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Choledochal cyst associated with extrahepatic bile duct atresia

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ABSTRACT

Three infants who presented with clinical features of neonatal cholestasis and clinically suspected to have biliary atresia were found on imaging studies and surgery to have extrahepatic bile duct atresia in association with choledochal cyst. All patients were treated by bilio-enteric bypass procedure. Post operatively jaundice cleared in only two patients. The prognosis of this association depends on the pre-existent liver damage due to biliary atresia.

KEY WORDS: Biliary atresia, Choledochal cyst

INTRODUCTION

Choledochal cyst is localized dilatation of the extrahepatic and/or intrahepatic biliary tree while biliary atresia is a type of obstructive cholangiopathy in which there is obliteration/sclerosis of the extrahepatic bile ducts. Choledochal cyst is a rare congenital anomaly with a reported incidence of 1 in 100,000-150,000 live births.[1] The incidence of biliary atresia is about 1 in 10,000-12,000 live births.[2] Hence, choledochal cyst and extrahepatic biliary atresia are individually rare congenital malformations and their association in a single case is still rarer. Very few cases have been reported in the literature.[3,4]

MATERIALS AND METHODS

We report three infants, aged 2, 2.5 and 3 months, with association of choledochal cyst and extrahepatic biliary atresia who were treated at our institute. The clinical presentation was similar to that of biliary atresia in all the three infants – clay colored stool, high colored urine and icterus since birth. Imaging evaluation was done with ultrasonography (USG), magnetic resonance cholangiopancreatography (MRCP), 99mTc iminodiacetic acid hepatic scan (HIDA) and peroperative cholangiography (POC). Biopsy of the liver was taken in all the three cases.

RESULTS

Dilatation of the extrahepatic bile ducts was evident on USG and MRCP preoperatively and HIDA scan did not show gut activity in all the three cases (Figure 1, 2). POC

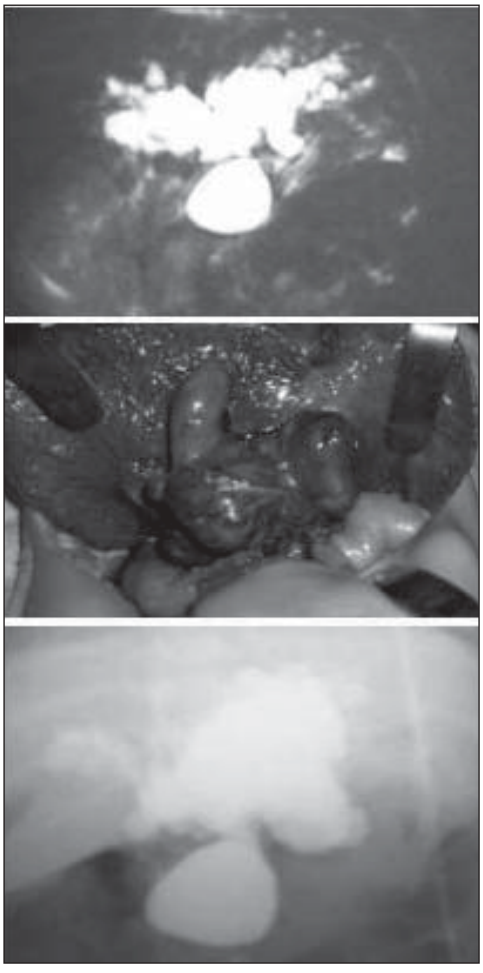

Figure 1: Pre-operative MRCP showing choledochal cyst; operative picture showing atresia of common bile duct and operative cholangiogram showing the same findings as the MRCP with no passage of dye in the duodenum. Case 1

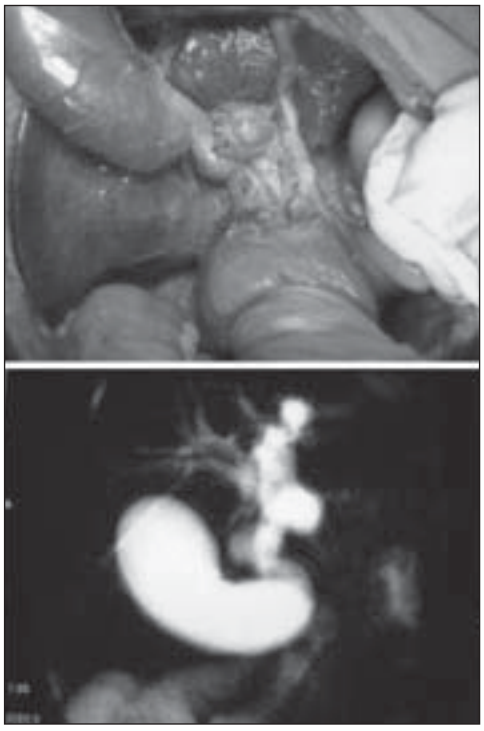

Figure 2: MRCP and operative picture showing atresia of the common bile duct. Case 3

failed to demonstrate patency of the distal common bile duct while liver biopsy showed classical features of biliary atresia in each case ie cholestasis, lobular disarray, bile duct proliferation, portal tract inflammation and extensive fibrosis bordering on cirrhosis. All the three infants were treated surgically with bilio-enteric bypass procedures. Three months post operatively jaundice cleared in only two cases.

Comments

Biliary atresia is a type of obstructive cholangiopathy in which there is obliteration/sclerosis of the extrahepatic ducts. Pathologically, the liver shows periportal inflammation and fibrosis, bile-duct proliferation, with variable absence of the extrahepatic bile ducts. Choledochal cyst is a localized dilatation of the common bile duct with obstruction to flow of bile into the duodenum and occasionally hepatocellular damage. The association of biliary atresia and choledochal cyst is extremely rare with clinical presentation mimicking that of biliary atresia. Diagnosis can be suspected on preoperative imaging and investigations. Liver histology is that of biliary atresia. Prognosis after surgical treatment is poor because of the pre-existing hepatic damage.

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