

Local Excision of Intraductal Papillary Mucinous Neoplasm of the Papilla of Vater

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Citation

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Abstract

Tumors of the ampulla of Vater are rare histopathological conditions. These tumors vary between 0.063 and 0.21% on autopsy investigations but the frequency of their diagnosis is increasing because of the extensive use of flexible endoscopy. A 55-year-old man presented with a history of abdominal discomfort, loss of appetite and nausea for 6 months' duration. An upper gastrointestinal series showed narrowing of pylorus. Upper abdominal ultrasound showed dilated intrahepatic and extrahepatic bile ducts and a distended gallbladder. Magnetic resonance cholangiogram showed dilatation of the biliary tree and obstruction at the level of the ampulla. The patient underwent surgery and local resection of ampullary neoplasm was performed. The histopathological examination of the mass of the ampulla of Vater demonstrated intraductal papillary mucinous neoplasm. The patient's postoperative course was uneventful and he was discharged at the 12th day of operation. The patient had a follow-up for 2 years and he is asymptomatic since surgery.

INTRODUCTION

The trends between surgeons in the last decade are to perform less invasive operations with adequate results in surgical oncology. This inclination leads surgeons to minimal invasive surgery techniques. The same interest arises in pancreaticobiliary disorders. Tumors of the ampulla of Vater are rare histopathological conditions and have relatively good prognosis after resection. Neoplastic lesions of the ampulla vary between 0.063 and 0.21% on autopsy investigations but the frequency of their diagnosis is increasing because of the extensive use of flexible endoscopy (1). The treatment modalities differ from open or endoscopic local resection to pancreaticoduodenectomies. Since Halsted (2) described the first local resection of ampullary lesions in 1899, the authors still debate on adequate surgery in this disease. We report a case of mucin-secreting biliary neoplasm of the ampulla of Vater diagnosed peroperatively because of unsuccessful endoscopy due to pyloric stenosis, and successfully treated with transduodenal local excision.

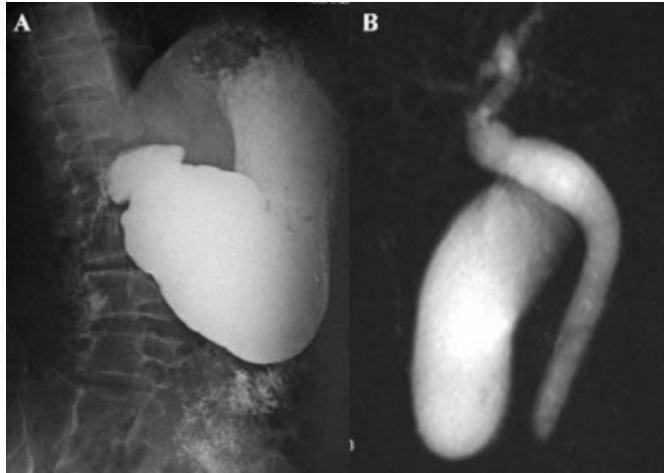
CASE REPORT

A 55-year-old man presented with a history of abdominal discomfort, loss of appetite and nausea for 6 months' duration. Physical examination at the time of admission was unremarkable except for moderate tenderness without any

rebound or peritoneal signs. He was afebrile, and his vital signs were stable. He had no history of trauma, alcohol addiction, drugs or biliary lithiasis. The laboratory results were unremarkable, except for a serum bilirubin of 2.2 mg/dL (normal range: 0.3 to 1.2 mg/dL). The tumor markers were within normal range. An upper gastrointestinal (GI) series was performed. Contrast duodenography showed narrowing of pylorus (Figure 1).

Figure 2

Figure 2: (A) Papillary lesion with abnormally dispersed glandular structures (HEX50), (B) Dysplasia of the epithelium with extended, pseudostratified hyperchromatic nuclei of crowdy epithelial cells with mucin depletion (HEX200).



Upper gastrointestinal endoscopy failed to examine the second part of the duodenum and the ampulla of Vater due to pyloric stenosis. Upper abdominal ultrasound (US) showed dilated intrahepatic and extrahepatic bile ducts and a distended gallbladder. Magnetic resonance (MRI) cholangiogram showed dilatation of the biliary tree and obstruction at the level of the ampulla (Figure 1). Computed Tomography (CT) revealed intrahepatic and extrahepatic bile duct dilation. CT investigation did not show any space-occupying mass in pancreas or duodenum. In view of the patient's clinical and radiological data, an operation was planned for the patient. During surgery the common bile duct and choledochus were found to be dilated. Kocher maneuver was then performed. A mass at the lower part of the choledochus was palpated through the duodenal wall. There was no invasion to the portal vein, hepatic artery or surrounding structures. Lateral duodenotomy was then performed to the opposite side of the mass. Stay sutures were placed in the duodenal wall, and the ampulla was identified. A circumferential resection of duodenal mucosa and ampulla of Vater was undertaken. Frozen section examination (FSE) revealed free tumor margins and absence of carcinoma. After local resection, the bile and pancreatic ducts were reconstructed. The duodenotomy was then closed transversely. Cholecystectomy was added to the operation. After surgery, the histopathological examination of the mass of the ampulla of Vater demonstrated an intraductal papillary mucinous neoplasm (Figure 2).

{image:2}

The patient's postoperative course was uneventful and he was discharged on the 12th day after the operation. The patient had a follow-up for 2 years and he is asymptomatic since surgery.

CONCLUSION

The ampulla of Vater is a cylindrical-shape structure consisting of three types of epithelia; the epithelium of the bile duct, of the pancreatic duct, and the duodenal mucosa, which was first described by a German anatomist Abrahamus Vater in 1720 (3). Benign tumors of the ampulla of Vater are lipomas, hamartomas, lymphangiomas, hemangiomas, leiomyofibromas, neurofibromas, villous and tubulovillous adenomas (3,4). Papillary neoplasms of the bile ducts are rare tumors and can be located in the intra- or extrahepatic portion of bile tree (4,5). Kim et al. first used the term of intraductal papillary mucinous neoplasm (IPMN) for nine cases of mucin-hypersecreting bile duct tumors in 2000 (6). The clinical findings of mucin-secreting bile duct tumors have a lot of similarities to those of pancreatic intraductal papillary-mucinous neoplasms, but little data is available because of the rarity of these tumors. Radiological investigations such as US, CT, and MRI are helpful techniques to examine these tumors but those techniques are unable to reveal small tumors. Dilatation of the common bile duct and main pancreatic duct and obstruction at the level of the ampulla are the main radiological findings with these methods. In the current case, US, CT and MRI were unable to detect the lesion but bile duct dilatation and cut-off sign at the level of the ampulla were revealed. In case of excessive mucin production, CT, US or MRI can detect dilatation of the intra- and extrahepatic bile ducts (3,7). Endoscopic ultrasound (EUS) and endoscopic retrograde cholangiopancreatography (ERCP) are the most useful techniques. ERCP shows the dilated ducts and filling defects. Endoscopic appearance of a mucin-secreting orifice is a specific finding of papillary mucinous tumor. Histopathological examinations with brushing and biopsies are available with this technique but they are thought to miss the diagnosis in up to 60% of cases (11). EUS is one of the most important modalities in detecting the structures of the periampullary region and can accurately diagnose the presence of tumors of this region (12). EUS and ERCP could not be performed in our case due to pyloric stenosis.

The clinical presentations of patients with this tumor are abdominal pain, jaundice, bleeding, and biliary sepsis due to tumor obstruction. Authors still debate on even the treatment of malign tumors of the ampulla of Vater; thus, the surgical

intervention for intraductal papillary mucinous neoplasm still remains unclear. The most accepted operative modalities are pancreaticoduodenectomy (PD), transduodenal excision (TDE), and endoscopic snare excision (ESE). PD is the most aggressive approach which has a mortality of 3%-9% in high volume and experienced centers (7,8,13). Five years survival after PD varies from 21% to 60% in published data (13-15). TDE and ESE are organ-preserving procedures. The indications of TDE are benign tumors smaller than 3-4cm, adenomas of the papilla with high-grade dysplasia and tubular, villous or tubulovillous adenomas of the papilla (3,16). Beger et al. (17) defined the method of TDE and reported zero operative mortality. The touchstone in TDE is to obtain adequate tumor-free margins with FSE during operation in order to prevent recurrence (18,19). ESE is an evolving method among surgeons. As in TDE, ESE has complications such as bleeding, cholangitis, and pancreatitis. The mortality rates were reported zero in ESE with recurrence rates up to 25% for benign papillary tumors (20, 21).

In conclusion, intraductal papillary mucinous neoplasms of bile duct are rare, benign tumors. In selected cases, TDE or ESE can be the most satisfactory method with low morbidity and mortality.

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