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Surgical management based on EUS findings in a rare case of Intraductal papillary mucinous neoplasm of the bile duct (IPMN-B) with invasive adenocarcinoma

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Intraductal papillary mucinous neoplasm of the bile duct (IPMN-B) is a usually non-invasive macroscopic neoplasm that can precede biliary cancer[1], such as cholangiocarcinoma, a rare biliary adenocarcinoma associated with poor outcomes. It is determined by a dilated extrahepatic or intrahepatic bile duct lined with papillary or villous neoplastic glands with a delicate fibrovascular stalk[2]. This neoplasm can occasionally be invasive, being known as IPMN-B associated with invasive carcinoma, a feature that is shared with intraductal papillary mucinous neoplasms (IPMNs), a pancreatic counterpart of IPMN-B[3]. There are four classifications for IPMN-B, based on histological and immunohistochemical features, being the pancreatobiliary type the most common on Western countries[4], usually found in patients between 60 and 70 years-old, and with almost equal distribution between men and women[1], with the five-year survival about 10%[5]. Most patients are asymptomatic in early stages, and present biliary obstruction symptoms such as jaundice, abdominal pain and weight loss on advanced stage disease[6]. The diagnostic is made by laboratory analysis (CA 19.9) and endoscopic cholangiography (brushing, cytology or FISH)[6]. Surgery resection is the preferred treatment, but only ⅓ of the patients have early stage disease that is amenable surgical resection with curative intention[7].

A 64-years-old male patient complained of superior abdominal pain for 2 months, associated with choluria, fecal acolia and jaundice for 5 days. The patient was a social drinker. His clinical physical exam was normal except for the jaundice. His laboratory tests revealed high levels of hepatic enzymes and bilirubin, especially direct bilirubin. Also, his CA-19.9 was at 238 U/mL and carcinoembryonic antigen at 8.99 ng/mL. Ultrasound showed normal gallbladder and dilated intra and extra hepatic bile ducts, and upper gastrointestinal endoscopy showed mild erosive gastritis. Computed tomography showed a mass in the distal portion of the common bile duct, close to the duodenal papilla, with dilation of the bile ducts (FIGURE 1). An endoscopic retrograde cholangiopancreatography was performed. The contrast-enhanced image revealed a filling defect of the supra pancreatic segment of the

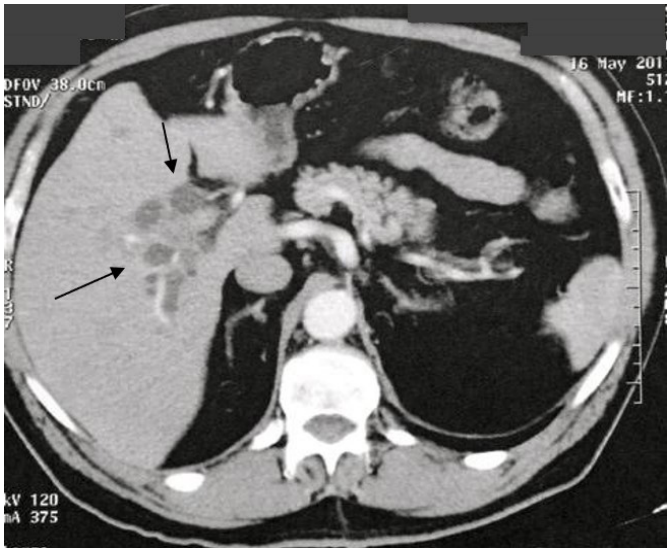

FIGURE 1. Computerized tomography shows a distal common bile duct unspecified mass associated with dilatation of intra and extra hepatic bile ducts.

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***E-VIDEO: https://youtu.be/iKf7CRYIXMI**

common bile duct (FIGURE 2). The papillotomy was done, the bile was drained, and the plastic stent positioned (FIGURE 3). Due to the uncertain diagnosis an EUS was performed (FIGURE 4 and E-VIDEO*). The lesion extended from the intrapancreatic common bile duct up to the hepatic ducts junction, but vascular structures were preserved. A complete resection of the common bile duct and Roux-en-Y reconstruction were done. During surgery, a frozen biopsy of the safety margins of the resection was performed, with a negative result. Pathological exam revealed tubulo-papillary structures with fine fibrovascular cores covered by biliary epithelial cells with high grade dysplasia. The result of paraffin section was positive for the left biliary margin. The final diagnosis was a intraductal tubulo-papillary adenocarcinoma with intraluminal and exophytic growth, classified as pT1N0. The disease recurrence occurred after 3 years and left hepatectomy was done. The patient died 5 years later.

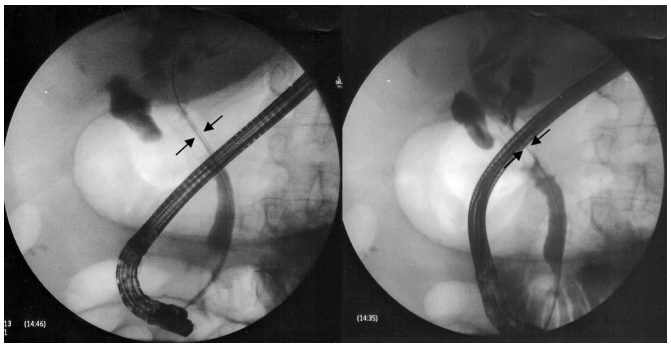

FIGURE 2. Endoscopic retrograde cholangio-pancreatography. Filling failure of the suprapancreatic portion of the common bile duct.

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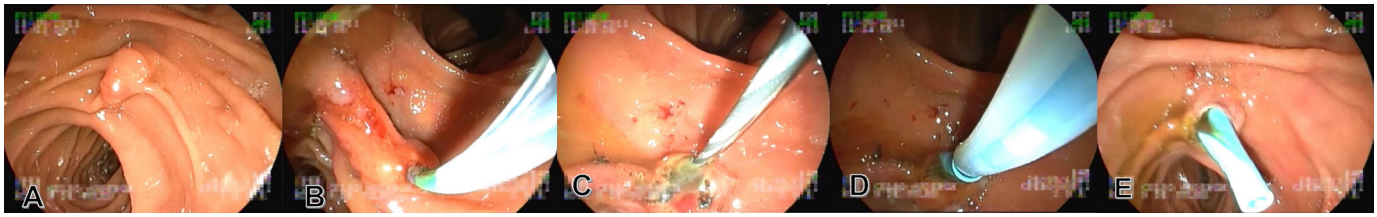

FIGURE 3. Endoscopic papillotomy. **A.** Spherical papillae with type I infundibulum. **B.** With a double-channel papillotome, the bile duct is selectively approached in a deep way. **C.** With the guide wire in place, the papillotome is removed. **D.** Passage of the 12 cm and 10 French plastic stent. **E.** Endoscopic view of the well-positioned stent.

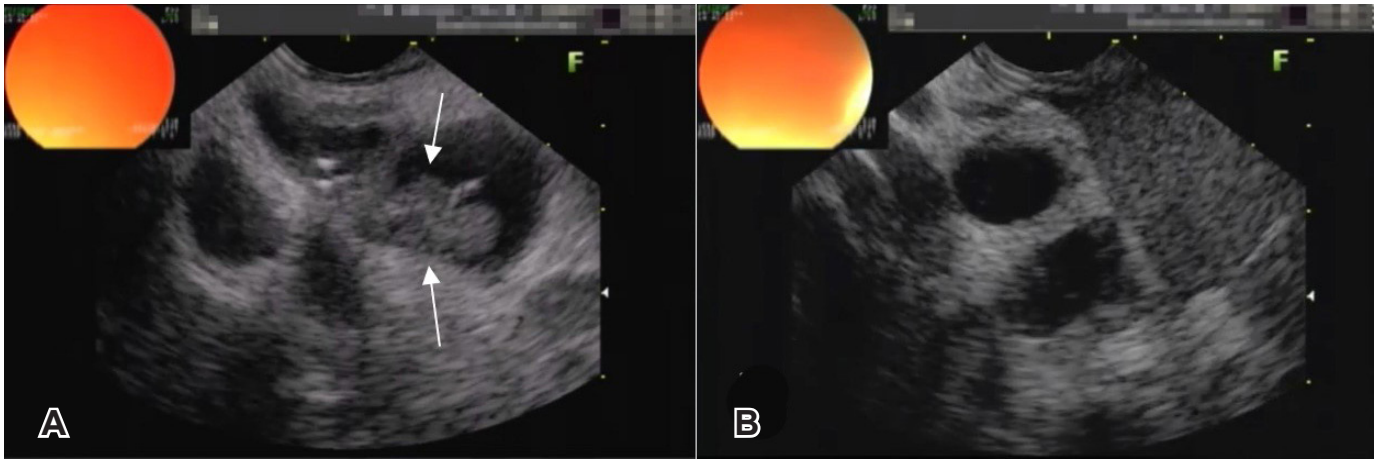

FIGURE 4. Echoendoscopy. **A.** Image shows the lesion that extends from the intrapancreatic common bile duct to the confluence of the right and left hepatic ducts. **B.** The vascular structures are free.

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Palavras-chave – Endosonografia; tratamento endoscópico; trato biliar; colangiocarcinoma; neoplasia papilar; diagnóstico.

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