and significant elevation of GGT and alkaline phosphatase to 944 and 589 U/L respectively. Her hepatitis screening turned out to be positive for Hepatitis-A IgM and negative for HbsAg, CMV and EBV. So, she was labelled as acute viral hepatitis A and managed with the supportive therapy.

After one week, she continued to have intermittent abdominal pain with off and on vomiting

and mild tenderness in the right hypochondrial region, however, after two weeks there was an elevation of ALT 322, AST 201, GGT 933, Alkaline phosphatase of 364 (U/L). A repeat ultrasound abdomen showed dilated common bile duct (CBD) measuring 0.93cm without any intrahepatic dilatation and suggested MRCP for further details. MRCP was reported as dilatation of CBD throughout its course with mild dilatation of right and left hepatic ducts, there was an oval shaped filling defect in the distal part of CBD with an additional finding of union of pancreatic duct with CBD before opening in the second part of duodenum which suggest choledochal cyst with an abnormal pancreaticobiliary junction as shown in MRCP figure 1. ERCP was done and confirmed MRCP findings without any stone but minimal amount of sludge was removed. She developed asymptomatic pancreatitis post ERCP which was managed conservatively. She was discharged home and seen at follow up, 6 weeks' post ERCP, her LFT's and pancreatic enzyme were normal however GGT and Alkaline phosphatase stayed high (823, 477 U/L) respectively.

She was readmitted for laparoscopic exploration; a full excision of dilated cyst with cholecystectomy was performed. CBD was significantly dilated with mild dilatation of right and left hepatic dilatation as well. Biliary diversion was made in the form of hepatico-duodenostomy. She was discharged home with normal LFT's and GGT. Histopathology findings were, gall bladder without any significant pathology, normal biliary epithelium without any dysplastic change in the bile duct specimen.

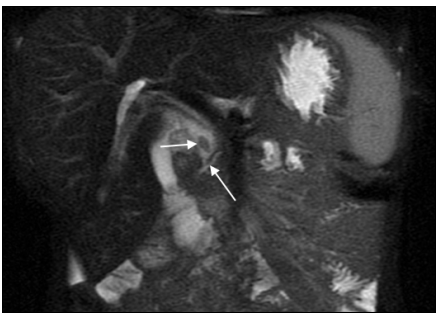

Figure-1: MRCP showing dilated CBD s with a stone in the distal part (upper arrow), pancreatic duct joining CBD before opening into small bowel APBJ (lower arrow).

DISCUSSION

Choledochal cyst or biliary cysts may be single or in multiples involving both extra and intrahepatic ducts according to the type of cyst.[2,6] The incidence is around 1:100,000 but in some Asian countries it is as high as 1:1000.[7] CC's may be isolated or with some other anomalies like biliary atresia, multi-septate gall bladder, duodenal and colonic atresia. Abnormal pancreaticobiliary junction is associated with 70% of the biliary cysts as similar to our case.[8]

The presentation is variable and depends upon the age, majority of them present in childhood and only 25% are detected in adult life. Infants present with cholestasis with or without acholic stool. Children and adolescent have varied presentation with recurrent jaundice or cholangitis, intermittent abdominal pain, pancreatitis and perforation of bile duct. Triad of abdominal mass, pain and jaundice is present in less than 20% of children. Complications of CC's include strictures in the biliary tree, stone formation due to stasis of bile in the cystic part of bile duct, biliary cirrhosis and portal hypertension if not managed at appropriate time, perforation of bile duct and malignant changes in the cyst tissue.[8,9]

Co-infection with hepatotropic viruses can be devastating in any established or chronic liver disease, although we were unable to find any documented case reports about co-infection of CC's with acute viral hepatitis. It is possible that co-infection like hepatitis A may unmask the underlying liver or biliary disease as in our patient. There are case reports about co-infection of hepatitis B and C with acute hepatitis A or E leading to decompensation of the existing silent liver disease.[10]

LFT's may remain normal or mildly elevated with an element of obstruction. Ultrasound is first modality of choice if performed by an expert sonologist (70–90% of detection) as it was missed on the first ultrasound in our patient. CT has advantage of detecting both extrahepatic as well as intrahepatic cysts and extent of biliary system involvement. MRCP is non-invasive and very sensitive for all types biliary cysts along with detection of APBJ if present without the risk of cholangitis and pancreatitis as associated with direct cholangiography and ERCP.[11–13] For difficult cases ERCP is always a great help especially in type I and IV where distal extent should always be seen before surgery.[14] Endoscopic ultrasound can be helpful in detection of CC's and APBJ but not a common procedure in children.

Management of these children is complete excision of the cyst with biliary diversion. Complete excision of cyst will prevent malignant transformation as had been reported with the previous mode of treatment in the form of cysenterostomy or external drainage of cyst. Roux-en-Y hepatico-enterostomy is the procedure of choice in these children either in the form of heptico-jejunostomy or

hepaticoduodenostomy.[15] Risk of malignancy after surgery dropped to 0.7–6% from 26% if left untreated or cyst tissue is left there.[17] Histopathology of this cyst tissue may be normal or densely fibrotic with an element of acute and chronic infection.

Surgical conditions like choledochal cyst should be considered within the context of hepatocellular elevation with or without cholestasis, as surgical excision early on can decrease the incidence of drastic complication of malignancy in these cysts.

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Received: 29 October, 2016	*Revised: 18 February, 2017*	*Accepted: 27 February, 2017*

Address for Correspondence:

Anjum Saeed, Department of Paediatrics, Division of Gastroenterology, Hepatology & Nutrition, The Children's Hospital & Institute of Child Health, Lahore-Pakistan.
Email: anjuj2002@gmail.com