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Acinar Cell Carcinoma Arising from an Ectopic Pancreas

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Abstract
We herein report a rare case of ectopic pancreatic acinar cell carcinoma (ACC) which presented as a submucosal tumor of the pylorus. A 73-year-old man came to our hospital presenting with epigastralgia. Esophagogastroduodenal endoscopy showed no mucosal lesions, but a submucosal tumor was observed around the pylorus. Abdominal computed tomography revealed two round masses. One was located in the pylorus, while the other was found between the portal vein and the inferior vena cava. An examination of a biopsy specimen was inconclusive. We diagnosed a gastrointestinal stromal tumor or malignant lymphoma preoperatively, and decided to perform an operation in order to confirm the diagnosis and select the optimal treatment. Intraoperatively, the mass in the pylorus invaded the pancreatic head, and the lymph node in the hepatoduodenal ligament was swollen. We performed a pancreaticoduodenectomy as a radical excision. The resected specimen showed the 7.6 × 4.9-cm size tumor to mainly originate from the pylorus. Histopathologically, the tumor was identified as pancreatic ACC with lymph node metastasis. The tumor cells were labeled by immunohistochemical staining for α1-antitrypsin. Because of the tumor location, we considered the tumor to have originated from the ectopic pancreatic tissue in the stomach. This is only the second case of ACC originating from an ectopic pancreas reported in the literature.

Key words Pancreatic acinar cell carcinoma · Submucosal tumor · Pylorus · Ectopic pancreas

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Introduction

Pancreatic acinar cell carcinoma (ACC) is a rare neoplasm, accounting for only 1%–2% of all exocrine tumors of the pancreas.[1] Pancreatic ACC can occur at any location in the pancreas, although most are found in the head or neck of the gland.[2] Pancreatic ACC usually appears as an exophylic, oval or round, well-marginated, and hypovascular mass on computed tomography (CT) and magnetic resonance imaging (MRI).[3] Because of the lack of any clinical findings and laboratory abnormalities for this disease, pancreatic tumors tend to be large at the time of diagnosis. The prognosis of pancreatic ACC is better than that of ductal adenocarcinoma, but worse than that of pancreatic endocrine tumors. The mean survival duration of pancreatic ACC is 18 months, with 1- and 3-year survival rates of 57% and 26%, respectively.[4]

Neoplasms arising from an ectopic pancreas are extremely rare. Adenocarcinomas originating from an ectopic pancreas have been reported in fewer than 30 cases in the literature.[5] There has been only one previous report[6] of ACC occurring from an ectopic pancreas. We herein report an extremely rare case of pancreatic ACC presenting as a submucosal tumor of the pylorus, which is considered to have originated from ectopic pancreatic tissue.

Case Report

A 73-year-old man came to our hospital presenting with epigastralgia. The physical findings were within the normal ranges. A physician noted no cardiac insufficiency, but two abdominal masses by abdominal computed tomography (CT). The two masses were located in the pylorus, and between the portal vein and the inferior vena cava (Fig. 1). The images of the masses were strongly and heterogeneously enhanced. Except for

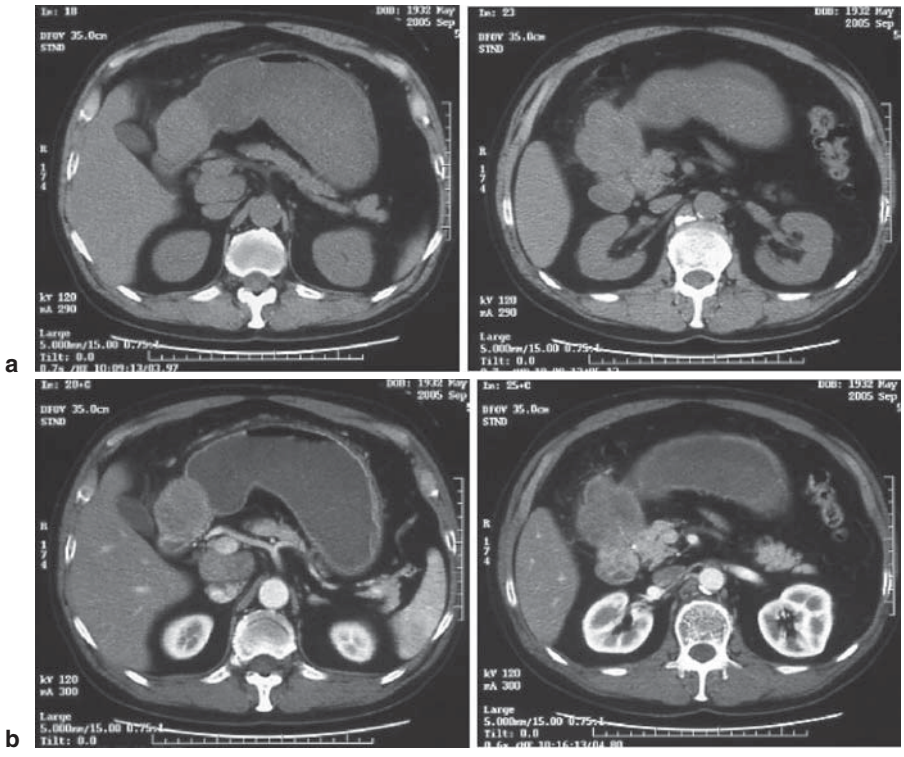

Fig. 1. a Plain computed tomography (CT) showed two masses, one located around the pylorus and another between the portal vein and inferior vena cava. **b** Enhanced CT showed the mass to have a strong enhancement heterogeneously

high interleukin (IL)-2 receptor (1300 U/ml) and lactate dehydrogenase (432 IU/l) concentrations, the laboratory findings were within the normal ranges including those of carcinoembryonic antigen (CEA) and carbohydrate antigen (CA) 19-9. Esophago-gastroduodenal endoscopy revealed a submucosal tumor in the pylorus with no mucosal lesions. The patient underwent ^{18}F-fluorodeoxyglucose positron emission tomography (^{18}F-FDG-PET). The standard uptake values of the two spots were 30 and 16, respectively. Magnetic resonance cholangiopancreatography showed no stenosis of the common bile or pancreatic duct. Abdominal angiography showed a round staining area fed by the gastroduodenal artery. An examination of a biopsy specimen was inconclusive. Our preoperative diagnosis was a gastrointestinal stromal tumor (GIST) or malignant lymphoma (ML), and we decided to perform an operation to confirm the diagnosis and select the optimal treatment. Intraoperatively, the mass in the pylorus had invaded the pancreatic head, and the lymph node in the hepatoduodenal ligament was swollen. We performed a pancreaticoduodenectomy for a radical excision. The resected specimen included a yellow, solid 7.6 × 4.9-cm tumor (Fig. 2). The tumor was mainly located in the pylorus, and a piece of the tumor had invaded the pancreatic head. Histopathologically, the tumor showed an acinar pattern composed of small lumina. The tumor cells had hyperchromatic round nuclei on hematoxylin–eosin staining, and they were positive

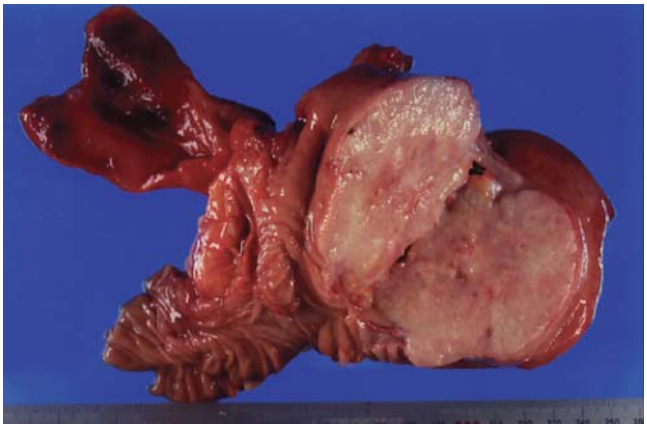

Fig. 2. The resected specimen included a yellow, solid tumor that measured 7.6 × 4.9 cm in size

for α1-antitrypsin (Fig. 3). The tumor was identified as pancreatic ACC with lymph node metastasis. The tumor cells were labeled by immunohistochemical staining for α1-antitrypsin. Because of the tumor location, we considered the tumor to have originated from ectopic pancreatic tissue in the stomach. The patient was discharged 65 days after the operation. Seven months after the operation, follow-up CT revealed tumor metastasis to the liver. He underwent chemotherapy by hepatic arterial infusion (gemcitabine, 1000 mg/day; cisplatin, 5 mg/day; 5-fluorouracil, 500 mg/day; days 1, 8,

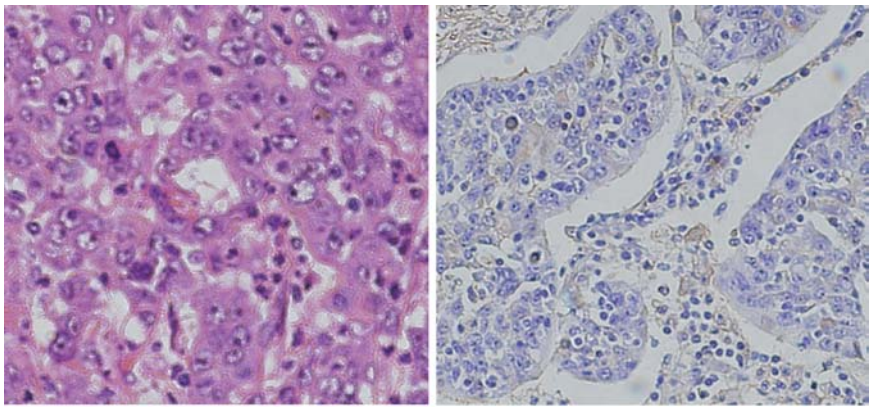

Fig. 3. a Microscopically, the tumor demonstrated an acinar pattern composed of small lumina, and the cells had hyperchromatic round nuclei (H&E stain). **b** The tumor cells were positive for α1-antitrypsin

15). After three courses, the size of the tumors dramatically decreased. He has remained healthy during a close follow-up as an outpatient for 11 months after the operation.

Discussion

None of the common tumor markers have been consistently noted in ACC, but elevated serum concentrations of α-fetoprotein (AFP), CEA, and CA19-9 have been.[7] In our case, the serum concentration of AFP was not measured. The serum concentrations of CEA and CA19-9 were within the normal ranges. Regarding any abnormalities in the laboratory findings in our case, a high IL-2 receptor concentration was noted. IL-2 receptor was reported to be a new tumor marker of pancreatic adenocarcinoma.[8] Although a correlation between pancreatic ACC and IL-2 receptor concentration was not clarified, IL-2 receptor was the only tumor marker in our case that showed a high concentration.

Although the CT and MRI features of pancreatic ACC have not been well defined, Tatli et al. reported that the tumor is usually observed as an exophylic, oval or round, well-marginated, and hypovascular mass on CT and MRI.[3] In our case, the morphological features of the tumor were compatible with this description; however, they were strongly enhanced.

The diagnosis of ACC requires the confirmation of acinar differentiation histochemically, immunohistochemically, and ultrastructurally. Acinar differentiation is defined as the production of pancreatic enzymes (e.g., trypsin, chymotrypsin, lipase, amylase), and the antigens α1-antitrypsin and α1-antichymotrypsin have been reported to be positive in ACC.[6] In our case, α1-antitrypsin was positive and we thus diagnosed the patient to have pancreatic ACC.

Matsuyama et al. reported a case of ACC of the pancreas eroding the pylorus and duodenal bulb.[9] The patient showed a submucosal tumor extending from the pylorus to the duodenal bulb by endoscopy, as in our case. They performed a pancreaticoduodenectomy under the diagnosis of a nonfunctioning pancreatic endocrine tumor or gastric cancer. Similarly, we performed a pancreaticoduodenectomy under a diagnosis of GIST or ML. A resection is usually selected for the treatment of ACC, and the prognosis of unresectable cases is poor. Riechelmann et al. reported the effect of weekly paclitaxel therapy on pancreatic ACC.[10] Kobayashi et al. reported a case of pancreatic ACC that was successfully treated by an en bloc resection and intraperitoneal chemotherapy for peritoneal relapse.[11] Other authors reported concurrent chemoradiation to be effective for treating pancreatic ACC.[12–14] These reports were successful cases, but in most cases of pancreatic ACC, chemotherapy and radiation therapy tend to be not very effective.[2] Our case showed a relapse with metastasis to the liver, and we performed chemotherapy by hepatic arterial infusion. Hepatic arterial infusion of gemcitabine, cisplatin, and 5-flourouracil was very effective. Although we failed to make a correct preoperative diagnosis, we considered that the treatment including the operative procedures was reasonable.

Ectopic pancreatic tissue is relatively uncommon, and it occurs most frequently in the stomach.[6] Furthermore, neoplasms arising from an ectopic pancreas are extremely rare. Adenocarcinomas occurring from an ectopic pancreas were reported in fewer than 30 cases in the literature.[5] Sun and Wasserman reported the only other case of ACC occurring from an ectopic pancreas.[6] The patient underwent a partial gastrectomy with Billroth II reconstruction. Because the additional treatment and the prognosis of their patient were not described, we could not refer to their procedures. Because typical pancreatic ACC eventually develops recurrence in approximately 70% of all patients,[2] a close follow-up is essential.

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