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Adenomyoma of Ampulla of Vater or the Common Bile Duct
– A Report of Three Cases –

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Adenomyoma is a rare non-neoplastic lesion of the biliary tract. Here we report on three cases of adenomyoma; one located in the ampulla of Vater and two located in the common bile duct. Although preoperative endoscopic and radiological evaluations could not determine whether lesions were benign or malignant, intra-operative frozen section histologic examinations aided the differential diagnosis. Microscopic features of a lobular gland architecture with basally located nuclei and the absence of desmoplastic stromal reaction were found to be characteristic in frozen and paraffin sections.

Key Words : Adenomyoma; Ampulla of Vater; Common bile duct

Adenomyomas of the extrahepatic bile ducts, including the ampulla of Vater are rare. This disease is most frequently located in the gallbladder, and is also referred to as adenomyomatous hyperplasia or adenomyomatosis.[1] Because of its location, adenomyoma of the ampulla of Vater or of the extrahepatic biliary tree characteristically present clinical signs of obstructive jaundice and mimic malignant neoplasms.[2-4] Approximately 20 cases of adenomyoma of the ampulla of Vater or extrahepatic bile duct have been reported in the literature.[1-8] The most effective methods of treatment are reported to be endoscopic ampullectomy and surgical resection. However, no useful diagnostic methods, other than histologic examinations, that can distinguish between an adenomyoma and a malignancy are known. Here, we report the pathologic findings of a single ampullary adenomyoma and two cases of adenomyoma in the distal common bile duct because of the rarity of this disease and the diagnostic pitfalls involved.

CASE REPORT

Case 1

A 56-year-old man with a 9-year history of noninsulin-dependent diabetes was admitted with an uncontrolled blood glucose level. A physical examination on admission revealed mild jaundice, and a laboratory examination revealed a serum total bilirubin level of 2.1 mg/dL, an alkaline phosphatase (ALP) level of 350 U/L, and a gamma glutamyl transpeptidase (GGT) level of 514 U/L. Serum levels of aspartate aminotransferase (AST), alanine aminotransferase (ALT), CA 19-9, and carcinoembryonic antigen were normal. Ultrasonography (USG) showed an incidental ampullary mass, and computed tomography (CT) of the abdomen revealed an ampullary mass with a dilated biliary tract. Endoscopic retrograde cholangiography (ERC) demonstrated a distal bile duct obstruction, and a sessile mass in the ampulla of Vater was observed by endoscopy (Fig. 1). A histologic examina-

tion of the endoscopic biopsy showed focal nuclear atypia in some mucosal glands without evidence of malignancy. Even though a pathological examination was performed, it remained uncertain whether the ampullary mass represented a benign stricture or a malignancy. Thus, a laparotomy was performed. Frozen section demonstrated atypical glandular structures in surface mucosa (Fig. 2A), and the submucosal area revealed dilated glands with the proliferation of smooth muscle bundles (Fig. 2B). Although malignancy was suggested by frozen section, only an ampullectomy was performed since the ampullary mass was well demarcated and showed no evidence of distal common bile duct involvement. Grossly, there was an ill-defined, whitish, submucosal mass, measuring 1.5 × 1 × 1 cm with intact mucosa. Histologically focal atypical glandular structures were present in the surface mucosa, which corresponded with the endoscopic and frozen biopsy findings. These atypical glands were negative by avidine-biotin-peroxidase complex immunostaining for monoclonal CEA (DAKO; 1:100) and p53 (DAKO; 1:400). The submucosal lesion consisted of cystically dilated glands with a lobular arrangement and smooth muscle bundle proliferation. These glands were lined by a single-layer of cuboidal or columnar cells, which showed no nuclear atypia or mitotic features, confirming the diagnosis of ampullary adenomyoma. The patient had an uncomplicated postoperative course, and was well at the 9-month follow up.

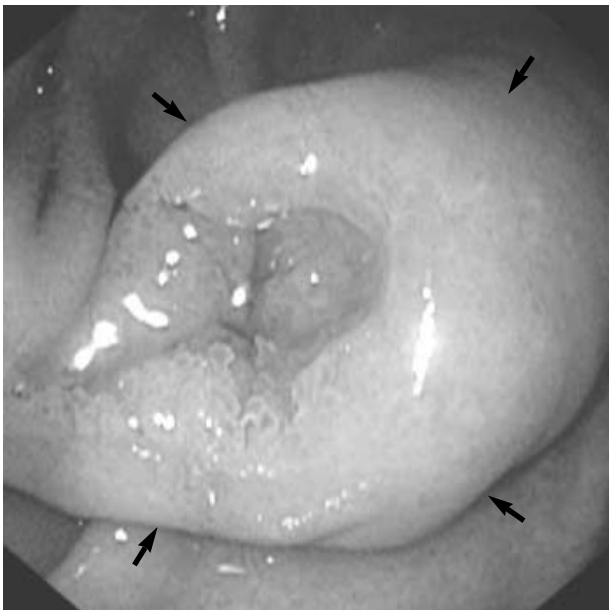

Fig. 1. Endoscopy reveals a polypoid sessile mass of ampulla of Vater (arrows).

Case 2

A 56-year-old woman with a history of hypertension was admitted to our hospital with an incidentally detected polypoid lesion of the distal common bile duct. USG, CT, and magnetic resonance cholangiopancreatography revealed diffuse dilatation of the biliary tract and a small intraluminal polypoid lesion in the distal common bile duct. A laboratory examination showed

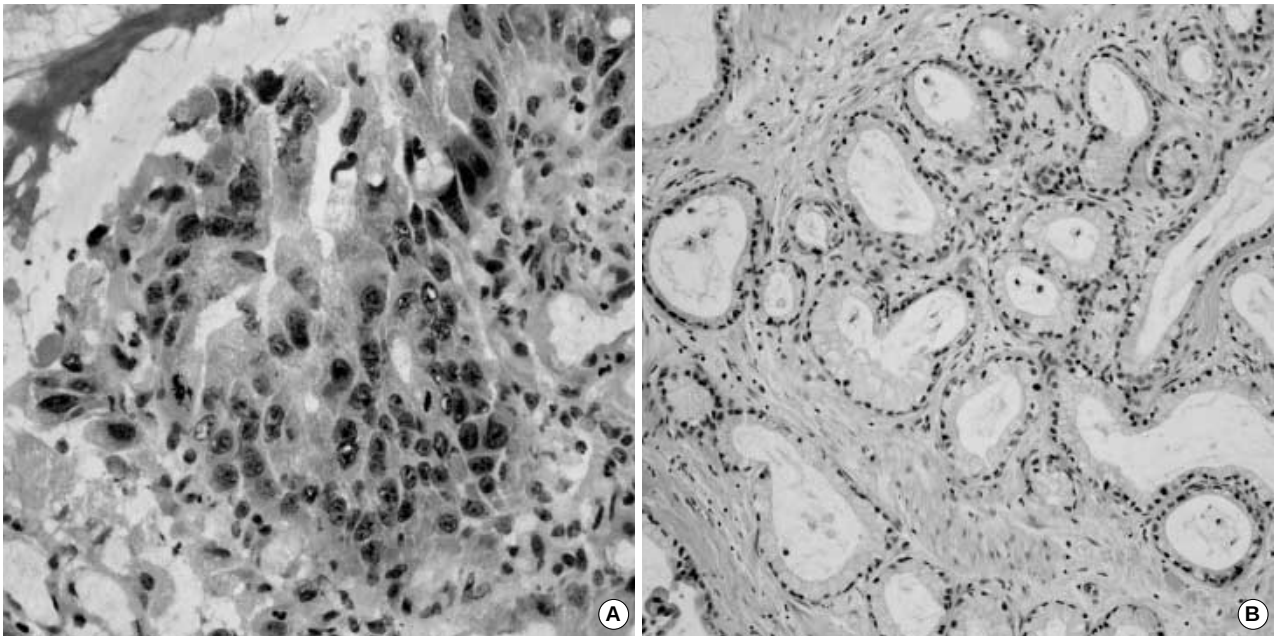

Fig. 2. (A) Microscopic features of atypical glandular structure of surface epithelium on frozen section. (B) At high magnification, the submucosal glandular lesions are composed of columnar epithelial cells with basally located nuclei without atypia.

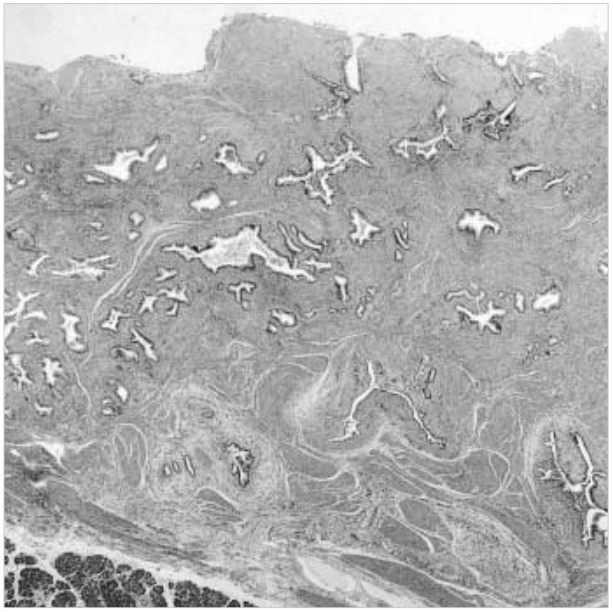

Fig. 3. At low magnification, thickened wall of distal common bile duct consists of glandular structures with lobular architecture and smooth muscle bundles.

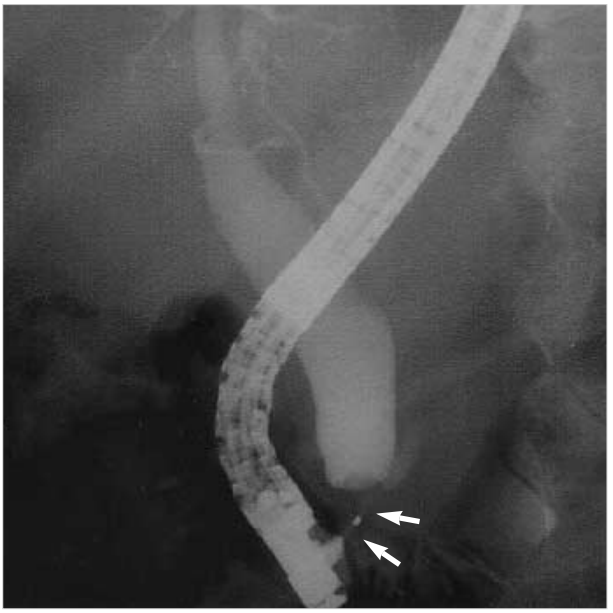

Fig. 4. Endoscopic cholangiography of case 3 shows abrupt luminal narrowing in the distal common bile duct (arrows) and dilatation of proximal common bile duct.

elevated ALP and GGT levels of 201 U/L and 250 U/L, respectively, and a high AST and ALT levels of 48 U/L and 108 U/L, respectively. Total and conjugated bilirubin, CEA, and CA19-9 levels were within normal limits. Under the impression of a distal common bile duct cancer, a laparotomy was performed. Although no mass lesion was identified in the lumen of the distal common bile duct, pylorus-preserving pancreaticoduodenectomy was carried out since malignancy could not be completely excluded. Grossly, the distal common bile duct revealed segmental wall thickening, measuring 0.4 cm, and a granular mucosal lesion, measuring 2×0.9 cm. The remaining mucosa of the proximal common bile duct was intact and dilated. Histologically, this segmental wall thickening of the distal common bile duct consisted of a benign glandular hyperplasia and proliferation of smooth muscle bundles (Fig. 3). There was no evidence of malignancy. The patient's postoperative recovery was uneventful, and she was well at the 7-month follow-up.

Case 3

A 78-year-old woman was admitted with mild epigastric pain and jaundice. USG, CT, and ERC revealed a diffuse dilatation of extrahepatic biliary tree with abrupt distal common bile duct narrowing (Fig. 4). A laboratory examination showed an elevated serum total bilirubin level of 2.6 mg/dL, ALP and GGT levels of 145 U/L and 247 U/L, respectively, and high AST and ALT levels of 159 U/L and 86 U/L, respectively. CEA, and CA19-9 levels were within normal limits. Pylorus preserving pancreaticoduodenectomy was carried out under the impression of distal common bile duct cancer. On gross examination, the distal common bile duct revealed segmental wall thickening (measuring 0.3 cm), with luminal narrowing (measuring 1.5×0.5 cm). The proximal bile duct was dilated with intact mucosa. The fundus of the gallbladder showed a polypoid mass, measuring 2.5×2×1.5 cm. Histologically, this lesion was an adenomyoma, and consisted of a lobular arrangement of benign glands and proliferated smooth muscle fibers. The polypoid mass of the gallbladder was a well-differentiated adenocarcinoma with proper muscle invasion. This patient was also well at 6-month follow up.

DISCUSSION

Adenomyoma of the extrahepatic bile ducts or ampulla of Vater is rare. According to the WHO classification, adenomyoma is defined as duct-like structures accompanied by hyperplasia of smooth muscle bundles.[1] It is difficult to evaluate the real incidence of adenomyoma of the biliary tract since many alternative terms are used, e.g., myoepithelial hamartoma, adenomyomatous hamartoma, adenomyomatous hyperplasia, and adenomyosis. Most adenomyomas are nodular tumor-like masses, although some exhibit an ill-defined diffuse configuration. Because only

a small number of cases of adenocarcinoma have been reported in association with periampullary adenomyoma, the latter lesion has been suggested to have a neoplastic potential.[2-4] It remains unclear whether adenomyomas themselves carry a high risk of malignant transformation or whether they are coincidental lesions in cases of adenocarcinoma. A preoperative endoscopic biopsy was performed in case 1, but did not result in a positive diagnosis because the specimen did not include the submucosal lesion. Intra-operative frozen examinations may be helpful in the differential diagnosis of adenomyoma and adenocarcinoma. However, most pathologists have limited experience of frozen sections of adenomyoma. In case 1, it was difficult to differentiate the cystically dilated glands of adenomyoma from well-differentiated adenocarcinoma, and the atypical glandular structure of the covering mucosal epithelium raised the possibility of adenocarcinoma by frozen section. However, most nuclei of the dilated glands were basally located without atypia or mitosis, and the lobular architecture of glands and thickened smooth muscle bundles were identified at low magnification in frozen and permanent sections. Although the atypical glandular structures showed cellular atypia, this lesion was confined to the surface mucosa and no multiple serial sections revealed a residual atypical glandular lesion. Based on histologic features of our cases, it appears that adenomyomas do not have malignant potential. In most case reports, patients received ERCP and endoscopic procedures that could damage the surface mucosal epithelium of ampulla of Vater. Thus, it seems that the atypical glandular lesion of the surface mucosal epithelium represents a reactive regenerative atypia rather than a true dysplastic lesion. Negative immunostaining results for CEA and p53 supported that the lesion is a reactive change rather than a dysplastic lesion. In fact, most reported cases of adenomyoma showed a lobular configuration of ducts and ductules without dysplastic change. Only three cases was reported to have dysplasia in periampullary adenomyoma.[5] To differentiate adenomyoma from adenocarcinoma, glands with basally located monotonous nuclei and a lobular arrangement of small ducts without desmoplastic reaction may represent adenomyoma, because most adenocarcinomas of the common bile duct, even the well-differentiated type, show intense desmoplastic reaction. The treatment of benign lesions of the biliary tract is variable. In some cases, multiple endoscopic biopsies of suspicious lesions have been advocated.[6] However, if a diagnosis is not established, intra-operative frozen examinations are indicated before proceeding to extensive surgery such as pancreaticoduodenectomy.[7,8]

In conclusion, adenomyoma of the ampulla of Vater and distal common bile duct are a rare lesion of clinical importance. Although the lesion is benign, it is often treated by extensive surgery. Therefore, careful histological evaluation is mandatory for a definite diagnosis. Adenomyoma should be included in the differential diagnosis of glandular lesions of the ampulla of Vater or distal common bile duct.

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