

Benign Parapapillary Choledochoduodenal Fistula Associated with Ampullary Carcinoma: Case Report and Literature Review

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Abstract

Parapapillary and periampullary choledochoduodenal fistulas (CDF) are rare conditions. We here report a case of benign parapapillary choledochoduodenal fistula associated with early ampullary carcinoma (pT₁NoMo). A 54-year-old Jordanian man had recurrent cholangitis without clinical jaundice. He had marked elevation of the alkaline phosphatase (AP) and Gamma Glutamyl Transferase (GTT). Abdominal ultrasonography showed distension of the gallbladder with dilatation of the common bile duct (CBD). Duodenoscopy showed a swollen ampulla with no intraduodenal growth. Cannulation the orifice of the papilla of Vater failed. But endoscopic biopsy revealed adenocarcinoma. Magnetic resonance cholangiopancreatography showed a dilated bile duct with a filling defect in its most distal part. With the diagnosis of ampullary carcinoma, the patient underwent pylorus-preserving pancreaticoduodenectomy (PPPD). The gross and microscopic examination of the resected specimen showed a dilated CBD, a small-sized benign choledochoduodenal fistula to the CBD above the tumor, and a non-dilated pancreatic duct that opened above the tumor. The small-sized fistula was missed by all used diagnostic procedures. This communication presents a unique case of association between ampullary carcinoma with benign small sized parapapillary CDF that caused cholangitis with marked elevation of AP and GTT but without clinical jaundice. The benign CDF may cause cholangitis and together with the high bilio-pancreatic junction may increase the risk of biliary cancer.

Keywords: Choledochoduodenal Fistula, Ampullary Carcinoma.

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Introduction

Ampullary carcinoma accounts for less than 7 percent of the neoplasms that obstruct the distal bile duct⁽¹⁾. The presenting symptom is most often fluctuating jaundice without cholangitis⁽²⁾. Very rarely periampullary or parapapillary choledochoduodenal fistula occurs concurrently with ampullary carcinoma.

Here, we report a patient with non infiltrating papillary adenocarcinoma of the ampulla of Vater that occurred concurrently with a benign parapapillary choledochoduodenal fistula. The patient presented with recurrent cholangitis. Jaundice was not seen throughout the entire clinical course. We discuss the relationships among these conditions in patients with ampullary carcinoma.

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Case Report:

A 64-year-old Jordanian visited Jordan University Hospital (JUH) complaining of high-grade intermittent fever of 40 days duration. The fever was associated with rigors, chills and profuse sweating. He had no pain, loss of appetite or weight loss. He noticed that his urine got darker during the attacks. He has been under treatment for gout, ischaemic heart disease and essential hypertension. His physical examination revealed a hepatosplenomegally but wasn't jaundice. His laboratory data showed normal leukocytic count (10.400), high

erythrocytic sedimentation rate (90mm/h), normal total and direct bilirubin, elevated serum AP (670 iu/l), and elevated GGT (1112 iu/l). Screening for viral hepatitis was negative.

Abdominal ultrasonography showed distended gallbladder and dilatation of the intra and extrahepatic biliary ducts but no cholelithiasis. Abdominal CT scan showed a well-defined hypodense lesion in the region of the head of the pancreas with dilatation of the common bile duct, distension of the gallbladder, hepatosplenomegally, and intrahepatic biliary dilatation (Figure 1).

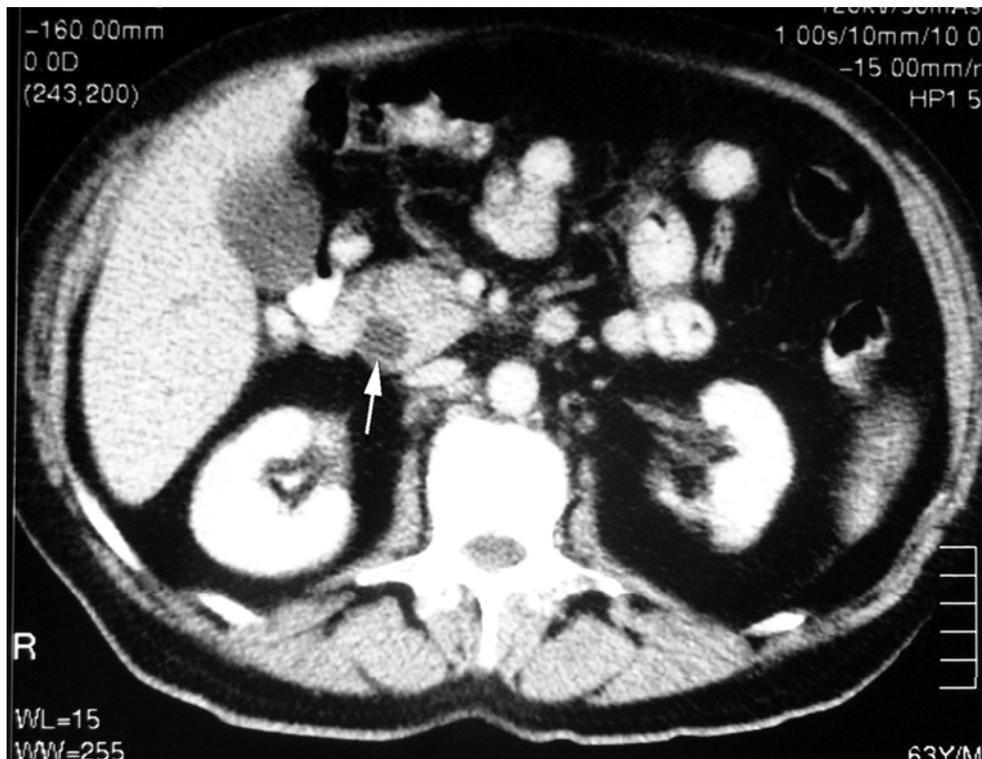


Figure 1: Abdominal computed tomography (CT) showed a hypodense lesion in the region of the head of the pancreas and a dilated intra and extrahepatic bile ducts

Magnetic resonant cholangio-pancreatography (MRCP) revealed that the bile duct is dilated with a filling defect in its most distal part (Figure 2). The filling defect was thought to be a stone versus a tumour. Endoscopic retrograde cholangio-pancreatography (ERCP) showed a

swollen ampulla of Vater with no intraduodenal growth. Its cannulation failed. However, endoscopic biopsies of the ampulla showed adenocarcinoma.

Tumor markers, including carcnoembryogenic antigen, the carbohydrate

antigens 19.9, 15.3, 125 were all within normal range.

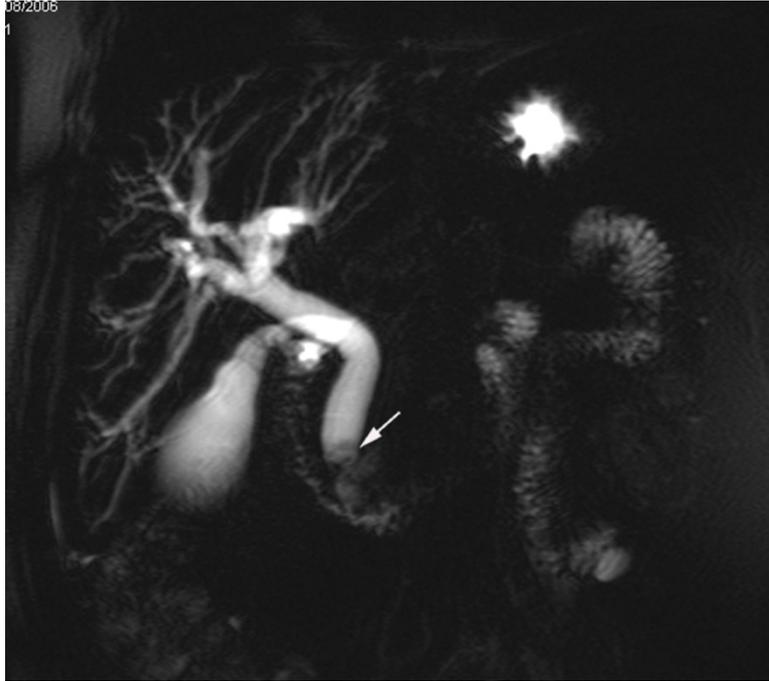


Figure 2: MRCP showed a filling defect in the most distal part of the CBD

After cardiac evaluation and with the diagnosis of ampullary carcinoma, pylorus-preserving pancreatico-duodenectomy (PPPD) was performed. Macroscopically, the ampulla was obstructed by a tumour that extended into

the lower part of the common bile duct. A fistula tract measuring 1.0 x 0.2 x 0.2 cm was seen just above the ampulla connecting the bile duct to the duodenum (Figure 3).

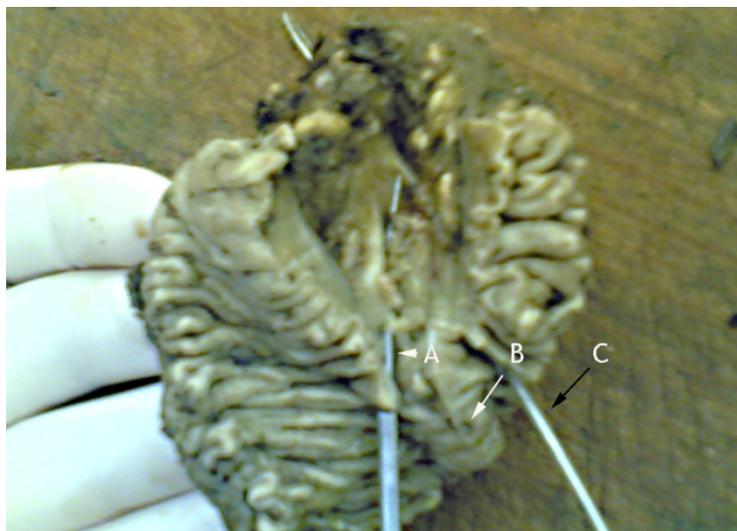


Figure 3: Resected specimen, showing (B) swollen papilla with no intraduodenal growth (A) choledochoduodenal fistula marked by probe and (C) pancreatic duct marked by a probe

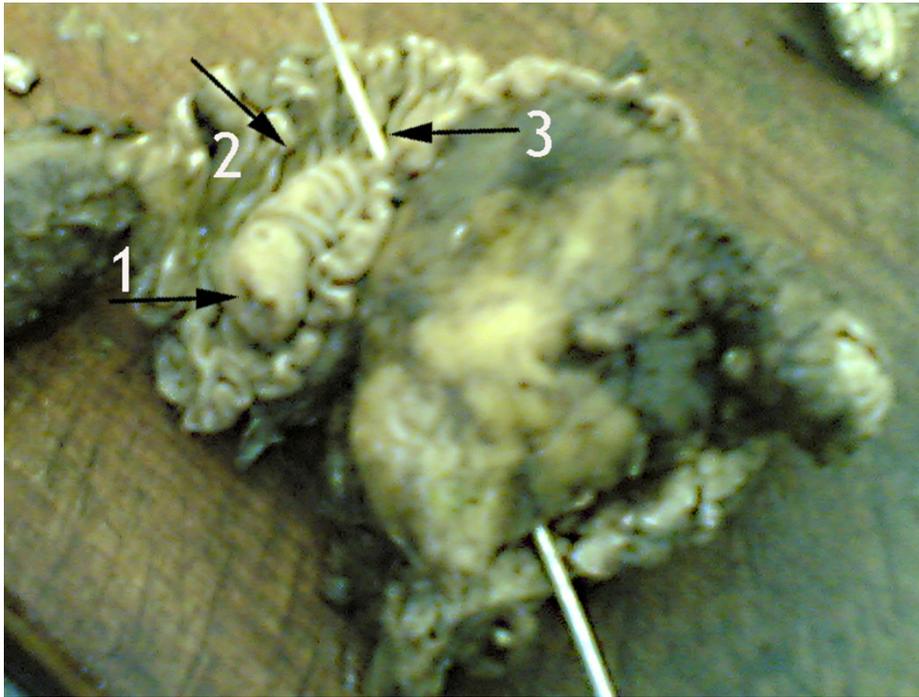


Figure 4: Resected specimen showing (1) swollen papilla (2) choledochoduodenal fistula (3) pancreatic duct joining the common hepatic duct above the tumor

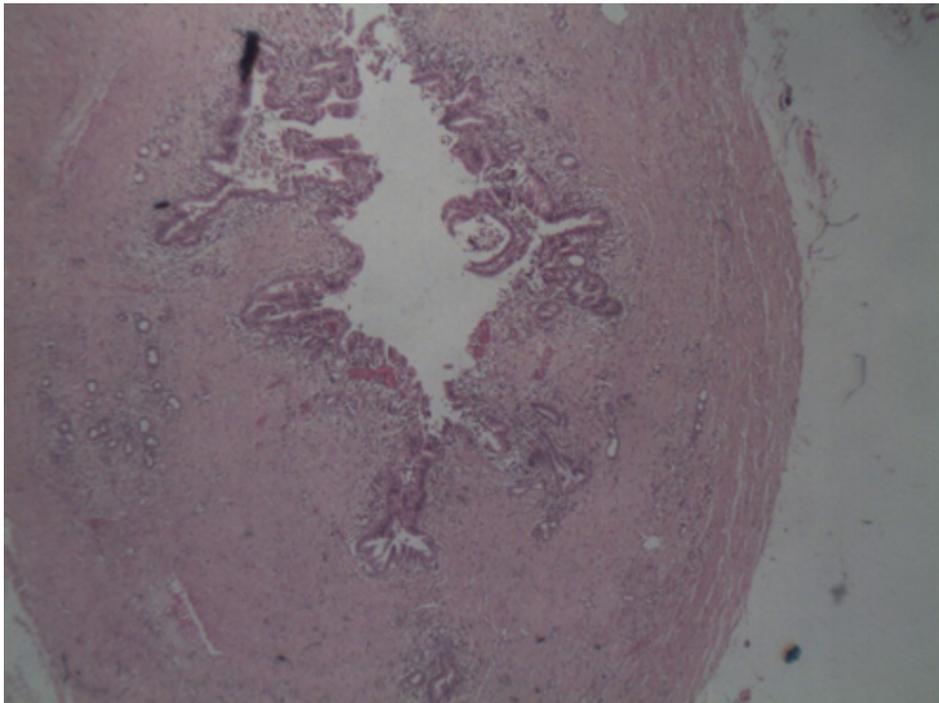


Figure 5: The slide shows a cross section through the fistula tract with a thick fibromuscular wall and a lining of epithelial cells forming villous and glandular structures (100X)

The ampullary tumour had papillary configuration and measuring 2.0 x 1.0 x 0.2 cm. The histopathological diagnosis was a moderately differentiated adenocarcinoma of the ampulla, pT₁NOMx. The pancreatic duct was not dilated and it did not join the bile duct above the ampullary tumour (Figure 4). Random sections from the fistula tract were free of malignancy (figure 5). Thus, the small-sized CDF was benign and was missed by all investigations carried out.

The patient's postoperative course was uneventful except for transient urine retention due to benign prostatic enlargement, and he was discharged on postoperative day 14.

Discussion:

Choledochoduodenal fistula (CDF) is a relatively rare condition that shows an abnormal communication between the biliary tract and the duodenum. It represents a complication of choledocholithiasis⁽³⁻⁶⁾, peptic ulcer disease⁽⁷⁻⁹⁾, prior surgery⁽¹⁰⁻¹¹⁾, and duodenal tuberculosis⁽¹²⁻¹³⁾. Rarely CDF occur concurrently with periampullary carcinoma.^(2, 4, 14-17) Also, it has been suggested that parapapillary choledochal fistula may be a risk factor for development of biliary tract carcinoma.⁽¹⁸⁻¹⁹⁾ When CDF is present in a Patient with ampullary carcinoma, the fistula itself can be benign as in our patient or it can be malignant as been reported by others^(17,18). our patient with ampullary carcinoma concurrently had benign choledochoduodenal fistula.

The presenting symptom of adenocarcinoma of the ampulla of Vater is most often jaundice. However, ampullary cancer can present without jaundice⁽²⁰⁾. Fluctuation of jaundice has been reported to be characteristic of ampullary carcinoma, and it is attributed to intermittent obstruction by a

movable tumour, transient disappearance of edema in the ampulla of Vater, or ulceration of ampullary tumour releasing the obstruction of the biliary tree. Hirata S, et al.⁽¹⁴⁾ reported that obstructive jaundice, cholangitis, and abdominal pain had subsided spontaneously within few days due to formation of a choledochoduodenal fistula at the base of an ulcerating ampullary tumours. Imaeda K, et al.⁽¹⁵⁾ reported a patient with multiple parapapillary choledochal fistulas with ampullary carcinoma. Jaundice was not seen throughout the entire clinical course of their patient. He postulated that the fistulas played a major role in bile drainage and had prevented development of obstructive jaundice. In both reports, the fistulas were malignant in nature. Our patient presented with episodes of cholangitis for 40 days but without clinical jaundice. His AP and GGT were markedly elevated. His fistula was proven to be of benign nature; but because of its small-size, it was missed on duodenoscopy and on MRCP. Also, it did not open into the bile duct proximal to ampullary carcinoma. The bile duct was grossly dilated. However, the pancreatic duct was not dilated and did not join the common hepatic duct high above the ampullary tumor, indicating a high bilio-pancreatic junction. Though the ampullary tumor was diagnosed at an early stage (pT₁NoMo), it totally occluded the ampulla leading into failure of its cannulation during attempted ERCP. Our patient had no biliary stones and no history of peptic ulcer disease, tuberculosis, or prior surgery. We postulated that the benign fistula formation had drained the bile duct partially and thus prevented appearance of clinical jaundice. Also, we postulated that the fistula together with the ampullary obstruction were risk factors for development of recurrent cholangitis. We share the view of others⁽²¹⁻²³⁾,

that both the high bilio-pancreatic junction and reflux of intestinal juice into the biliary tree may be a risk factor for development of biliary and/or ampullary carcinoma.

Benign parapillary and periampullary choledochoduodenal fistulas can occur in patients with ampullary carcinoma. The fistula can prevent the development of clinical jaundice, but may increase the risk of development of cholangitis and biliary tract cancer. The best investigation to detect the presence of CDF is the ERCP provided that

the procedure is carried out by an experienced endoscopist who had it in mind. MRCP and CT scanning are needed to evaluate changes in the biliary cyst in the CDF patients such as dilatation, stone formation, and others. The fistula can be too small and will be missed on duodenoscopy and MRCP. Therefore, when ampullary carcinoma has produced cholangitis without clinical jaundice, the presence of parapapillary or periampullary choledochal fistula should be considered and searched for.

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ناسور القناة الصفراوية الإثني عشرية حول انبور فاتر مصاحبا لسرطانة انبور فاتر

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الملخص

يعد ناسور القناة الصفراوية الإثني عشرية الواقع حول انبور فاتر وحوالي الحليمة من الحالات النادرة. ونقدم هنا تسجيلاً لحالة ناسور القناة الصفراوية الإثني عشرية الحميد المصاحب لسرطانة انبورية مبكرة (pT1N0M0). رجل أردني عمره 54 عاماً، كان لديه إلتهاب القنوات الصفراوية الراجع بدون يرقان سريري. وكان لديه ارتفاع عالٍ للفسفوتاز القلوية ولناقلة الجاما قلو تامايل. وقد أظهر تخطيط الصدى البطني وجود انتفاخ في المرارة مع توسع في قناة الصفراء الجامعة. وأظهر تنظير الإثني عشر وجود انبور منتفخ ولكن بدون نمو داخل الإثني عشر. ولقد فشل اقناء حليمة فاتر ولكن الخزعة بواسطة المنظار أظهرت وجود سرطانة غدية. وبين تصوير القنوات الصفراوية بالرنين المغناطيسي توسع القنوات الصفراوية مع وجود امتلاء معيب في أسفل القناة الصفراوية الجامعة. وبعد الوصول لتشخيص السرطانة الغدية، أجريت للمريض عملية القطع البنكرياسي الإثني عشرية مع الإبقاء على البواب. وأظهر الفحص العياني والمجهري للعينة المستتصلة وجود قناة صفراوية جامعة متوسعة مع ناسور صغير الحجم وحميد ما بين القناة الصفراوية الجامعة والإثني عشر، واقع إلى الأعلى من الورم. وأما قناة البنكرياس فلم تكن متوسعة ما فوق الورم. وبهذا كان الناسور صغير الحجم فائتاً بالرغم من استعمال جميع الوسائل التشخيصية المتاحة. ويقدم هذا التقرير حالة فريدة لتصاحب بين السرطانة الانبورية لفاتر مع ناسور حميد حول الحليمة ما بين القناة الصفراوية الجامعة والإثني عشر وقد تسبب هذا بارتفاع عالٍ للفسفوتاز القلوية وناقلة الجاما قلو تامايل وبدون يرقان سريري. إن هذا النوع من الناسور قد يسبب إلتهاب القنوات المرارية ويتكونه مع وجود التقاء عالٍ ما بين قناة الصفراء وقناة البنكرياس فقد يزيد من احتمال الإصابة بالسرطانة المرارية.

الكلمات الدالة: ناسور القناة الصفراوية، سرطانة أنبور فاتر.