Pancreatic Acinar Cell Carcinoma with Diffuse Whole-organ Involvement: an unusual presenting pattern

WEN-HUI CHAN  CHIEN-MING CHEN  MING-YI HSU  KUANG-TSE PAN  SUNG-YU CHU  JENG-HWEI TSENG

Department of Medical Imaging and Intervention, Chang Gung Memorial Hospital, Linkou, and College of Medicine, Chang Gung University, Taoyuan, Taiwan

ABSTRACT

Pancreatic acinar cell carcinoma is a rare tumor arising from exocrine components of pancreas. Several case reports and case series studies have reported the imaging characteristics of the pancreatic acinar cell carcinoma, including computed tomography (CT) and magnetic resonance imaging (MRI). It is generally considered to be an exophytic, well-circumscribed mass arising from pancreas without pancreatic ductal dilatation. We report an unusual case of pancreatic acinar cell carcinoma with diffuse organ involvement initially mimicking a duodenal tumor. The diagnosis was confirmed histologically after a total pancreatectomy and the specimen showed diffuse pancreatic involvement of the tumor. To the best of our knowledge, diffuse whole-organ involvement of pancreatic acinar cell carcinoma has never been reported in the scientific literature.

CASE REPORT

A 70-year-old woman presented to the emergency department with passage of tarry stool for three days. She had a history of peptic ulcer and diabetes mellitus under regular oral antidiabetic medication for six years. On physical examination, she was anemic but not jaundiced. There were no skin nodules or arthritis. Esophagastroduodenoscopy revealed a duodenal mass with fresh blood clots. Endoscopic biopsy demonstrated a moderately differentiated adenocarcinoma. Complete blood count showed a normocytic anemia (hemoglobin: 5.5 g/dL). Biochemical profile showed normal levels of total bilirubin (0.4 mg/dL) and liver enzyme. Tumor markers including CEA (2.3 ng/mL), CA19-9 (10.93 U/mL) and alpha-fetoprotein (1.4 ng/mL) were within normal limits.

Contrast-enhanced abdominal CT scan (Fig. 1a) revealed a huge pancreatic tumor involving the entire pancreas and invading the duodenum. Magnetic resonance imaging (Fig. 1b-1e) showed diffuse enlargement of the pancreas with lobulated contour and direct invasion to the duodenum. The lesion appeared hypointense on T1-weighted images and mildly hyperintense on T2-weighted images. The lesion showed moderate enhancement on arterial phase and an enhancing peripheral capsule on delayed
phase. There was no peripancreatic lymphadenopathy or infiltration of the peripancreatic fat planes. The portal trunk and superior mesenteric vein were encased. Small stellate necrosis in the pancreas was visualized. Magnetic resonance cholangiopancreatography (MRCP) showed mild compression of common bile duct. The pancreatic duct was difficult to depict.

The patient subsequently underwent a total pancreatectomy. Gross examination of the specimen showed a hard encapsulated tumor at the pancreatic head and uncinate process extending to the whole length of the pancreas (Fig. 2a). Microscopic examinations showed acinar configuration of the cells and immunohistochemistry demonstrated negative CD 56, chromogranin, synaptophysin, CEA, PR and HER-2, confirming the diagnosis of acinar cell carcinoma (Fig. 2b). No normal pancreatic tissue was found. Postoperatively, the patient developed intractable hyperglycemia that required insulin therapy. Poor performance status prevented her from receiving chemotherapy. The patient died one year later from massive gastrointestinal bleeding.

DISCUSSION

The pancreas is composed of endocrine and exocrine components. Pancreatic acinar cell carcinoma arises from the exocrine component of pancreas and demonstrates characteristic acinar configuration. Acinar cell carcinoma is traditionally considered to be a disease of older adults and typically occurs in the 5th to 7th decades of life [3]. There is a strong male predominance [5]. The neoplastic cells excrete pancreatic enzyme that may circulate systemically and cause a myriad of symptoms. The clinical manifestations are diverse, including epigastric pain, abdominal mass, back pain and painful skin nodules presumably fat necrosis resulting from hyperlipasemia [6]. Despite the classic presentation, the prevalence of the so-called “lipase hypersecretion syndrome” is found in less than 10% of patients [4]. Unlike pancreatic ductal adenocarcinoma, jaundice is rare. When the tumor invades the duodenum, gastrointestinal bleeding may ensue [7], as did our patient. Interestingly, our patient presented with diabetes mellitus.

Figure 1

Figure 1. a. Contrast-enhanced CT scan on arterial phase shows a lobulated pancreatic mass with diffuse whole-organ involvement. The tumor showed moderate enhancement. b. Magnetic resonance imaging T1-weighted images show diffuse hypointensity of the pancreas. c. On T2 weighted fat-saturated image, the mass was mildly hyperintense while the stellate necrosis showed foci of high signal intensity. d. Contrast-enhanced images on arterial phase demonstrated moderate enhancement of the mass. e. Delayed phase shows a peripheral thin-enhancing capsule.
Diffuse pancreatic acinar cell carcinoma

Figure 2

2a 2b

Figure 2. a. Surgical specimen demonstrated diffuse involvement of the entire pancreas. The resected specimen measured 15 x 9.5 x 4.5 cm in size. The tumor involved the entire pancreas, abutting the ampulla of Vater and the common bile duct, causing mild ductal dilatation with ulcerated duodenal mucosa (not shown). b. Low-power view photomicrograph showed acinar configuration of tumor cells (asterisk) and invasion to the duodenal mucosa (D in the figure).

for the last six years. It is postulated that diffuse-whole organ involvement of tumor caused replacement of endocrine component of pancreas, resulting in diabetes mellitus requiring antidiabetic treatment.

Despite several previous reports that described the CT and MR characteristics of pancreatic acinar cell carcinoma, prospective imaging diagnosis is still difficult because of its rarity and diverse imaging presentation [2-5]. In fact, acinar cell carcinoma was not considered in the preoperative imaging diagnosis of our patient. Acinar cell carcinoma was traditionally described as uniformly or partially circumscribed mass with thin enhancing capsules. Intratumoral hemorrhage, amorphous calcification and cystic components have also been depicted. On MRI, the tumor exhibits T1 hypointensity and T2 iso- to hyperintensity. Heterogeneous enhancement was shown on CT or MR dynamic study.

Although the imaging presentation is diverse, there are still some clues for differentiating acinar cell carcinoma from other pancreatic neoplasms. Unlike pancreatic ductal adenocarcinoma, acinar cell carcinoma usually presents with relative larger size, exophytic nature, well-defined enhancing capsule, cystic or necrotic change, the lack of pancreatic ductal dilatation, and lack of vascular encasement [5]. On rare occasion, intraductal and papillary variant of acinar cell carcinoma may show small intraductal polypoid mass with dilatation of pancreatic duct, mimicking ductal adenocarcinoma [8]. When the tumor contains cystic components, differential diagnosis also includes neuroendocrine tumor, intraductal papillary mucinous neoplasm (IPMN) or other cystic tumor [9]. However, acinar cell carcinoma are usually not overtly hypervascular compared against neuroendocrine tumor [9]. On the other hand, IPMN usually shows dilatation of the pancreatic duct.

As far as tumor markers are concerned, levels of CEA and CA 19-9 are low in acinar cell carcinoma, as opposed to ductal adenocarcinoma [1]. There are few reports regarding elevated alpha-fetoprotein level in acinar cell carcinoma, which is rarely elevated in ductal adenocarcinoma [10]. However, the mechanism by which pancreatic tumors secrete alpha-fetoprotein is still not elucidated [10].

In past series, pancreatic acinar cell carcinoma showed a slight predilection for the pancreatic head. Diffuse whole-organ distribution of the tumor, as in our case, has never been reported. This may suggest an unusual indolent growth pattern allowing the tumor to grow without any symptoms.

In conclusion, we present a case with diffuse whole-organ involvement of pancreatic acinar cell carcinoma with initial presentation of gastrointestinal bleeding. This case illustrates an unusual presenting pattern of this rare pancreatic tumor.
REFERENCES