Adenoendocrine Carcinoma of the Accessory Papilla of the Duodenum: Report of a Case

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Abstract
This report describes a very rare case of an adenoendocrine carcinoma of the accessory papilla of the duodenum. A 70-year-old woman was admitted to the hospital complaining of epigastralgia. Gastrointestinal endoscopy showed a protruding tumor with ulceration at the accessory papilla of the duodenum. A biopsy revealed a small-cell carcinoma. Computed tomography showed a highly enhanced tumor in the early phase. No metastatic lesions were shown. Magnetic resonance cholangiopancreatography showed dilatation of the pancreatic duct, but a normal common bile duct. A pylorus-preserving pancreaticoduodenectomy was performed with lymph node dissection. Microscopically, the tumor was a small-cell neuroendocrine carcinoma with adenomatous differentiation. An immunohistochemical analysis showed positive staining for synaptophysin, chromogranin A, CD56, and carbohydrate antigen 19-9. The final diagnosis was an adenoendocrine carcinoma with lymph node metastasis. The postoperative course was uneventful and the patient is now doing well as an outpatient after 14 months of follow-up.

Key words Adenoendocrine carcinoma · Accessory papilla of the duodenum · Immunohistochemistry

Introduction

Neuroendocrine cell (NEC) carcinoma of the gastrointestinal tract accounts for <5% of all tumors of the alimentary tract.1 Neuroendocrine cell carcinoma shows strong cell atypism as well as abundant mitotic figures, vessel invasion, and metastasis in comparison to classical carcinoid tumors.2 The coexistence of NEC carcinoma and adenocarcinoma in the duodenum seems to be extremely rare. Morphologically, such lesions are classified into two subgroups: namely, composite-type tumors and collision-type tumors. Two hypotheses have arisen regarding the mechanism of association between adenocarcinoma and NEC carcinoma. One is that both are derived from a common multipotential epithelial stem cell and the NEC carcinoma component results from differentiation of the adenocarcinoma to an endocrine cell phenotype during the course of tumor progression. Another hypothesis is that the adenocarcinomas and NEC carcinomas arise from a multipotential epithelial stem cell and a primitive endocrine cell, respectively, coincidentally existing next to each other.3 Only nine cases of duodenal adenoendocrine cell carcinoma have been reported in Japan.4–11 The lesions were located in the ampulla of Vater in all of the reported cases and no such lesions have previously been reported in the accessory papilla of the duodenum. This report presents a case demonstrating adenoendocrine carcinoma of the accessory papilla of the duodenum.

Case Report

A 70-year-old woman was admitted to the hospital complaining of epigastralgia. A physical examination revealed no palpable surface lymph nodes or abdominal masses. Gastrointestinal endoscopy showed a protruding tumor with an ulceration of the accessory papilla in the second part of the duodenum. Computed tomography (CT) showed a low echoic mass in the submucosal layer and dilatation of the main pancreatic duct (Fig. 1). A biopsy specimen showed the presence of small-cell carcinoma. Computed tomography (CT) showed the tumor to be highly enhanced in the early phase. No metastatic lesion was observed. Magnetic resonance cholangiopancreatography showed the dilatation of the pancreatic duct, but not of the common bile duct.
Angiography showed tumor staining of the duodenum. Laboratory data revealed no abnormalities in either blood chemistry or tumor markers (carcinoembryonic antigen [CEA], neuron-specific enolase [NSE] and progastrin-releasing peptide).

A pylorus-preserving pancreaticoduodenectomy with lymph node dissection was thereafter performed. In the resected specimen, the tumor measured 1.5 cm in diameter and showed an elevated lesion with a small central ulceration of the mucosal surface. Microscopically, the tumor was an endocrine carcinoma mainly present in the muscularis propria of the accessory papilla with pancreatic invasion and lymphatic duct infiltration. Areas of adenomatous differentiation were observed sporadically within the area of the neuroendocrine carcinoma (Fig. 3). Only one lymph node along the lower common bile duct was positive for metastasis. The metastatic lymph node contained only carcinoid cells. An immunohistochemical analysis showed positive staining for synaptophysin, CD56, chromogranin A, carbohydrate antigen (CA) 19-9, gastrin-releasing peptide, and serotonin. Both the carcinoid and adenomatous cells were

(Fig. 2). Computed tomography shows a strong early-phase enhancement of the tumor. Magnetic resonance cholangiopancreatography shows the dilatation of the pancreatic duct, but not of the common bile duct.

Fig. 1. Left Gastrointestinal fiberscopy showing an elevated tumor with ulceration of the accessory papilla in the second part of the duodenum. Right Endoscopic ultrasonography showed a low echoic mass in the submucosal layer and the dilatation of the main pancreatic duct.

Fig. 2. Computed tomography shows a strong early-phase enhancement of the tumor. Magnetic resonance cholangiopancreatography shows the dilatation of the pancreatic duct, but not of the common bile duct.