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CASE REPORT

Acinar cell carcinoma of the pancreas: A rare tumor with a particular clinical and paraclinical presentation

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Summary Acinar cell carcinoma (ACC) of the pancreas is a rare tumor with an extremely low incidence rate. While the number of reported patients with ACC is relatively small, a long-term survival rate has been noted in patients with neuroendocrine differentiation. A 39-year-old woman visited our emergency department for upper gastrointestinal bleeding. Endoscopy indicated extrinsic compression of the posterior body of the stomach, together with a large, 10-cm, central ulcer covered with necrotic tissue. Abdominal computed tomography (CT) indicated a lesion that involved the whole of the pancreas, with a fistula in the stomach, which was suspected of being a degenerative intraductal papillary mucinous tumor of the pancreas. Magnetic resonance imaging (MRI) of the pancreas was performed, and the results further strengthened our suspicions by demonstrating the presence of cystic lesions and tumor buds. A total duodenopancreatectomy, including total splenectomy and gastrectomy, was performed, along with two independent Roux-en-Y anastomoses (one esojejunal and one hepaticojejunal). The tumor also had a wide opening in the stomach. The patient's postoperative course was marked by partial thrombosis of the portal vein, which was treated medically. Histopathological examination provided evidence of pancreatic ACC. The diagnosis of ACC should be considered in the presence of cutaneous lesions, which were absent in the case of our patient, and colonoscopy is also highly desirable because of the various forms associated with familial adenomatous polyposis. The prognosis, which includes a 5-year survival rate of 45%, in the population with an R0 resection is better than that for ductular adenocarcinoma, thus prompting the more aggressive management of this type of tumor.

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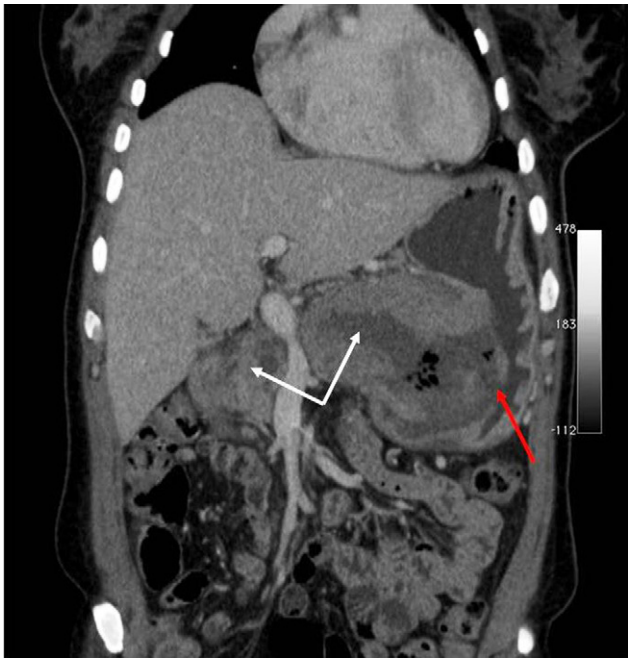

Figure 1 Abdominal computed tomography (CT), coronal view, shows solid and cystic lesions throughout the pancreas (white arrow) and an opening into the stomach (red arrow), with compression of the superior mesenteric vessels.

Introduction

Acinar cell carcinoma (ACC) of the pancreas is a rare tumor with a low rate of incidence. It also has clinical, diagnostic and prognostic characteristics that can distinguish it from pancreatic ductal adenocarcinoma.

Observation

A 39-year-old woman, an insulin-dependent diabetic of around 3 months' duration, visited our emergency department because of abundant upper gastrointestinal bleeding (hemoglobin [Hg] = 6 g/dL). After resuscitation and stabilization of the patient (Hg = 11 g/dL), upper digestive tract endoscopy was performed, which demonstrated extrinsic compression at the posterior aspect of the body of the stomach and a 10-cm central ulcer covered by necrotic tissue. However, the second part of the duodenum was normal. We considered a diagnosis of a nearby stomach tumor as the cause of the severe upper gastrointestinal bleeding. Abdominal computed tomography (CT) findings showed solid and cystic lesions throughout the whole of the pancreas, with an opening into the stomach, and compression of the superior mesenteric vessels (Fig. 1). Thus, an intraductal papillary mucinous tumor was suspected. Given the imminent risk of repeat bleeding, magnetic resonance imaging (MRI) was performed, which provided evidence of cystic lesions of the pancreatic duct with associated tumor buds (Fig. 2). Given the complicated nature of the tumor and the young age of the patient, we decided to treat her surgically.

The operation involved a bisubcostal laparotomy, and exploration of the tumor revealed involvement of the entire pancreas. Moreover, solid and cystic lesions were observed with the aid of an intraoperative ultrasound examination. These lesions were adherent to, but did not invade, the superior mesenteric axis. A total duodenopancreatectomy, involving total splenectomy and gastrectomy, and two separate Roux-en-Y anastomoses, was performed. Esojejunal and hepaticojejunal anastomoses were also created. The tumor had a wide opening into the stomach (Fig. 3). The patient's postoperative course was marked by partial thrombosis of the portal vein, which was treated medically. Histopathological examination was conclusive of pancreatic ACC (Fig. 4). The patient also received adjuvant chemotherapy, consisting of three cycles of cisplatin and gemcitabine. Eleven months after the operation, the patient remains well with no evidence of recurrence.

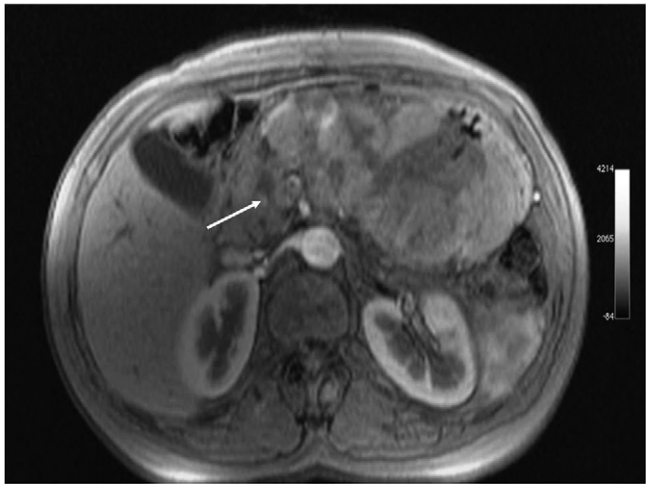

Figure 2 Magnetic resonance imaging (MRI), transverse section, shows the presence of bulging tissue in Wirsung's canal.

Discussion

Although acinar cells constitute most of the pancreatic parenchyma, ACC represents only 1% of all exocrine pan-

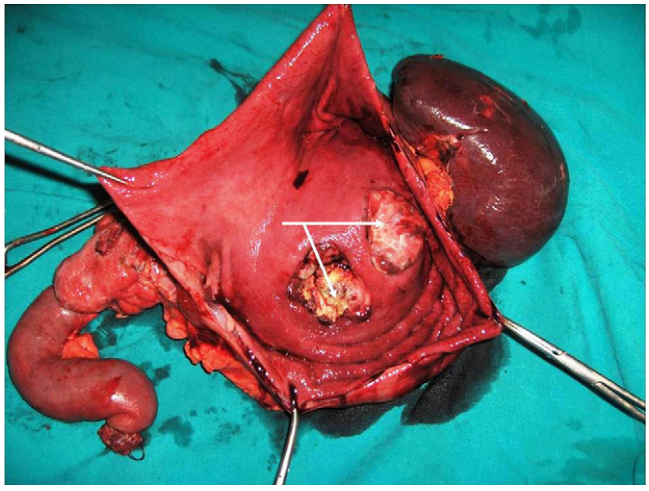

Figure 3 Macroscopic view of a tissue specimen taken from the whole of the pancreas, stomach and spleen reveals the large opening of the tumor in the posterior aspect of the stomach.