A Rare Case of Type 1 Choledochal Cyst with Type C Anomalous Pancreaticobiliary Maljunction in a 52-year-old Patient

Hunza Chaudhry[1], Timothy Wang[2], Jayakrishna Chintanaboina[2]

[1]Department of Internal Medicine, University of California, San Francisco- Fresno, California,

[2]Department of Gastroenterology and Hepatology, University of California, San Francisco-Fresno, California

UCSF Fresno

Introduction

- Choledochal cysts are anomalies of the bile duct that generally present in childhood but can rarely be delayed until adulthood in 20% of cases.
- They are often diagnosed incidentally or present with acute pancreatitis or cholangitis.
- We report a case of a type 1 choledochal cyst (T1CC) and type C anomalous pancreaticobiliary maljunction (PBM), successfully managed by a multidisciplinary approach.

Case Description

- A 52-year-old female presented with right upper quadrant pain, jaundice and vomiting. Labs revealed elevated lipase, bilirubin, AST, ALT and alkaline phosphatase.
- A CT of the abdomen showed no gallstones but a marked dilatation of extrahepatic bile duct with a transition point in the distal intrapancreatic portion (Figure 1a).
- ERCP showed fusiform dilation of the CBD consistent with T1CC (Figure 1b). Spyglass cholangioscopy revealed focal stenosis in the distal CBD joining above the ventral pancreatic duct opening within the long common channel, suggestive of type C anomalous PBM (Figure 1c).
- Plastic biliary stent was placed into the CBD. EUS guided fine needle biopsy of the stricture showed no evidence of malignancy.

Images

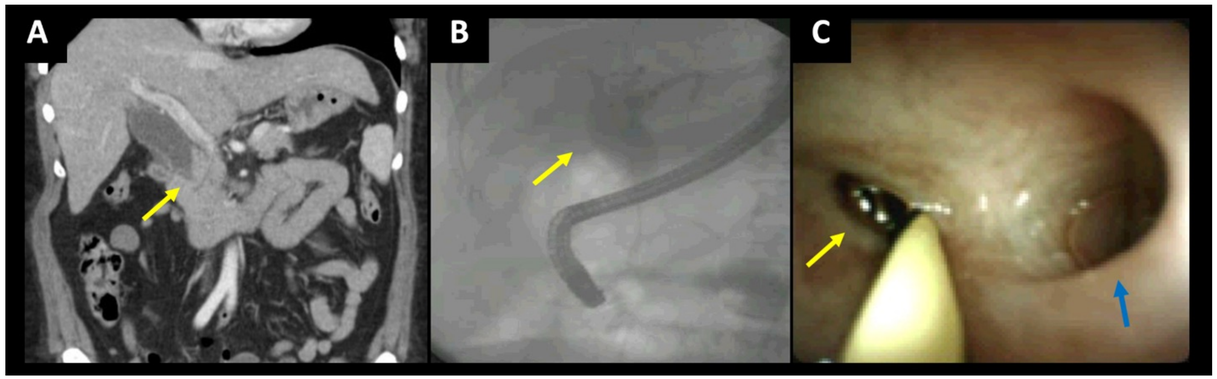

Figure 1: A. CT of the abdomen showing marked dilatation of extrahepatic bile duct with a transition point in the distal intrapancreatic portion. **B.** Cholangiogram showing Type 1 Choledochal Cyst and Type C Anomalous Pancreaticobiliary Maljunction. **C.** Spyglass cholangioscopy showing Jagwire™ within the CBD (yellow arrow) revealing focal stenotic CBD joining the common channel just above the ventral pancreatic duct (blue arrow).

Type	Description
Type I	Fusiform dilation of the CBD
Type II	Bile duct diverticulum
Type III	Saccular dilation of the intraduodenal portion of the CBD
Type IVa	Multiple cysts at intra and extrahepatic ducts
Type IVb	Multiple cysts at extrahepatic ducts only
Type V	Fusiform or saccular cystic dilation of intrahepatic bile ducts

Case Description (contd.)

- Her clinical course was complicated by multiple admissions due to stent occlusion.
- After multidisciplinary discussion, the patient underwent choledochal cyst resection with hepaticojejunostomy, cholecystectomy and biliary reconstruction.
- Intraoperative gallbladder specimen revealed poorly differentiated adenocarcinoma with plan for chemotherapy.

Discussion

- Etiology of choledochal cysts remains unclear and while some believe these cysts are congenital in nature, others propose they arise from anomalous PBM.
- Anomalous PBM leads to reflux of biliary secretions leading to chronic inflammation, increasing the risk of developing cholangiocarcinoma and gallbladder cancer, as seen in our patient.
- Placement of biliary stents can temporarily alleviate symptoms, but surgical excision is the definitive treatment.
- Our case highlights the importance of including type 1 choledochal cyst in the differential diagnosis even in adult patients with a significantly dilated CBD.
- A thorough endoscopic and radiological work up should be done to prevent progression to malignancy.