

Recurrent chemical pancreatitis complicating choledochal cystic disease

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Case report

A seventeen year old lady presented with a twelve year history of self-limiting attacks of severe epigastric pain, associated with occasional fever, normal total white cell count (WCC), normal or mildly elevated serum alanine transaminase (ALT) (13-200iu/l) and variable hyperamylasaemia (127-460iu/l). At age 5 years, abdominal ultrasonography had shown non-cystic dilatation of the intrahepatic and extrahepatic biliary system without choledocholithiasis. Despite endoscopic sphincterotomy of a normal ampulla with extraction of bile duct stones at age eleven years, followed by laparoscopic resection of a dilated gall bladder, the attacks increased in frequency. Subsequent endoscopic retrograde cholangio pancreatography (ERCP) confirmed biliary dilatation in the absence of stones but the pancreatic duct was poorly outlined. At age 15 years a magnetic resonance cholangiopancreatogram (MRCP) showed generalised dilatation of the extrahepatic biliary tree involving

the common hepatic duct and the porta hepatis with a large elongated choledochal cyst arising out of the lateral wall of the common bile duct (CBD). These findings are best seen in a coronal view (Figure 1) which also shows a normal pancreatic duct joining the CBD about 15mm proximal to the level of the ampulla. This suggests a long common pancreatobiliary channel running through the pancreatic head before entering the duodenal wall (Figure 2).

The patient underwent excision of the dilated biliary tract above the anomalous pancreatobiliary junction with Roux en Y hepatico-jejunostomy. A small cystic lesion in the jejunal wall was also excised. Histology of the resected specimens showed mildly inflamed choledochal malformations with no dysplasia. Postoperative recovery was uneventful. Follow up MRCP surveillance for neoplasia at six month intervals showed a patent hepatico-jejunostomy and an unchanging filling defect in the intapancreatic bile duct stump remnant with anterior meniscal indentation typical of a trapped air bubble (Figure 3).

Key words

Choledochal cysts, hyperamylasaemia, hepaticojejunostomy, pancreatitis



Figure 1: Coronal MRCP image showing gross dilation of intrahepatic and extrahepatic biliary tree (B), choledochal cyst (CC), pancreatic duct (PD), confluence of biliary and pancreatic ducts (C) at about 15 mm (double arrowed line) proximal to the duodenal wall (D).

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Table 1: Simplified Todani Classification of bile duct cysts

Type I	Extra hepatic duct segmental cylindrical or diffuse dilatation
Type II	Extra hepatic duct diverticulum cyst
Type III	“Choledochoceles” of the distal (intraduodenal) position of the CBD
Type IVA	Multiple cysts of intra hepatic + extra hepatic ducts
Type IVB	Multiple cysts of extrahepatic ducts
Type V	Single or multiple cysts of intrahepatic ducts

Discussion

Choledochal cysts are rare congenital malformations of the biliary tree, affecting mostly children and young adults that follow defective epithelialisation or recanalisation of the embryonic biliary plate. Biliary dilatation has been attributed to congenital weakness of the bile duct wall although hypertonicity of the sphincter of Oddi may coexist.¹ Choledochal cysts may present in infancy as obstructive jaundice that must be differentiated from biliary atresia, or intermittent cholangitis in childhood. The finding of gross extrahepatic bile duct dilatation that persisted in our patient despite sphincterotomy and stone removal was highly suggestive of a predominantly Type I

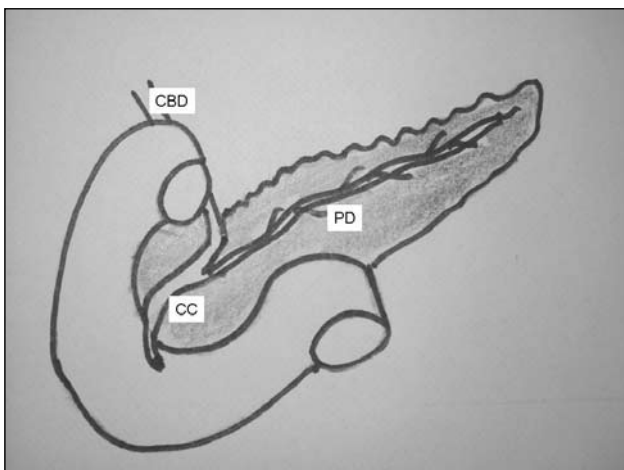


Figure 2: Anatomy of long pancreatobiliary common channel (CC) due to confluence of pancreatic duct (PD) and common bile duct (CBD) in pancreas at distance of more than 10 mm above ampulla of Vater.

choledochal cyst disease, the commonest type of choledochal cyst according to the Todani classification² (Table 1).

The patient's repeated attacks of hyperamylasemia and epigastric pain without gross elevation of WCC and ALT is now better understood. High anomalous insertion of the pancreatic duct into the CBD is now recognised as a frequent radiologic finding in choledochal cystic disease.³ The absence of a protective pancreatic duct sphincter allows free drainage of pancreatic juice into bile. Chemical cholangitis and bile duct epithelial injury may result from conversion of biliary phosphatidyl choline into lysophosphatidylcholine by pancreatic lipase A2.⁴ This admixture is also associated with a chemical self-limiting pancreatitis.

MRCP which uses very heavily T2-weighted sequences to image, serially and non invasively, the anatomy of the pancreatobiliary tree, is rapidly gaining popularity over ERCP.⁵ Axial and coronal reconstructions allow confident diagnosis and timely referral to a specialised hepatobiliary surgical unit. MRCP permits safe serial screening for malignant complications in the post operative years when ERCP is often made difficult by surgery.

Excision of as much of the ectatic or cystic extra hepatic biliary tree and restoration of biliary drainage using a Roux en Y hepaticojejunostomy to limit biliary reflux has replaced the older drainage procedures such as choledochoduodenostomy which have a high re-operation rate due to poor healing of the fibrous cyst walls. Failure to excise dilated biliary channels predisposes to recurrent cholestasis and does not remove the main source of dysplasia or malignancy.

The incidence of malignant transformation, usually adenocarcinoma is 14-18% in adults over the age of 20 years and reaches 50% at age 50 years. Most cancers are attributed to mitogenic bile. They occur in the choledochal cysts (58%) or



Figure 3: Postoperative follow-up axial MRCP image showing air in intrapancreatic CBD stump with anterior meniscus shaped air-fluid level.

gall bladder (40%)⁶, and are usually not associated with any biliary stones. A strong association between the frequent finding of an associated anomalous pancreatic biliary duct junction and gall bladder carcinoma has led different authors to advocate prophylactic cholecystectomy.⁷ Malignant transformation is reported less frequently in patients undergoing resection as compared to drainage procedures.⁸ Regular tumour surveillance by MRCP is recommended in all operated patients.

Conclusions

Recurrent abdominal pain in young patients associated with hyperamylasaemia and dilatation of the extrahepatic biliary system which persists despite endoscopic sphincterotomy and clearance of biliary calculi requires confirmation of Type I choledochal cystic disease with anomalous pancreatic duct drainage, preferably by MRCP, before consideration of cyst excision with Roux en Y hepaticojejunostomy followed by periodic surveillance for malignancy of the biliary tract.

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