



A rare case of periampullary carcinoma with ectopic ending of Vater's ampulla

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Abstract

A 71-year-old woman was referred to our department complaining of painless progressive jaundice for the last 3 mo. Magnetic resonance imaging and magnetic resonance cholangiopancreatography (MRCP) showed the ectopic hepatopancreatic ampulla draining into the fourth part of the duodenum adjacent to the duodenojejunal flexure; the irregular morphology of the duodenojejunal flexure likely due to a soft tissue mass. Laparotomy confirmed the presence of the abnormal ampulla of Vater located at the fourth part of the duodenum and a soft tissue tumor about 6 cm × 5 cm × 5 cm with a peduncle adjoining the ampulla. Resection of the tumor, including some peripheral tissue, and a Roux-Y loop anastomosis choledochojejunostomy were performed. Pathological examination indicated an intestinal villous adenoma accompanied by severe dysplasia and focal canceration. Periampullary carcinoma with ectopic ending of the Vater's ampulla into the fourth part of the duodenum is rather rare. The embryonic genetic background of this anomaly has not yet been fully explained. It is worth mentioning that MRCP is useful for demonstrating anomalies and anatomic variants of the biliary tract system and pancreatic duct.

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INTRODUCTION

The usual location of the major papilla of Vater is in the mid-portion of the descending duodenum, closer to the posterior wall than the anterior wall. On rare occasions it may be found in other sites, along a line extending from the stomach to the fourth part of the duodenum^[1-5]. Here we present a case of periampullary carcinoma combined with ectopic opening of the Vater's ampulla into the fourth part of the duodenum and a low confluence of the cystic duct into the common bile duct. To our knowledge, such a case has never been described previously, so the pathogenesis, diagnosis and treatment will be discussed in this report.

CASE REPORT

A 71-year-old woman was referred to our department complaining of painless progressive jaundice for the last 3 mo. Hypertension and diabetes mellitus were diagnosed 10 years previously, and a cholecystectomy was performed 18 years before admission. Physical examination did not show any abnormalities except jaundice. Laboratory data (Table 1) showed the levels of the serum total bilirubin, direct bilirubin, alanine aminotransferase, aspartate aminotransferase, alkaline phosphatase, and glutamyl transpeptidase were 138.5 μmol/L, 120.0 μmol/L, 78 IU/L, 184 IU/L, 1031 IU/L, and 944 IU/L, respectively (significantly higher than normal); tumor markers including serum α-fetoprotein, carcinoembryonic antigen, carbohydrate antigen 199 (CA199), and carbohydrate antigen 125 (CA125) were 1.44 ng/mL, 3.27 ng/mL, 34.54 U/mL, and 89.87 U/mL, respectively (the latter two were a little higher than normal); the hepatitis B & C markers were negative; blood routine test (BRT) was normal. Sonography of the upper abdomen revealed only hepatic biliary and main pancreatic duct dilation. Subsequently magnetic resonance imaging and magnetic resonance cholangiopancreatography

Table 1 Some lab tests of patient

Lab items	Values		Normal values
TBIL ($\mu\text{mol/L}$)	138.5	↑	1.7-17
DBIL ($\mu\text{mol/L}$)	120.0	↑	0-6.8
ALT (IU/L)	78	↑	10-40
AST (IU/L)	184	↑	10-40
ALP (IU/L)	1031	↑	40-110
GT (IU/L)	944	↑	< 50
AFP (ng/mL)	1.44		< 20
CEA (ng/mL)	3.27		< 15
CA199 (U/mL)	34.54	↑	< 33
CA125 (U/mL)	89.87	↑	0-35

TBIL: Total bilirubin; DBIL: Direct bilirubin; ALT: Alanine aminotransferase; AST: Aspartate aminotransferase; ALP: Alkaline phosphatase; GT: Glutamyl transpeptidase; AFP: α -fetoprotein; CEA: Carcinoembryonic antigen.



Figure 1 Magnetic resonance imaging showed the expansion of hepatic biliary duct and the ectopic hepatopancreatic ampulla (arrow) draining into the fourth part of the duodenum.

(MRCP) were performed (Figure 1) which showed the expansion of the extrahepatic and intrahepatic biliary ducts and the main pancreatic duct, the ectopic hepatopancreatic ampulla draining into the fourth part of the duodenum adjacent to the duodenojejunal flexure, a low confluence of the cystic duct into the common bile duct with a distance of approx 4 cm to the Vater's ampulla, and the irregular morphology of the duodenojejunal flexure; likely due to a soft tissue mass, which did not rule out the possibility of periampullary tumor resulting in the common bile duct stricture. Gastroscopy revealed chronic superficial antral inflammation without orifice or mucosal ulceration in the stomach and without any duodenal papilla in the descending duodenum.

Laparotomy confirmed the presence of an abnormal ampulla of Vater located at the fourth part of the duodenum and a soft tissue tumor about 6 cm \times 5 cm \times 5 cm with a peduncle joined to the ampulla (Figure 2). Intraoperative choledochoscopic examination displayed no mass in the common bile duct. Consequently, complete resection of the tumor including some peripheral tissue was performed from an incision of the jejunum, close to the duodenojejunal flexure. Frozen pathological examination indicated an intestinal villous adenoma accompanied by severe dysplasia and highly suspect focal canceration, however, no incisional margin tumor cells were seen. A Roux-Y loop anastomosis

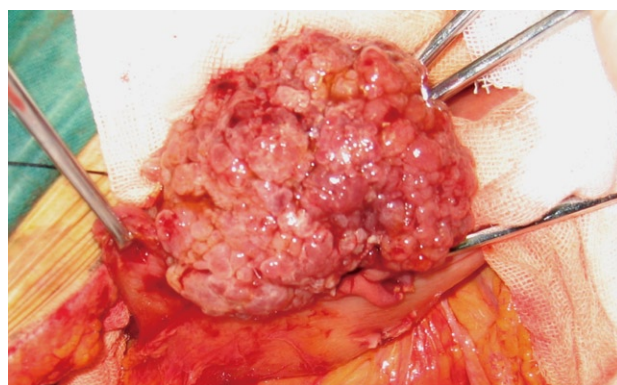


Figure 2 A soft texture tumor about 6 cm \times 5 cm \times 5 cm with a peduncle conjunct to the ampulla.

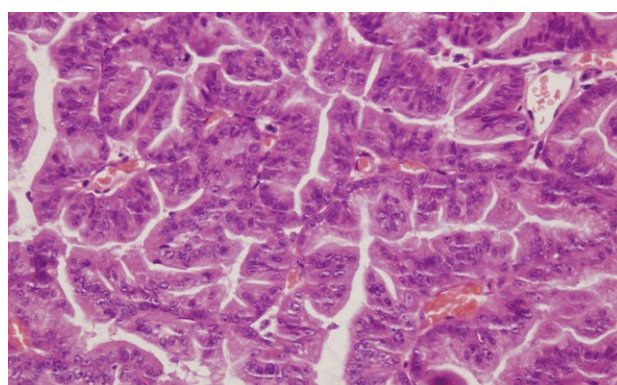


Figure 3 Microscopic examination (HE, \times 400) showed severe dysplasia and focal carcinomatous change.

at side-to-side choledochojejunostomy and end-to-side jejunojunctionostomy was carried out in order to prevent stricture at the end of the common bile duct. Pancreatoduodenectomy was not done. Paraffin block histopathologic examination confirmed a diagnosis of intestinal villous adenoma accompanied by severe dysplasia and focal carcinomatous change without incisional margin infiltration (Figure 3). After operation, the patient recovered without incident and liver function tests gradually reverted to normal levels.

DISCUSSION

The ectopic drainage of the biliary tract is a rare congenital anomaly, which consists of abnormal communication of the common bile duct, the cystic duct, or an intrahepatic duct with the gastrointestinal tract. As noted by previous authors, ectopic drainage of the common bile duct may be located along a line extending from the stomach to the fourth part of the duodenum^[1-5]. The frequency of this anomaly fluctuates between 0% and 13%^[1,6]. However, there only four cases with ectopic ampulla draining into the fourth part of duodenum were reported^[5,7]. Periampullary carcinoma combined with this anomaly has never described previously.

The embryogenetic background of this anomaly has not yet been fully explained. As is known, the extrahepatic biliary duct and the ventral pancreas arise

from the hepatic diverticulum of the end of foregut during the 4th to 6th week of embryo life^[8,9]. The dorsal pancreatic bud appears opposite the hepatic diverticulum. During development, as the duodenum rotates to the right and becomes C-shaped, the ventral pancreatic bud is carried dorsally with the bile duct. The ventral pancreatic duct joins the distal portion of the dorsal pancreatic duct to form the main pancreatic duct, which merges with the common bile duct, inserting into the duodenum *via* the papilla of Vater^[10]. Then, the proximal portion of the dorsal pancreatic duct gradually regresses and becomes obliterated at the junction with the ventral pancreatic duct. By the end of the 7th week, the proximal portion of the dorsal pancreatic duct may disappear or persist as the accessory pancreatic duct^[11,12]. Thus, we can make the hypothesis that the ampulla of Vater may be found in the sites along a line extending from the stomach to the fourth part of the duodenum, or in another words, along the end of the foregut. However, other factors may play a role in this process.

It is worth mentioning that MRCP is useful for demonstrating anomalies and anatomic variants of the biliary tract system and pancreatic duct^[13,14]. In our case, MRCP was of great value not only for depicting the ectopic common bile duct and pancreatic duct draining into the fourth part of duodenum but also for depicting the abnormal junction of the cystic duct and intraluminal small bowel lesion. According to Ruge's^[15] categorization, this variation of lower confluence of the cystic duct belongs to type II - the double-barreled type in which the cystic duct follows the hepatic duct for some distance before entering it. In addition to MRCP, gastroscopy should be used to assess the gastric mucosa and other malformations which could possibly exist.

Periampullary carcinoma includes cancer of the terminal end of the common bile duct, the ampulla of Vater and its adjacent duodenum. These lesions are similar to each other in many aspects, such as their clinical manifestations, changes noted by laboratory determinations and approach and technique of surgical excision. The prognosis is much better than for carcinoma of the head of pancreas. Endoscopic ampullectomy may be considered as a viable procedure in patients with small ampullary tumors who are unfit for surgery or who prefer a nonsurgical approach^[16-18]. But in our case, on the account of the ectopic hepatopancreatic ampulla and a likely major periampullary tumor located in the duodenojejunal flexure, endoscopic ampullectomy was unfit. So local excision, combined with frozen section, is a low morbidity and a valid alternative to pancreaticoduodenectomy^[19]. However, a choledochojejunostomy was carried out in order to prevent stricture of the lower end of the common bile duct after operation.

In conclusion, periampullary carcinoma with ectopic ending of the Vater's ampulla in the fourth part of the duodenum is rather rare. Although pathogenesis is not quite clear, accurate diagnosis and appropriate investigations are necessary for surgeons before operation. Use of MRCP is recommended for the diagnosis and

exclusion of associated anomalies of the biliary and pancreatic ducts.

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