Clinical Vignette

• 47 year old male with alcohol abuse and elevated liver enzymes.

• Ultrasound: dilated common hepatic duct measuring 9.5mm. No gallstone. (top image)

• CT: common duct dilatation as well as cecum malrotation. (middle image)

• MRCP and ERCP: moderate fusiform dilatation of the common hepatic duct with normal intrahepatic ducts and no stones.

• Management: referred to surgery for excision.

Definition

• Abnormal dilatation of the biliary ducts, intrahepatic and/or extrahepatic, which is congenital in nature.

• Incidence in the general population is 1:13000

Etiology

Abnormal formation of the biliary ducts in utero predisposes to reflux of bile and weakening of the duct walls.

Classification

Todani classification into 5 morphological types (Please refer to the table)

Clinical presentation

• 80% children, 20% adults

• Triad: jaundice, abdominal pain and mass in the right upper quadrant

Diagnosis

• Initial testing with ultrasound

• ERCP is definitive but invasive

• MRCP

  • Highly accurate, non-invasive
  • Demonstrates both extrahepatic and intrahepatic biliary ductal
  • Can demonstrate complications, such as cholangiocarcinoma, and used for pre-surgical planning.

Complications

• Cholangiocarcinoma (rare)

• Calculi, cholangitis, pancreatitis, rupture

Associations

• Situs anomalies (polysplenia)

• Gastrointestinal tract malrotation (few case reports, including our case)

• Duodenal atresia

Management

Surgical excision of the extrahepatic biliary tree + Roux-en-y anastomosis due to the risk of cholangiocarcinoma.

Conclusion

Choledochal cysts represent a spectrum of congenital anomalies revolving around abnormal dilatation of the biliary ductal system and pancreaticobiliary junction. It classically presents with a triad of jaundice, abdominal pain and mass, usually in children, but can be incidentally discovered in adults on imaging for other suspected causes of jaundice. It is associated with a number of congenital anomalies, of which gastrointestinal malrotation is a rarely reported association. Imaging is the mainstay of diagnosis (MRCP/ERCP), and management is surgical excision, due to the risk of cholangiocarcinoma.