Biliary atresia associated with choledochal cyst

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ABSTRACT

Choledochal cyst and biliary atresia are rare but important causes of neonatal jaundice. Both present with jaundice and acholic stool in neonatal period. Treatment and prognosis of both entities are very different. We are presenting a case in which choledochal cyst co-existed with biliary atresia; the antenatal diagnosis was that of choledochal cyst. Patient had cyst excision with hepatojejnostomy, but ultimately required portoenterostomy because of associated biliary atresia.

We conclude that choledochal cyst and biliary atresia are two entities with similar antenatal and postnatal presentation; they should be differentiated as management and prognosis are different.

Key words: Biliary atresia, choledochal cyst, cystic biliary atresia

INTRODUCTION

Choledochal cyst is a rare congenital anomaly with reported incidence of 1 in 100000-150000, while the incidence of biliary atresia is 1 in 10000-15000 live birth.[1-6] Presence of both choledochal cyst and biliary atresia in a patient is extremely rare. Though both present with jaundice and acholic stool in neonatal period, their treatment and prognosis is different.

We are reporting a case of biliary atresia with cyst at portahepatis who was diagnosed as simple case of choledochal cyst, which ultimately required portoenterostomy.

CASE REPORT

A full term normal delivered female child who was antenatally diagnosed as case of choledochal cyst presented at the age of one and half months with jaundice since birth, and passage of clay coloured stool.

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General examination showed jaundice; abdominal examination revealed a lump in right hypochondrium, measuring 4x4 cm, with ill-defined margin. Laboratory examination showed total bilirubin 5.2 mmol/l (direct bilirubin was 4.3 mmol/l); liver enzymes were markedly elevated. Magnetic resonance imaging report was that of type I choledochal cyst with blind ending distal common bile duct [Figure 1]. Radioisotope liver scan showed non excretion of dye in the duodenum even after 24 hours.

Intra-operatively the cyst was located in the subhepatic region and gall bladder was found to be communicating with it [Figure 2]; aspiration of the cyst yielded no bile, only mucinous fluid was aspirated. Complete excision of cyst with hepaticojejunostomy was done. Liver biopsy was also taken, which later showed reversible hepatic injury. Post operatively patient continued to pass clay colour stool and there was no fall in serum bilirubin. Radioisotope liver scan showed non excretion of dye. Patient was reexplored and hepaticoductoscopy showed obliterated duct, which necessitated portoenterostomy. Postoperatively stercobilinogen in stool became positive. Histopathological examination showed ductile size of 200μ. Patient is presently doing well.

DISCUSSION

Choledochal cyst associated with biliary atresia is very rare disease; only 88 cases have been identified till now. Type I biliary atresia is the most common variety associated with choledochal cyst.[1]

Neonates with choledochal cyst present with jaundice and acholic stools characteristic of biliary obstruction.[2,4] This clinical presentation is indistinguishable from neonatal hepatitis and biliary atresia, and differentiation from these more common entities requires radiologic evaluation. The presenting symptoms and diagnostic findings of our patient were typical of those reported by other authors.[5]

Preoperative diagnosis can be made with help of ultrasonography, magnetic resonance cholangiopancreatography (MRCP) and radioisotope liver scan. Ultrasonographic criterion for the diagnosis of biliary

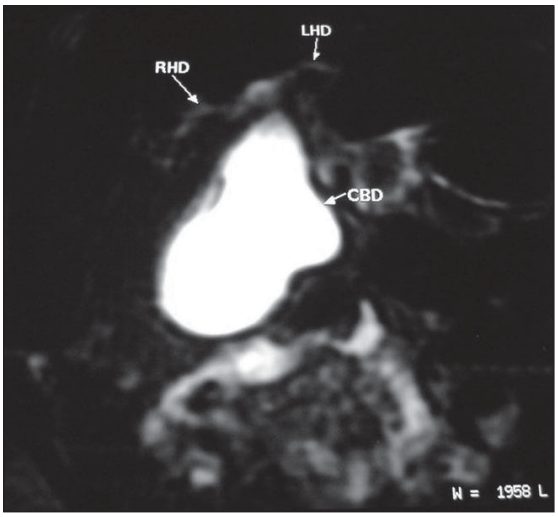


Figure 1: MRCP of the patient showing cystic lession in extrahepatic duct with well visalisation of both hepatic duct giving impression of patent duct

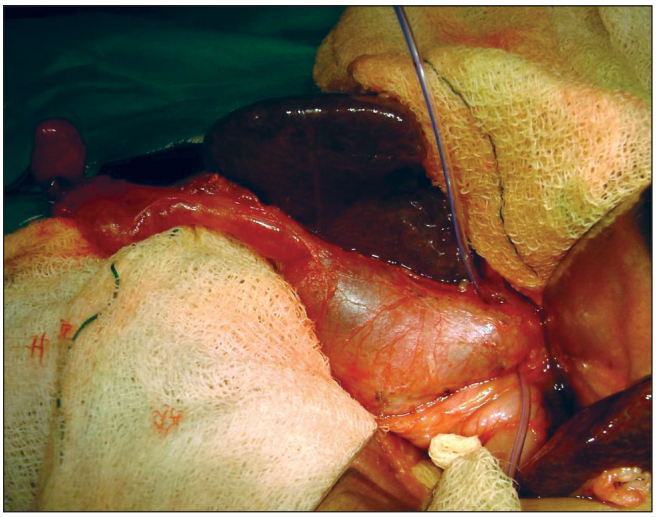


Figure 2: Operative photograph showing blind ending cytic lession resembling like choledochal cyst with non atretic gall bladder comunicating with the cyst

atresia is nonvisualisation of the gall bladder or the appearance of a small gall bladder less than 1.5 cm in length.[3] In our case, gall bladder was clearly visualised which is unusual for biliary atresia, but on radioisotope scan there was no excretion of dye in the duodenum and cyst, which indicate there was obliteration of duct proximal to cyst. On MRCP, intrahepatic duct are visualised and seen in continuity with cyst which is unusual for biliary atresia; this created false impression that it is case of choledochal cyst (MRCP overestimated patency of duct). Peroperatively, if there is no bile secretion from the proximal duct than duct patency should be checked. Hepaticoductoscopy can help in two ways first to ascertain patency of proximal duct second it helps in clearing sludge in proximal duct.

Although choledochal cyst with biliary atresia had been named as "correctable" type of biliary atresia in the past, it is known that there is no difference in treatment and prognosis between the correctable type and the "noncorrectable" type of biliary atresia.[5] The treatment of choice is complete surgical excision of the extrahepatic bile duct and Roux-en-Y portoenterostomy.[5]

Our case is unusual in that it is biliary atresia type III with a cystic structure that could be mistaken for correctable biliary atresia or choledochal cyst; even after resection of cyst with hepaticojejunostomy it does not show any excretion of dye. Similar finding was reported by Sookpotaram who reported two case of noncorrectable biliary atresia associated with cyst in portal region.[7]

In conclusion, cystic lesion of extra hepatic biliary apparatus should be carefully evaluated in infants because there is chances of associated biliary atresia as both can present similarly. MRCP and radioisotope liver scan, if done simultaneously, can help to differentiate between infantile form of choledochal cyst and cystic biliary atresia but confusion can occur with the use of MRCP which tend to overestimate the patency of duct. The definitive diagnosis can only be made peroperatively.

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