Papillary adenoma of the extrahepatic biliary tract – a rare cause of obstructive jaundice

O. Sotona[1], F. Čečka[1], Č. Neoral[5], A. Ferko[1,2], S. Rejchrt[3] , M. Podhola[4], Z. Šubrt[1,2], B. Jon[1]

(1) Department of Surgery, (3) Second Department of Internal Medicine, (4) Fingerland Department of Pathology, Faculty of Medicine and University Hospital Hradec Králové, Czech Republic ; (2) Department of Field Surgery, Military Health Science Faculty, Hradec Králové, Defence University Brno, Czech Republic ; (5) Department of Surgery, Faculty of Medicine and University Hospital Olomouc, Czech Republic.

Abstract

The authors present a case of papillary adenoma of the extrahepatic biliary tract presenting as obstructive jaundice. The diagnosis was based on the endoscopic retrograde cholangiopancreatography (ERCP), and above all cholangioscopy findings. The patient was treated by bile duct resection with Roux-en-Y hepaticojejunostomy. Adenoma of the bile duct is a rare entity. Only a few cases have been described in the world literature so far. (Acta gastroenterol. belg., **2010**, 73, **270-273**).

Key words : adenoma, biliary tract, obstructive jaundice, cholangioscopy.

Introduction

Benign causes of obstructive jaundice are most frequently gall stones and chronic pancreatitis. Most common malignant causes are gall-bladder cancer, carcinoma of the biliary tract and carcinoma of the pancreas. Other causes are less frequent.

Papillary or villous adenomas are benign epithelial tumours which can occur anywhere in the gastrointestinal tract. They are found mostly in the colon and rectum. In the upper gastrointestinal tract they occur less frequently and mainly in the duodenum and ampulla of Vater. Villous adenomas of the extrahepatic biliary tract are exceptional. They usually present with obstructive jaundice, abdominal pain and dyspepsia.

Case report

A 58-years old male presented with painless obstructive jaundice, which was gradually increasing. Laboratory investigations showed a total serum bilirubin of 13.8 mg/dL (normal value 0.3-1.7 mg/dL), alanine aminotransferase (ALT) of 1475 IU/mL (normal value 0-55 IU/mL), aspartate aminotransferase (AST) of 516 IU/mL (normal value 0-52 IU/mL), gamma gutamyltransferase (GGT) of 1838 IU/mL (normal value 0-125 IU/mL), alkaline phosphatase (ALP) of 548 IU/mL (normal value 47-167 IU/mL). Coagulation parameters were normal. Ultrasound scan showed intrahepatic dilatation, the extrahepatic biliary tract was not clearly visible. Endoscopic retrograde cholangiopancreatography (ERCP) showed dilatation of the biliary tract and a fixed formation in the left hepatic duct extending to the hepatic duct junction. After papillotomy, we performed brush cytology and dilatation of the stenosis. A plastic biliary stent (diameter 10 Fr, length 12 cm) was placed into the left lobar bile duct. The reason for placing the stent only to the left lobar bile duct was significant stenosis in the left lobar bile duct, with greater left-sided intrahepatic biliary dilatation. An accidental finding in the ERCP was a separate bile duct for the sixth and seventh liver segments (Fig. 1). The bilirubin decreased to 8.6 mg/dL after the ERCP. The brush cytology was negative.

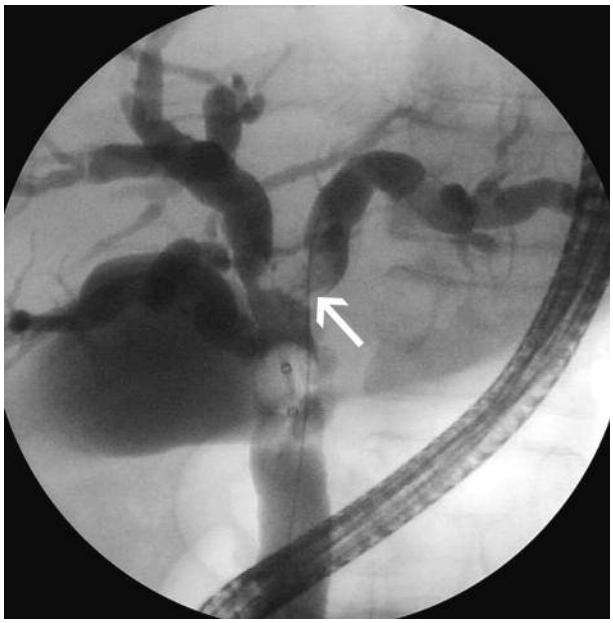

Fig. 1. — ERCP shows dilatation of both the intrahepatic and extrahepatic biliary tract. The arrow points to a fixed formation in the left hepatic duct growing to the hepatic duct junction. Accidental finding is a separate bile duct for the sixth and seventh liver lobes. The gall bladder was filled with the contrast substance through the cystic duct.

Correspondence to : Sotona Otakar, M.D., Department of Surgery, Faculty of Medicine and University Hospital Hradec Králové, Sokolská 581, 500 05 Hradec Králové, Czech Republic. E-mail : otakar.sotona@seznam.cz

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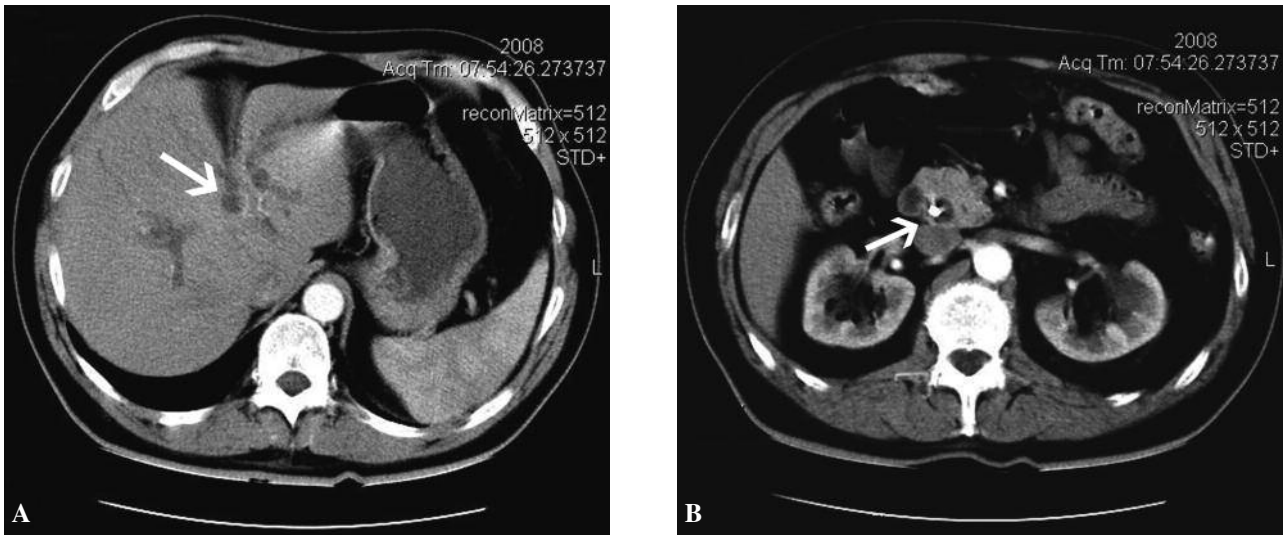

Fig. 2. — Contrast-enhanced CT scan shows dilatation of the intrahepatic bile duct (A). Dilatation of the extrahepatic bile duct is shown with the plastic stent in place (B).

Another ERCP was performed three weeks later in order to repeat brush cytology. This time two plastic stents were used, the first one (diameter 10 Fr, length 15 cm) was placed to the left lobar bile duct, and the second one (diameter 11.5 Fr, length 12 cm) was placed to the bile duct for the fifth and eighth segments. Brush cytology was again negative. The tumour marker CA 19-9 was elevated to 58 IU/mL (normal value 0-37 IU/mL). Ultrasound scan as well as CT scan showed dilatation of the intrahepatic and extrahepatic biliary tract. No gall stones were found (Fig. 2A, 2B). Neither pancreatic mass nor hepatic lesions were found.

Due to the unclear etiology of the bile duct stenosis we decided for a cholangioscopy, which was performed 3 days after the second ERCP. At that time laboratory data showed total serum bilirubin 1.2 mg/dL, ALT 105 IU/mL, AST 85 IU/mL, GGT 345 IU/mL and ALP 204 IU/mL. The cholangioscopy showed an ulcer just below the hepatic duct junction and a polypoid mass with considerable mucous secretion (Fig. 3). During cholangioscopy a biopsy was taken which showed only fragments of bile duct epithelial cells without dysplasia. Neither premalignant changes nor malignant structures were found.

Despite repeated ERCP and cholangioscopy with biopsy, we were not able to determine the histopatological characteristic of the polypoid tissue in the extrahepatic biliary tract. Due to mucin production of the tumour, elevation of CA 19-9 and weight loss of the patient, we decided for radical surgical resection at approximately two months after the initial presentation of the obstructive jaundice. After a standard retrograde cholecystectomy, we performed a longitudinal choledochotomy of the common bile duct. In the bile duct we found a considerable amount of mucinous substance. We found an ulcer at the posterior wall of the hepatic junction, and the left lobar duct was filled with a polypoid tumour. The common bile duct resection was performed. On the left side the resection line reached up to the segmental bile ducts for the second and third liver segments. A frozen section of the specimen showed low grade adenoma of the bile duct. The reconstruction was performed with Roux-en-Y hepaticojejunostomy.

The postoperative course was uneventful. The patient was discharged from the hospital on the ninth postoperative day. He made a good recovery and has been followed up in the outpatient department. The histology examination of the resected sample showed papillary adenoma with low-grade dysplasia and sporadically high-grade dysplasia (Fig. 4). One year after the surgical procedure the patient is in good general health with no complaints and no signs of recurrence of the disease.

Discussion

Benign tumours of the extrahepatic biliary tract are rarely diagnosed in clinical practise. Less than 200 cases had been published until 2000 in the English literature. Benign tumours occur in approximately 6% of all tumours of extrahepatic biliary tract (1-4). The first case was published by Saxe in 1988 (5).

From the morphological point of view we can divide benign adenomas of the biliary tract into papillary adenomas, pedunculated adenomas, and sessile adenomas (6,7). Histologically adenomas are classified into tubular adenoma, tubovillous adenoma, and villous adenoma. Thus papillary adenomas are histologically equivalent, or at least very close, to villous adenomas (8). In fact, some authors use papillary adenomas and villous adenomas interchangeably (9). Classification of these benign epithelial lesions is not uniform (10). Benign non-epithelial tumours of the biliary tract including

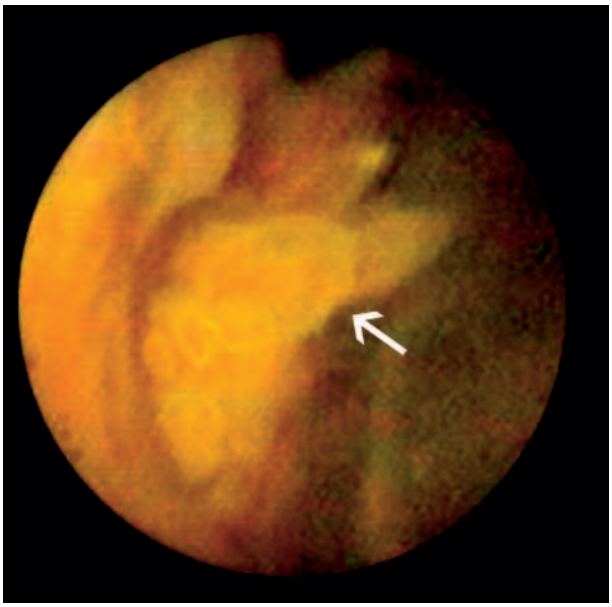

Fig. 3. — Cholangioscopic image of the bile duct adenoma

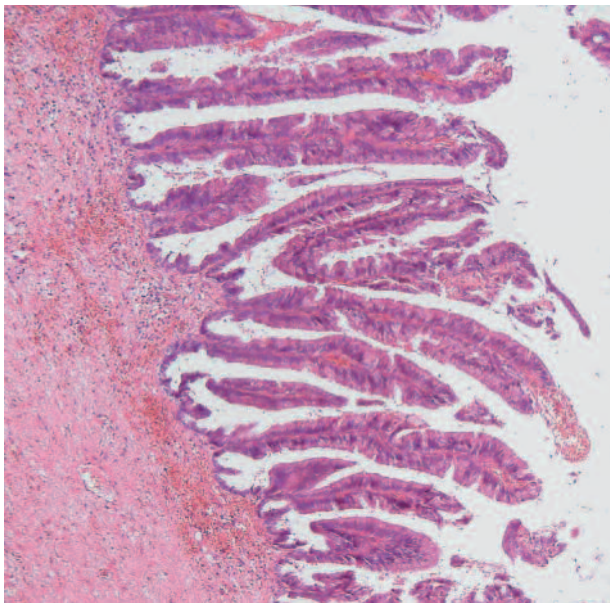

Fig. 4. — Histological examination of the surgical specimen shows the papillary tumour with cylindrical cells which resemble primary bile duct epithelium. There are no signs of invasion. Hematoxylin-eosin staining, magnification 100×.

cystadenomas, fibromas, lipomas, neuromas, myxomas and xanthomas are even more uncommon (11). The adenomas are most commonly localized in the common hepatic duct, intrahepatic lesions are rare (7). Several cases of synchronous occurrence of adenomas and congenital anomalies (e.g. choledochal cyst) have been reported (4,12).

Benign tumours of the extrahepatic biliary tract are prone to recurrence (10). A case of progression of a single benign bile duct tumour into multiple benign tumours has been reported. The initial papillary adenoma of the distal common bile duct over a 7 year period progressed to diffuse papillary adenomatosis, causing obstruction of the common bile duct (7). Even coexistence of intestinal and biliary polyps has been reported. This is the reason why some authors recommend routine imaging of the biliary tract in patients with familial adenomatous polyposis (FAP) (10).

The sequence of adenoma to adenocarcinoma in the large bowel is very well known. The large bowel adenomas are assigned various grade of dysplasia, which is used to describe structural and cytological alteration in the epithelium that predisposes an organ to cancer development. According to severity, dysplasia can be graded into mild, moderate and severe. Severe dysplasia in an adenoma is considered a selective marker for the increased risk of cancer and this is particularly true for lesions greater than 10 mm in diameter and with a marked villous component. The definition of different grades of epithelial dysplasia in adenomas is one of the most convincing pieces of evidence of the adenoma – carcinoma sequence. Most colorectal carcinomas evolve through stages of increasingly severe epithelial dysplasia before becoming invasive lesions. Many molecular abnormalities have been reported in colorectal adenomas, which include mutation of oncogenes (such as k-ras), inactivation of tumour suppressor genes (such as p53) and of mutator genes. Many of these genetics events may target the transition from normal mucosa to small adenomas and from these to large adenomas and infiltrating carcinoma (13,14). The risk of malignant change in the biliary tract has not been precisely described. It has been reported in only a few case reports. However, it is generally considered that villous adenomas of the biliary tract behave like those elsewhere in the gastrointestinal tract (2). There is one theory which says that the epithelium of the biliary tract can have the same cancerogenesis as in the large bowel (15).

Jaundice, abdominal pain and dyspepsia are the most common presenting signs of benign tumours of the biliary tract (3,4,11). Futami presented a case of adenoma of the bile duct presenting as recurrent acute pancreatitis (20). Preoperative diagnosis of these tumours is difficult (1,3,16). It is based on ERCP findings, which is able to localize the tumour in the biliary tract. It allows imaging of the biliary tract and the pancreatic duct, and preoperative biopsy removal (1,17). Another possibility is to use intraductal ultrasonography (IDUS). Current studies indicate that the presence of sessile tumour, tumour with size greater than 10 mm, and interrupted wall structure were positive factors in predicting malignancy on IDUS. When IDUS shows two of these three features, the patient should be judged as having malignancy even if the biopsy is negative. Preoperative diagnostic accuracy

can be substantially improved by a combination of IDUS and ERCP (21,22). On the other hand the drawbacks of IDUS include low depth of penetration, limited value in assessing lymph nodes and unability to provide the histopathological diagnosis (23). A cholangioscopic examination via transpapillary route is very important because bile duct tumours have characteristic cholangioscopic findings (1,18). Another beneficial method is magnetic resonance cholangiopancreatography (MRCP), although the sensitivity of this method in the diagnosis of bile duct tumours is unknown (11). MRCP has excellent overall sensitivity and specificity in demonstrating the presence and level of biliary obstruction. The major advantage of MRCP is the noninvasive nature of the procedure (23).

There are no guidelines for appropriate treatment of benign biliary lesions due to the rarity of this clinical entity. Local endoscopic excision by ERCP has been proposed, but the risk of recurrence is too high (19). However, it is a possibility for high risk patients or patients refusing surgical resection (8).

In our case the patient presented with painless obstructive jaundice. Imaging methods showed intrahepatic as well as extrahepatic biliary tract dilatation. They showed a mass located in the left hepatic duct. The precise preoperative diagnosis was not achieved in spite of repeated ERCP with brush cytology. Cholangioscopy showed a characteristic picture of bile duct adenoma, but the biopsy taken at cholangioscopy was negative. Bile duct resection with Roux-en-Y reconstruction was performed. Peroperatively we found a tumour with mucin production, which together with intraductal growth of the tumour, caused bile duct obstruction. The patient is in good general health one year after the surgical procedure, with no signs of recurrence of the disease.

In conclusion, benign tumours of the biliary tree are rare. The diagnosis of these tumours preoperatively is often difficult and cholangioscopy offers a great advantage. They are considered premalignant lesions with a definite risk of recurrence and risk of progression to cholangiocarcinoma if left untreated. Surgical resection with tumour-free margins should therefore be performed.

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