Primary Adenocarcinoma of the Minor Duodenal Papilla

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A 70-year-old man was admitted to our institution due to aggravation of blood-sugar level control and because an abdominal CT showed dilatation of the main pancreatic duct. Upper gastrointestinal endoscopy revealed a flat elevated tumor with central ulceration in the second portion of the duodenum. Subsequent duodenoscopy for a more detailed examination showed that the tumor had originated in the minor duodenal papilla. A biopsy specimen showed moderately differentiated adenocarcinoma. Endoscopic retrograde pancreatography via the major duodenal papilla revealed a slightly dilated main pancreatic duct and obstruction of the accessory pancreatic duct. Endoscopic ultrasonography showed a hypoechoic mass in the minor duodenal papilla with retention of the muscularis propria of the duodenum. These findings suggest that the tumor existed only to a limited extent in the minor duodenal papilla, and that the tumor did not infiltrate into the pancreas. For treatment, pylorus-preserving pancreatoduodenectomy was performed, and histological findings revealed a well-differentiated adenocarcinoma that originated in the minor duodenal papilla. Primary adenocarcinoma of the minor duodenal papilla is extremely rare. Our case is the first report of primary adenocarcinoma of the minor duodenal papilla at an early stage with no infiltration into muscularis propria of the duodenum and pancreas.

Key Words: Adenocarcinoma, minor duodenal papilla, endoscopic ultrasonography

INTRODUCTION

The minor duodenal papilla is an opening for the accessory pancreatic duct. Some cases of tumors in the minor duodenal papilla have been reported, such as adenoma, carcinoid, gangliocytic paraganglioma, and adenomyoma. To our knowledge, however, primary adenocarcinoma of the minor duodenal papilla is extremely rare; only one case report has been published. That scarcity of reports might be explained by the difficulty in diagnosing whether a tumor is derived from the minor duodenal papilla or pancreatic parenchyma by endoscopic and histologic findings. We report an apparent primary adenocarcinoma of the minor duodenal papilla.

CASE REPORT

A 70-year-old man was admitted to our institution in September 2003 because of aggravation of blood sugar control. Subsequent abdominal computed tomography (CT) showed dilation of the main pancreatic duct. He had no other symptoms. No other abnormality was apparent by CT, except dilation of the main pancreatic duct. Although the fasting blood sugar level and HbA1c were abnormal (230 mg/dL, normal range: 60 - 110 mg/dL, and 6.3%, normal range: 4.3 - 5.8%), other laboratory findings, including tumor markers (CEA and CA 19-9), were normal. Screening upper gastrointestinal endoscopy showed a flat elevated tumor with central ulceration in the 2nd portion of the duodenum (Fig. 1). Subsequently, duodenoscopy using a side-view endoscope showed that the tumor was located in the descending duodenum where the minor duodenal papilla should have been situated (the major duodenal papilla was normally visible); furthermore, absence of the
minor duodenal papilla elsewhere was confirmed (Fig. 2). Biopsy results indicated moderately differentiated adenocarcinoma. Endoscopic retrograde pancreatography (ERP) via the major duodenal papilla revealed no apparent abnormality except a slightly dilated main pancreatic duct and obstruction of the accessory pancreatic duct (Fig. 3). To assess the tumor’s origin and staging, endoscopic ultrasonography (EUS, 7.5 MHz, UM230; Olympus Medical Systems Corp., Tokyo, Japan) was performed using a de aerated water-filled method in the duodenum. Those results revealed an elevated hypoechoic mass in the minor duodenal papilla with retention of the muscularis propria of the duodenum (Fig. 4), suggesting that the tumor existed only to a limited extent in the minor papilla and did not infiltrate into the pancreas. According to TNM classification of the tumor in the duodenal major papilla, this case was inferred to be T1.

For treatment, pylorus-preserving pancreaticoduodenectomy was done. The tumor was found in the minor duodenal papilla and macroscopic examination of the resected specimen showed that the tumor was 11 mm × 8 mm. Microscopic examination showed a well-differentiated adenocarcinoma, and the tumor cells surrounded the orifice of the minor papilla with a slight invasion into the submucosa (Fig. 5). There was no finding of lymph node metastasis or infiltration into the
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pancreas parenchyma (Fig. 6). These findings were thought to support the EUS findings. Because of the above-mentioned findings, we diagnosed a primary adenocarcinoma of the minor duodenal papilla. No relapse of the tumor after surgery was identified for 32 months.

DISCUSSION

Primary adenocarcinoma of the minor papilla is extremely rare. Actually, the incidence of adenocarcinoma of the minor duodenal papilla, especially at an early stage, might be low, but it seems that the main reason for the rarity of reports in the literature is that most tumors in this area are identified in an advanced stage; they seldom present symptoms in the early stage. It is therefore difficult to diagnose whether a tumor has originated from the minor duodenal papilla or pancreatic parenchyma.

The minor duodenal papilla is the opening for the accessory pancreatic duct. It consists of: the accessory pancreatic duct; pancreatic tissue of the dorsal pancreas, which penetrate the muscularis propria of the duodenum; and the surrounding fibrous connective tissue. Therefore, primary adenocarcinoma of the minor duodenal papilla is defined as a tumor derived from the epithelium covering the minor papilla or the accessory pancreatic duct within the duodenal wall. In the present case, the tumor cells surrounded the orifice of the minor duodenal papilla with a slight invasion into the submucosa and no infiltration into the pancreas parenchyma. Although it remains unclear whether the tumor had originated in the epithelium covering the minor duodenum papilla, the accessory pancreatic duct, or the ectopic pancreatic tissue penetrating the muscularis propria, the tumor was classified as a primary adenocarcinoma of the minor duodenal papilla.

Our patients had no symptoms or abnormalities aside from aggravation of blood sugar control and dilatation of the main pancreatic duct by CT. These findings are not associated with tumors of the minor duodenal papilla. Among previous reports regarding tumors of minor duodenal papilla, 2 patients had no symptom, 2 patients had transient epigastralgia, and 1 patient had acute relapsing pancreatitis. No specific symptom of the tumor of the minor duodenal papilla existed; in all patients, including the patient described herein, tumors were discovered incidentally by upper gastrointestinal examination. When routine endoscopic examination of the upper gastrointestinal tract is performed, careful observation of ampulla, including the minor duodenal papilla, is necessary.

Although CT is useful for evaluation of tumor staging, Yamao et al. described the usefulness of EUS for tumors of the minor duodenal papilla. In addition, in the presented case, EUS with 7.5 MHz revealed the tumor as a hypoechoic mass and...
duodenal muscularis propria as a hypoechoic layer. Consequently, we were able to verify that no infiltration into the pancreatic parenchyma had occurred. Diagnosis of whether the carcinoma infiltrated into the pancreatic parenchyma was an important matter because infiltration into the pancreatic parenchyma is an important predictive factor of prognosis.9

Recently, endoscopic resection (papillectomy) has been reported in patients with a tumor of the major duodenal papilla and a few malignant cases have been treated by papillectomy.10-12 Some investigators have asserted that the indication of papillectomy should be limited to benign cases.13,14 Regarding endoscopic treatment for tumors of the minor duodenal papilla, a few cases of papillectomy have been reported,4,5 these also were all benign. Our case showed malignancy in spite of the T1 stage. Therefore, we did not choose endoscopic treatment. Expansion of indications of papillectomy for malignancy in duodenal papilla will require further accumulation of case information.

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