

Mixed Choledochal Cyst (Type I and II) Associated With a Malformation of the Pancreatobiliary Junction. A Case Report and Review of the Literature^{☆,☆☆}

Quiste de colédoco mixto (tipo I y II) asociado a malformación de la unión pancreatobiliar. Reporte de un caso y revisión de la literatura

Choledochal cysts are a rare disease, with an incidence in western countries of 1 in 100 000–150 000 births.¹ The rate is considerably higher in Asian populations, where the reported incidence is 1 in 1000 births. The exact etiology is still unknown, although they predominantly affect females with a ratio of 4:1.¹ According to the Todani classification, the distribution of the different types of choledochal cysts is: type I 50%–80%; type II 2%; type III 1.4%–4.5%; type IV 15%–35%; type V 20%.¹ Mixed type I and II choledochal cysts are extremely rare and represent less than 1.1% of cases. A review of the literature has found only 6 reported cases.^{2,3} Around 80% present with symptoms before the age of 10. The described symptomatic triad of abdominal pain, jaundice and a palpable abdominal mass occurs in less than 20%.⁴

The complications of choledochal cysts include bile stasis, the formation of calculi, recurring infections and inflammation. The initial study of these patients involves ultrasound, which is able to establish a diagnosis in most cases. The pancreato-biliary malformations entail a common canal between the junction of the pancreatic duct and the bile duct, which are outside the duodenal wall.⁵ These malformations are a risk factor for neoplasms of the biliary tract. Pancreatic juices and bile combine constantly, producing toxic substances. Due to this, the mucus of the bile tract is repeatedly damaged and repaired on many occasions, which causes an acceleration of proliferative activity and multiple gene mutations. In turn, this causes histological changes, such as hyperplastic epithelium, metaplastic epithelium, and dysplastic epithelium, which lead to carcinogenesis of the bile duct.⁶

The frequency of anomalies at the pancreato-biliary junction ranges between 0.08% and 3.2% of the population.⁷ Kimura defined 2 types of anomalies of the pancreato-biliary junction. In type 1, the pancreatic duct empties into the common bile duct at an acute angle and the distal bile duct does not have stenosis. In type 2 (95%), it is the common bile duct that empties into the pancreatic duct and the distal bile duct usually presents stenosis; therefore, there is greater and earlier dilatation of the bile duct.⁸ In this article, we report the case of a mixed type I and II choledochal cyst associated with a Kimura type 2 malformation of the pancreato-biliary junction.

Case Report

The patient is a 20-year-old male with a history of episodes of acute pancreatitis of undetermined etiology that had been treated in primary care clinics, as well as several episodes of jaundice. He was referred to our unit for study and treatment. Abdominal ultrasound showed evidence of dilatation of the intra- and extrahepatic bile duct, a choledochal cyst and pancreas with inflammation. Abdominal tomography showed dilatation of the intra- and extrahepatic bile duct up to the ampullary region. Surgical findings were: a gallbladder measuring 8×4×2 cm, which drained through the cystic duct to a diverticulum measuring 2×2×1 cm of the common bile duct, which was dilated to 2.5 cm, approximately (Fig. 1). Transcystic cholangiography was performed, and a dilated type II choledochal cyst was observed. We proceeded with diverticulectomy/cholecystectomy, choledochorrhaphy and Kehr tube placement. The histopathology study reported a gallbladder measuring 7×3×1 cm, with a 3 mm wall and hemorrhagic-looking mucosa, drainage of the cystic duct to a

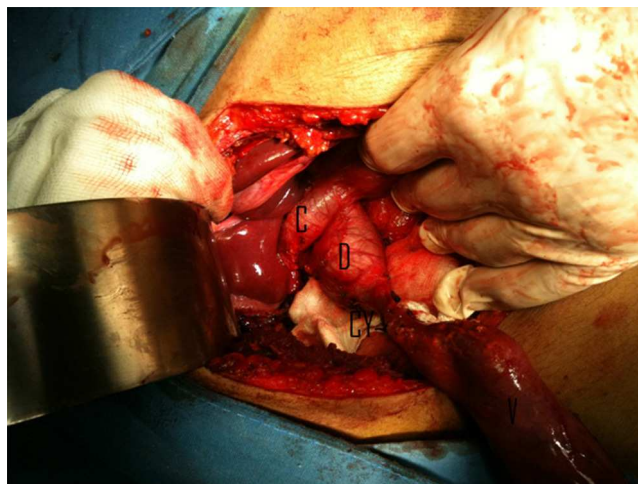


Fig. 1 – Exploratory laparotomy: dilated common bile duct (C), diverticulum (D), cystic duct (CY), and gallbladder (V).

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cystic lesion measuring 2×1×1 cm with irregular outer surface and thickened walls.

During post-op, there was a high level of bile discharge through the Kehr tube of up to 2100 cc in 24 h. Cytology was ordered of the bile liquid, which reported 183,654 IU amylase and 159.617 IU lipase. Cholangiography was performed through the Kehr tube, and a common bile duct of 18 mm was observed as well as pancreato-biliary malformation (Fig. 2). There was no evidence of residual calculi and adequate passage of contrast medium to the duodenum was observed. We therefore decided to close the Kehr tube and scheduled an endoscopic retrograde cholangiopancreatography (ERCP) and sphincterotomy in order to reduce pancreato-biliary reflux, both of which were done without incident. Cytology of the bile liquid showed that the pancreatic enzymes had doubled over the previous analysis, so the diagnosis of pancreato-biliary reflux was established. We decided to perform exeresis of the main bile duct with Roux-en-Y hepaticojejunal anastomosis, with findings of type I choledochal cyst. The histopathology study reported a common bile duct measuring 3.5×2.5 cm, follicular lymphoid hyperplasia and no evidence of malignant cells. The patient presented an uneventful recovery and, 12 months after surgery, has had no new episodes of pancreatitis or jaundice.

In a review of 356 patients over a period of 40 years, Kaneyama reported 4 cases in children who presented dilatation of the main bile duct with the presence of a diverticulum in the middle third of the duct, and the gallbladder drained by means of a cystic duct directly to the diverticulum.² In 2003, Katsinelos reported a similar case of a mixed type I and II cyst with pancreato-biliary junction malformation in a 72-year-old woman that had begun with an

episode of pancreatitis. The patient was managed conservatively with endoscopic cholangiopancreatography and sphincterotomy.³ In our case, we opted to perform cholecystectomy and resection of the diverticulum with Kehr tube placement; afterwards, we performed an endoscopic cholangiopancreatography study with extensive sphincterotomy, which showed evidence of a long pancreato-biliary common duct measuring 24.5 mm (normal range 4.6±2.6 mm).⁹ With these criteria, we established the diagnosis of pancreatic reflux. In our case, there was evidence of that the sphincterotomy did not diminish the pancreato-biliary reflux. Given the evidence of pancreato-biliary malformation associated with dilatation of less than 20 mm, complete exeresis of the bile duct was indicated.

Congenital pancreato-biliary malformations have a risk of malignization of 10%–30%, which is 20–30 times more frequent than in the general population.¹⁰ In these cases, it is essential to utilize precise imaging techniques and to evaluate the risk-benefit of preoperative ERCP to document pancreato-biliary malformations.

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Fig. 2 – Cholangiography through the Kehr tube; pancreato-biliary malformation, Kimura 2.

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From bariatric surgery to a radical total gastrectomy: A change in the proposed surgical procedure due to intraoperative diagnosis of carcinoid tumour[☆]

De cirugía bariátrica a gastrectomía total radical: Cambio del procedimiento quirúrgico previsto por hallazgo operatorio de un tumor carcinoide

Gastric carcinoid tumors (GCT) are benign neuroendocrine cell tumors of the glands of the body and fundus of the stomach. Some 70% of these tumors are located in the digestive tube (more in the small intestine and appendix). The probability of lymph node invasion depends on tumor size: 2% in tumors smaller than 1 cm, between 10% and 15% in those measuring 1–2 cm, and 60%–70% in tumors larger than 2 cm.^{1–4} The most frequent locations are the body and fundus of the stomach; when there is associated pernicious anemia, 50% are multifocal. Treatment depends on size, possibility of lymph node involvement and whether there are multiple foci. In our case, in a patient who was a candidate for bariatric surgery, preoperative gastroscopy revealed a gastric wall lesion, whose final pathology was GCT. Gastroscopy before bariatric surgery can significantly reduce the number of potentially malignant gastric lesions, which may inadvertently remain in the gastric remnant in cases of bypass surgeries without gastric resection, such as gastric bypass.³

The patient is a 28-year-old woman who had been referred to the Obesity Unit due to progressive weight gain after her first pregnancy and failed attempts to lose weight with low-calorie diets and physical activity (weight 110.5, height 152 cm, BMI 47). She reported having extrinsic asthma. She provided a gastroscopy report from a study done in another hospital 1 year before, which described a 3 mm polypoid lesion in the prepyloric antrum (biopsy: compatible with chronic antral gastritis with intestinal metaplasia and *Helicobacter pylori*+). After being assessed by the Obesity Unit, she was considered

as candidate for bariatric surgery, and preoperative studies were initiated in accordance with the hospital protocol. *H. pylori* was eradicated.

The study of the gastric mass was completed with another gastroscopy, which detected a raised, umbilicated lesion measuring 2 cm on the posterior side of the body of the stomach. Biopsy was non-specific. Endoscopic ultrasound showed a subepithelial mass on the posterior side of where the body and fundus meet, with central ulceration and slight depression. In the area of the lesion, a hypoechoic image was observed with central hyperechogenicity; it was round, measured 14 mm×10 mm, and appeared to depend on the longitudinal portion of the fourth layer or *muscularis propria* (Fig. 1).

The remaining preoperative studies showed no notable alterations.

The case was discussed with the Digestive Department, and the most probable diagnosis of the lesion was thought to be a 1.4 cm GIST tumor. Its location would enable a vertical gastrectomy to be done with resection of the lesion in the gastrectomy specimen as it seemed to be located on the posterior side toward the greater curvature of the stomach at the junction of the body and fundus.

The findings from the gastroscopy and endoscopic ultrasound were explained to the patient. Bariatric surgery was proposed, including resection of the gastric mass. The most likely surgical options would be either vertical sleeve gastrectomy or gastric bypass with gastric resection, and we explained to the patient that the final technique would depend on the intraoperative findings.

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