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Intraductal Papillary Neoplasm of the Bile Duct (IPN-B) with Low-Grade Dysplasia in a Choledochal Cyst Type I: A Case Report

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Intraductal papillary neoplasm of the bile duct (IPN-B) is a rare premalignant lesion and precursor of cholangiocarcinoma. Very few cases of IPN-B in a choledochal cyst have been reported, with none published in the Philippines. Reported here is the case of a 53-year-old female with a Type I choledochal cyst diagnosed in childhood who previously underwent choledochal cystoduodenostomy. She remained asymptomatic for decades until she presented with intermittent cholangitis. Work-up and imaging suggested biliary stricture, and ERCP with biliary stenting was performed. After resolution of cholangitis, she underwent elective biliary reoperation with complete excision of the choledochal cyst and Roux-en-Y hepaticojejunostomy. Histopathology demonstrated IPN-B with low-grade dysplasia in the choledochal cyst. Despite nonspecific preoperative findings, neoplasia should be a consideration in the evaluation of unresected choledochal cysts. Complete cyst excision and long-term surveillance are essential to reduce the risk of cholangiocarcinoma.

Key words: Cholangitis; bile duct; choledochal cyst; cholangiopancreatography, endoscopic retrograde

Intraductal papillary neoplasm of the bile duct (IPN-B) is a rare premalignant neoplasm of cholangiocarcinoma and accounts for only 4 to 15% of bile duct tumors.[1] Its high malignant potential was documented in 30 to 80% of patients with invasive components.[2] Clinically, patients usually have intermittent abdominal pain, jaundice and cholangitis.[3] Diagnostic work-up for any biliary tract disease can be done and imaging may demonstrate an intraductal mass, bile duct dilatation, or a cystic lesion.[1,3] Among the identified risk factors are hepatolithiasis, cholecystolithiasis, choledocholithiasis, liver fluke (Clonorchis or Opisthorchis) infection, primary sclerosing cholangitis, familial adenomatous polyposis or Gardner syndrome, and biliary malformations including choledochal cyst.[1,3]

This case presents a rare occurrence of IPN-B with low-grade dysplasia in an unresected Type I choledochal cyst in a patient previously managed with a biliary-enteric bypass. It highlights the need to maintain a high index of suspicion for premalignant transformation in patients with recurrent cholangitis despite nonspecific preoperative imaging findings, and it emphasizes the need for complete surgical resection of choledochal cysts and long-term postoperative surveillance to reduce the risk of progression to cholangiocarcinoma.

This case is reported in line with the SCARE checklist.[4]

The Case

A 53-year-old female, single, who is a known case of choledochal cyst type I diagnosed during childhood and underwent a biliary surgery more than 40 years ago, sought consultation due to symptoms of intermittent cholangitis (abdominal pain, fever and jaundice). The patient had poor recall of her father's account of the medical condition and operation she had during childhood. However, the patient was apparently well thereafter and thus lost to follow-up. Interval history of one year prior to admission, she presented with intermittent epigastric pain associated with occasional vomiting and fever. She was treated as a case of cholangitis with antibiotics, a pain reliever and a proton pump inhibitor.

Five months before admission, the persistence symptoms along with the onset of jaundice prompted consultation. Ultrasound was unremarkable, and a

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subsequent CT scan with contrast revealed mildly dilated intrahepatic and extrahepatic bile duct radicles, pneumobilia, and a dilated common bile duct (CBD) with a transverse diameter of 2.6 cm (Figure 1). With the impression of a distal biliary tract stricture, the patient was treated as a case of recurrent cholangitis. Intravenous antibiotics were given, and she underwent endoscopic retrograde cholangiopancreatography (ERCP) with stenting for biliary decompression. After ERCP, magnetic resonance cholangiopancreatography (MRCP) was done for definitive surgical planning. It showed a cystic dilatation of the CBD (up to 2.9 cm in diameter) with intracystic signal suggestive of sludge settled in the posterior aspect of the cyst; the stent was seen within the CBD (Figures 2 and 3). No discrete mass could be identified in the CBD. A stricture, however, could not be entirely excluded from the choledochal cyst (Type 1) down to the preampullary segment. Additionally, serum markers CA19-9 and CEA were not elevated. With the resolution of cholangitis, the patient was discharged and scheduled for an elective definitive surgical intervention.

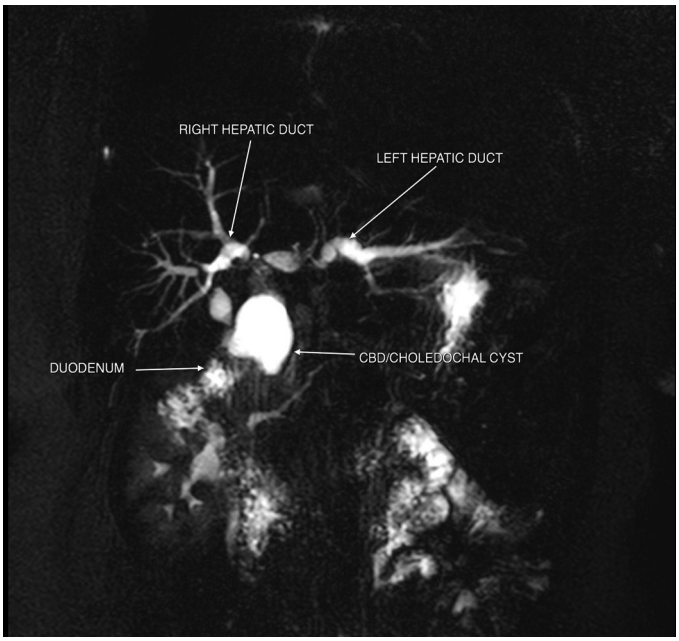


Figure 2. The targeted area in sagittal, axial and coronal view (counterclockwise) shows the choledochal cyst (crosshair) in its widest transverse diameter at 2.6 cm in the CT scan with contrast.

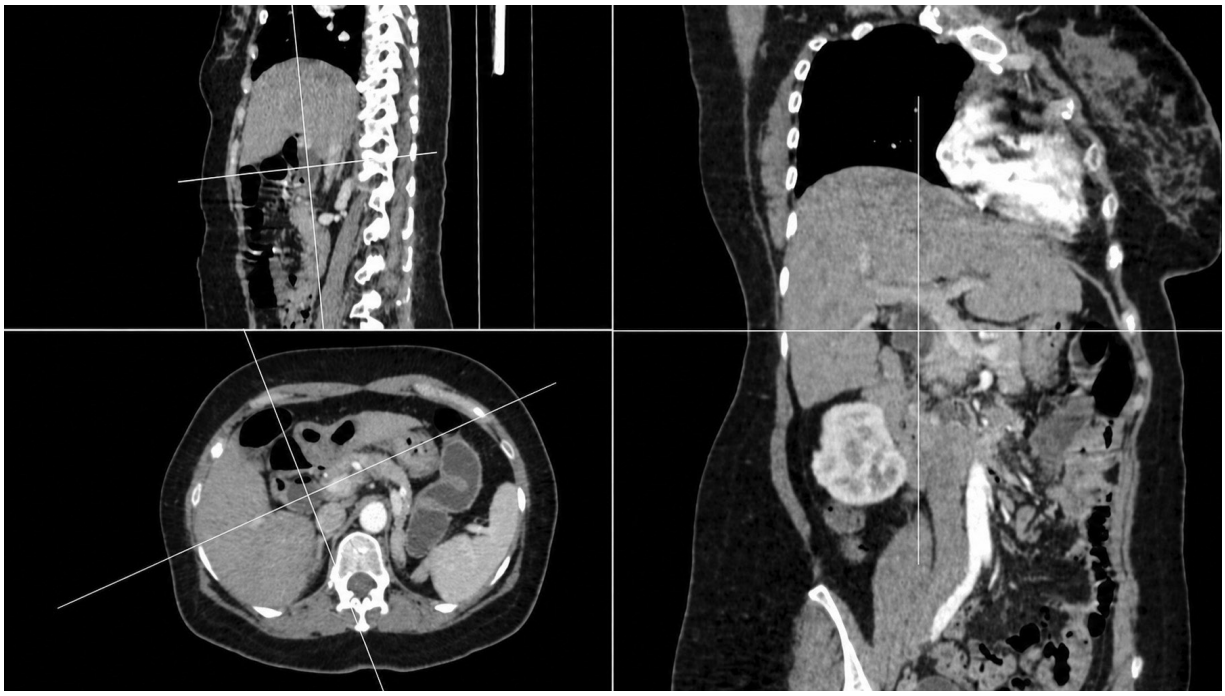


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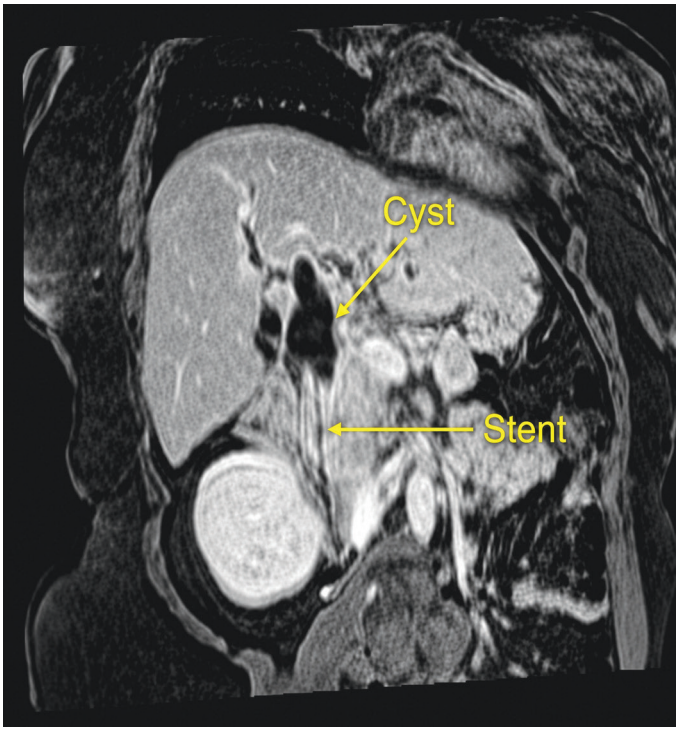


Figure 3. Coronal MRCP demonstrating a patent stent within the common bile duct (CBD) extending to the ampulla.

Except for a total abdominal hysterectomy and bilateral salpingo-oophorectomy (TAH-BSO) five years prior for abnormal uterine bleeding from multiple uterine myomas, her past medical, personal, and family histories are unremarkable.

On admission, the patient was awake, ambulatory, conversant, afebrile, and with stable and normal vital signs. Her BMI was 25.1 kg/m^2 (overweight). She was not jaundiced. There was a right subcostal abdominal incision scar. The rest of her physical examination was unremarkable.

The patient was scheduled for surgery under general anesthesia that same day of admission with a pre-operative diagnosis of recurrent cholangitis probably secondary to biliary stricture, S/P ERCP with stenting, status post biliary surgery (1984) for choledochal cyst Type I. An extended right subcostal incision through the previous incision was done. There were moderate adhesions in the portahepatis area. The choledochal cyst was also adherent to the surrounding structures, including the portal vein and the hepatic artery. After the initial dissection of the gallbladder and cystic duct, a transcystic cholangiogram (Figure 4) was performed to assess how

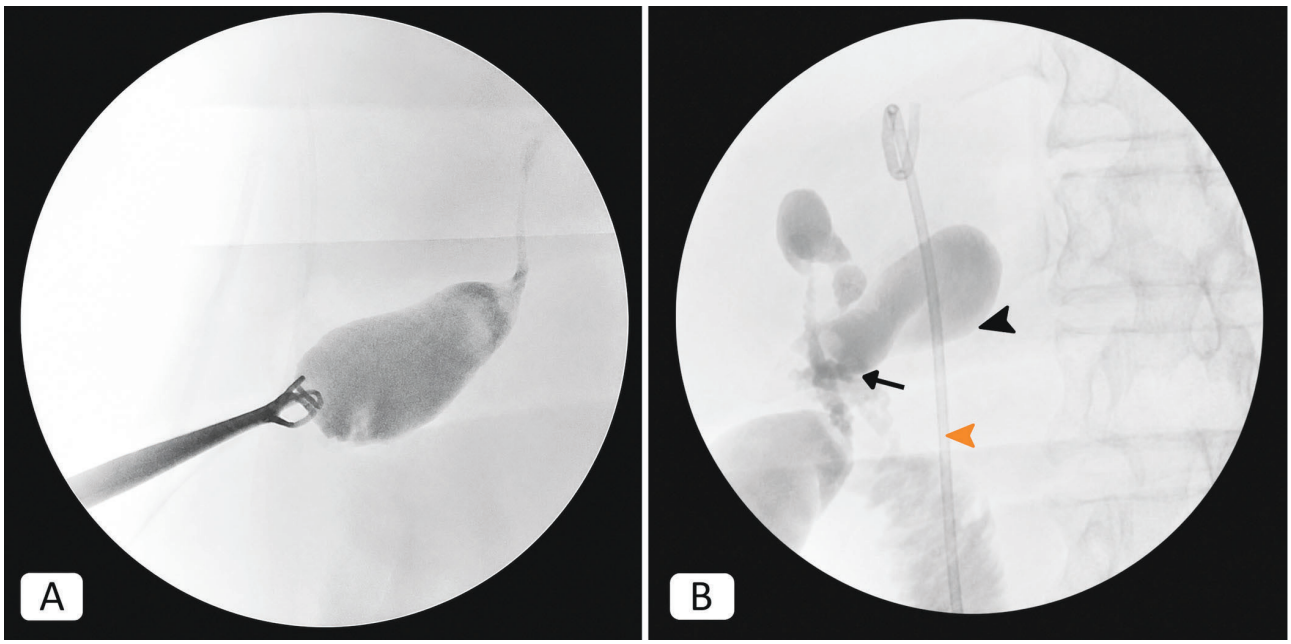


Figure 4. Intraoperative cholangiogram. (A) Image demonstrates opacification of the gallbladder. (B) A fusiform dilatation of the common bile duct is visible (black arrowhead), showing preferential flow into the duodenum via the cystoduodenostomy (black arrow). A stent (orange arrowhead) is seen running through the cyst into the duodenum.

the previous surgery had altered the patient's anatomy. This study revealed a fusiform dilatation of the common bile duct about 5 cm x 3 cm with an intact stent. There was also a strictured choledochal cystoduodenostomy about 5 to 6 mm in diameter and a long cystic duct draining into the choledochal cyst (Figure 5). The surgery proceeded with cholecystectomy, takedown of the choledochal cystoduodenostomy, duodenorrhaphy, excision of choledochal cyst (Lily's technique), Roux-en-Y hepaticojejunostomy, and drain placement. Lily's technique involves excision of the thick inner lining of the posterior wall of the choledochal cyst. The thin outer layer is retained to minimize injury to the portal vein or hepatic artery commonly adherent to this after previous inflammation or surgery.[5] The patient had an unremarkable postoperative course and was discharged on the fifth postoperative day.

The histopathology report on the submitted choledochal cyst wall was consistent with an intraductal papillary neoplasm with low-grade dysplasia, arising in a choledochal cyst with acute and chronic inflammation (Figures 6 and 7). The tumor size was 1.5 cm and was noted in only one of the cyst wall fragments.

On follow-up, there was no further recurrence of abdominal pain, vomiting, fever, and jaundice. Since the resection margin was not checked by frozen section intraoperatively nor confirmed on final biopsy, the patient was advised about the importance of a regular long-term follow-up every 3 to 6 months for clinical examination, blood examination (CA 19-9, CEA, liver function tests, bilirubin profile), and ultrasound. Any suspicious findings would be further worked up with MRCP.

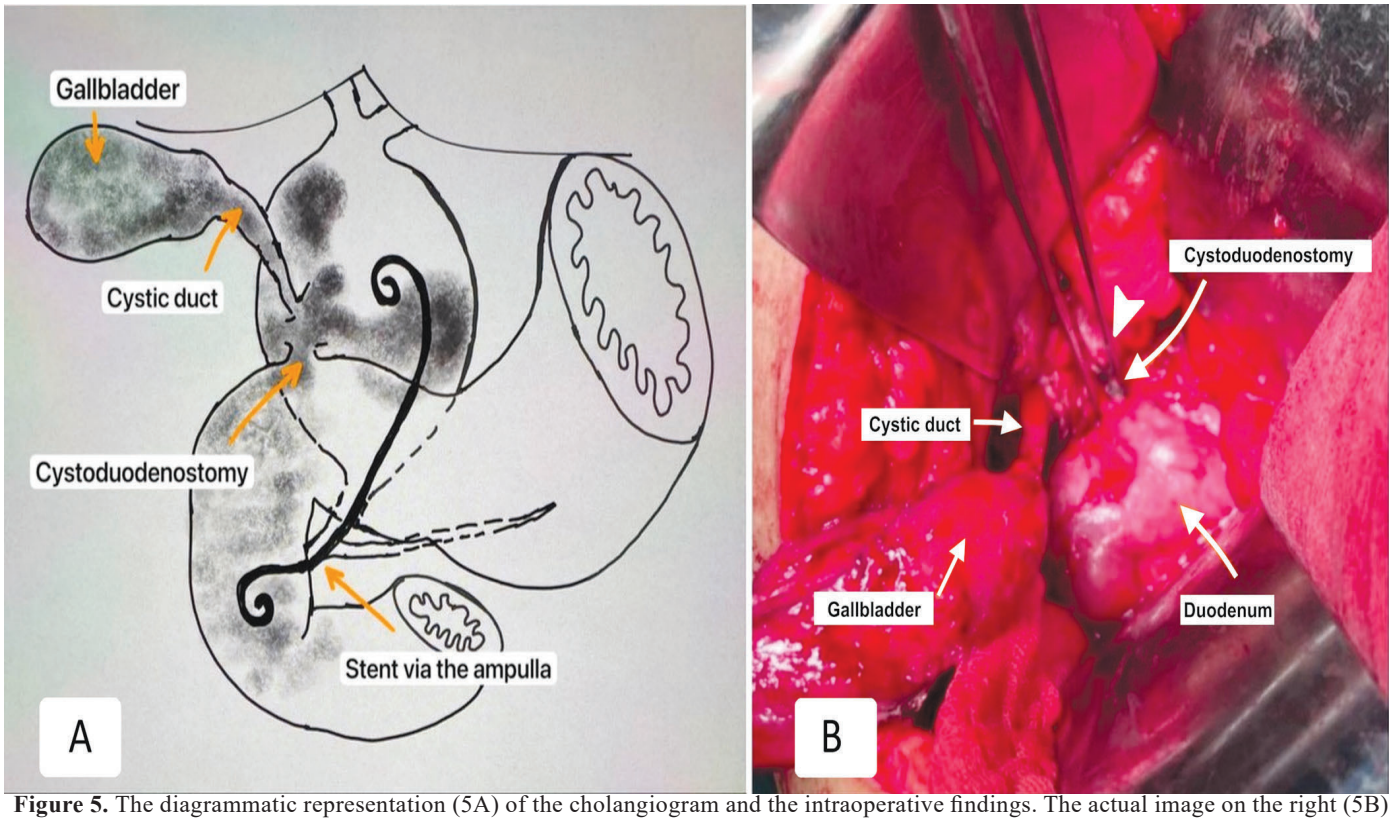


Figure 5. The diagrammatic representation (5A) of the cholangiogram and the intraoperative findings. The actual image on the right (5B) shows the Debakey forceps holding the cystoduodenostomy measuring about 5 to 6 mm and a long cystic duct draining into the choledochal cyst (arrowhead).

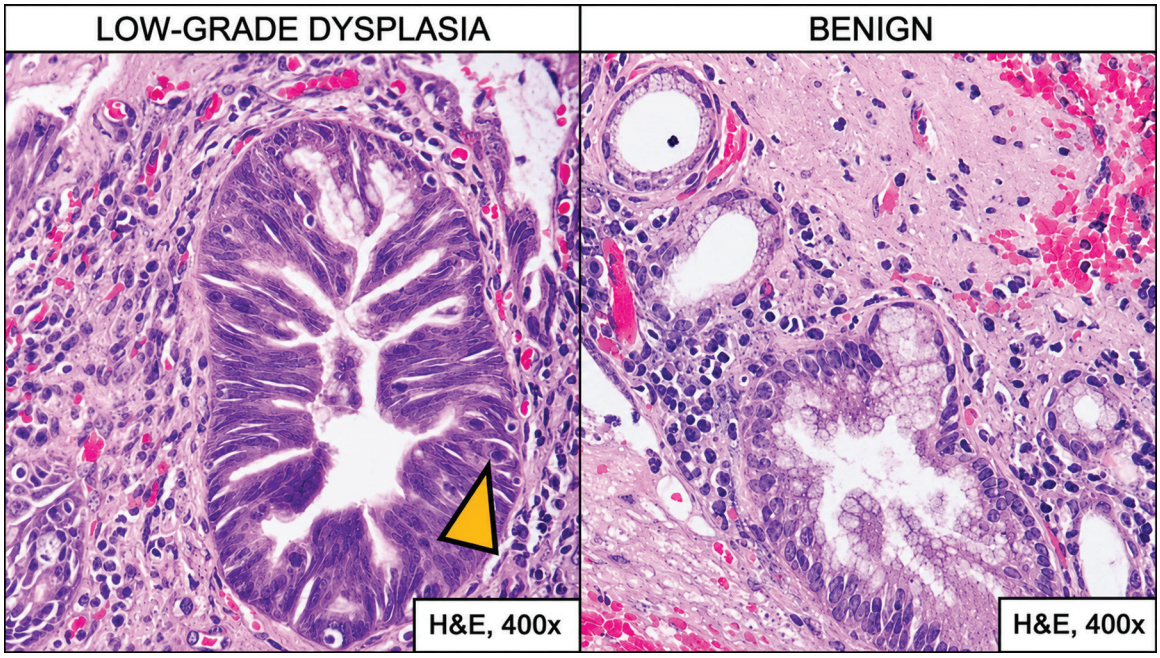


Figure 7. Low-grade dysplasia in IPNB. High-magnification view (400×) identifies low-grade dysplasia of the neoplastic lining epithelium (yellow arrowhead), displayed adjacent to benign, morphologically normal epithelium for comparison.

Discussion

Intraductal papillary neoplasm of the bile duct (IPN-B) was first proposed as a distinct entity in the 2010 revision of the World Health Organization (WHO) classification for liver and intrahepatic bile duct tumors.[1] IPN-B features intraductal papillary or villous glands lined by neoplastic epithelium with fine fibrovascular stalks in the dilated intrahepatic or extrahepatic bile ducts.[3] In 2019, IPN-B was classified by the WHO into four histological subtypes: intestinal IPN-B, gastric IPN-B, pancreaticobiliary IPN-B and oncocytic IPN-B.[1,6] The pancreaticobiliary subtype is usually associated with high-grade dysplasia and aggressive behavior while the gastric subtype is more commonly with low grade dysplasia.[1]

A subclassification of IPN-Bs was proposed by the Japan-Korea Pathologist Group and was further differentiated by Aoki et al.[1,6] Type 1 was the "non-invasive phenotype" associated with intestinal and oncocytic subtypes, overt mucin production, intrahepatic duct development, KRAS mutation, and relatively good prognosis. On the other hand, Type 2 was the "invasive phenotype" with pancreaticobiliary subtype, less often mucin hypersecretion, extrahepatic duct development, mutations in TP53 and SMAD4, aberrant expression of p53 and SMAD4, and poorer prognosis than Type 1.[1,6] Given the low-grade dysplasia and intestinal differentiation finding in the case of this patient, her IPN-B subtype is most likely the non-invasive Type 1 intestinal subtype.

Comparison was also made between IPN-B and intraductal papillary mucinous neoplasm (IPMN) of the pancreas because of shared features. Both have prominent intraductal papillary proliferation, four histological types, duct-associated cyst formation, ductal dilatation and possible mucin overproduction.[1] Type 1 intestinal IPN-B and oncocytic IPN-B may be considered the biliary counterparts of intestinal-type pancreatic IPMN and oncocytic pancreatic IPMN, respectively, due to similar genetic and molecular characteristics.[1] However, IPN-B is generally distinct from pancreatic IPMN because of a

more advanced stage, a higher histologic grade, a worse prognosis, a higher incidence of associated invasive cancer and some differences in genetic alterations.[1]

In a meta-analysis by Gordon-Weeks, et al. of 276 IPN-B patients, only 18% had adenoma, 20% had dysplasia as in the patient in the present case, 20% had carcinoma-in-situ and 43% had invasive carcinoma.[7] The tumor was confined to the bile duct wall in 63% of cases, transmural in 28%, invaded adjacent organs in 8% and sometimes had lymphatic dissemination in 9% of cases.[7] Our patient's tumor was confined to the bile duct wall like in most cases.

Clinically, patients with IPN-B are typically between 50 and 70 years of age, with slight male predominance and present with intermittent abdominal pain, jaundice and acute cholangitis[3,6,8], however, up to 30% of cases can be asymptomatic.[6] The symptoms can be due to intermittent obstruction from friable lesions that are easily detached from their origin or from the abundant mucin secreted by the tumor.[3] The high viscosity of the mucin can also contribute to bile duct obstruction and dilatation.[1] The patient's profile was consistent with the age bracket; however, being a female partly contributes to the rarity of this case. Symptoms of intermittent acute cholangitis were also present in the patient, though the strictured biliary-enteric anastomosis could be another factor contributing to the biliary obstruction.

Diagnostic workup for IPN-B is the same as with other biliary tract disease which includes ultrasound, computed tomography (CT), magnetic resonance imaging (MRI), magnetic resonance cholangiopancreatography (MRCP), endoscopic retrograde cholangiopancreatography (ERCP), cholangioscopy, endoscopic ultrasound (EUS), intraductal miniprobe ultrasound (IDUS) and contrast-enhanced ultrasound.[1] The radiologic features of IPN-B are intraductal mass, cystic lesion, bile duct dilatation with both upstream and downstream dilatation for mucin overproduction subtypes, and periductal invasion including macroinvasion of the liver.[1,3] However, challenges may be encountered because mucin production may prevent opacification of the entire biliary tree in ERCP and percutaneous transhepatic cholangiography (PTC) while conventional imaging may overlook small papillomas.[3] This was demonstrated in the present case since ultrasound and CT scan did not show any intraductal mass or papilloma. Preoperative biopsy can also be done if solid lesions can be identified using percutaneous ultrasound-guided needle biopsy, ERCP forceps biopsy, cholangioscopic mini forceps biopsy, EUS fine needle biopsy or brush cytology from stenosed areas.[1] However, there was a report of cutaneous needle tract seeding 23 months after a needle biopsy procedure.[1] Additionally, the use of different markers such as CA 19-9, CEA, alpha-fetoprotein (AFP), and various mucins (e.g., MUC1 and MUC5AC) may be used in combination to improve diagnostic accuracy.[6]

Radical surgical resection with adequate margins is the first line and standard treatment for IPN-B without metastasis.[3,9] The tumor location, extension, depth of invasion, and lymph node metastasis dictate the type of surgery[8] which may include excision of the extrahepatic biliary tree, pancreaticoduodenectomy, major hepatectomy, or liver transplantation.[3,9] Frozen section biopsy of the bile duct margin plays a role in ensuring complete resection and excluding dysplasia or tumor involvement.[3,8] For patients who are high-risk or are not candidates for surgery, radiofrequency ablation (RFA), photodynamic therapy (PDT), or argon plasma coagulation (APC) are viable options.[1,6,9] Ongoing research in immunotherapy with checkpoint inhibitors and microbiome-targeting therapies has also shown promising results.[6]

The overall survival and postsurgical outcomes of IPN-B are better than conventional flat intraductal neoplasia-associated cholangiocarcinoma.[2,3] The 5-year survival after curative resection was 81% for IPN-B while for cholangiocarcinoma it was less than 51.6%.[2] The prognostic factors affecting long-term survival are the depth and percentage of invasive component, resection margin status, and cellular type of IPN-B.[3] After surgical resection, benign IPN-B has a recurrence rate of approximately 20%, while its malignant counterpart has around 60% recurrence.[2] Recurrence can be attributed to preoperatively undetected lesions on conventional imaging.[2,3]

Hence, to achieve better outcomes, a meticulous assessment of the tumor's extent before surgery, complete removal of the tumor, and regular monitoring every 3 to 6 months are essential.[2] Long-term monitoring with MRCP is also advised due to the high recurrence rate.[10]

A similar case was reported by Tarhan, et al. in Turkey where a 31-year-old female presented with abdominal pain and was diagnosed with choledochal cyst Type 1 on MRI/MRCP. The patient underwent total extrahepatic bile duct excision, cholecystectomy, and Roux-en-Y hepaticojejunostomy.[11] Final biopsy revealed intraductal papillary neoplasm in the excised extrahepatic bile duct.[11] The authors planned to conduct a surveillance of annual blood tests (CA 19-9 and parameters of cholestasis) and abdominal ultrasound, and MRI/ MRCP for any suspicious findings.[11]

Conclusion

With the challenges in the diagnosis of IPN-B in an unresected choledochal cyst, a high index of suspicion should be maintained in patients presenting with repeated bouts of cholangitis and biliary obstruction, even with no identified intraductal mass on conventional imaging. Appropriate use and interpretation of diagnostic imaging workup preoperatively can be a key to adequate surgical treatment planning. This can be further aided by a preoperative biopsy if solid lesions are identified on imaging. Additionally, an intraoperative frozen section biopsy to check for negative resection margins should be requested. Optimal management for the best possible outcome includes complete surgical resection of the choledochal cyst, including the removal of any abnormal wall findings on imaging studies whenever possible, followed by regular long-term surveillance.

Clinical Images

These were taken and presented herein with the consent of the patient.

Acknowledgement

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