Pancreatic Acinar Cell Carcinoma Presenting with Diffuse Pancreatic Enlargement

Hongwei Li[1], Yaping Wu[1], Xiaodan Xiang[1], Jun Zhou[2] and Qian Li[1]

[1]*Department of Gastroenterology, Xiangya Hospital, Central South University, China*
[2]*Department of Biliary and Pancreatic Surgery, Xiangya Hospital, Central South University, China*

ABSTRACT

Pancreatic acinar cell carcinoma (PACC) is a rare tumor of the exocrine pancreas. It accounts for only 1% of all pancreatic malignancies. A 39-year woman sought treatment for repeated abdominal pain and jaundice. The computed tomography (CT) and magnetic resonance imaging (MRI) suggested diffuse enlargement of the pancreas. 18 F-fluorodeoxyglucose (FDG) positron emission tomography (PET)/CT showed diffuse enlargement of the pancreas and increased glucose uptake. The endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA) biopsy of the pancreas was performed, and the case was finally diagnosed as PACC. After the sixth course of chemotherapy with gemcitabine and albumin paclitaxel, a CT scan showed significant shrinkage of the pancreas. PACC very rarely presents with diffuse enlargement of the pancreas. This case illustrates a rare presentation of PACC with diffuse pancreatic enlargement, which was effectively treated with gemcitabine and albumin paclitaxel chemotherapy.

Key Words: *Pancreatic acinar cell carcinoma, Positron emission tomography/computed tomography, Chemotherapy.*

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INTRODUCTION

Pancreatic acinar cell carcinoma (PACC) is a rare pancreatic tumor that accounts for only 1% of all pancreatic malignancies.[1] PACC may exhibit different clinical symptoms, morphological features, and outcomes, leading to difficulties in the clinical diagnosis of the tumor.[2] It usually presents with non-specific symptoms, including abdominal pain and weight loss.[2,3] The imaging features of PACC usually appear as an elliptical or circular, well-circumscribed exogenous mass. The computed tomography (CT) and magnetic resonance imaging (MRI) enhancement show low-density enhancement relative to normal pancreatic tissue.[4]Diffuse enlargement of the pancreas is very rare in pancreatic tumors. We report a case of PACC, which presented with diffuse enlargement of pancreas due to the extensive infiltration of tumor cells in the pancreas.

Although PACC has invasive clinical manifestations, it has a better prognosis than pancreatic ductal carcinoma; thus surgical intervention is recommended early in the disease.[5] Chemotherapy may be considered for the inability to perform radical resection due to advanced disease. Due to the rarity of the disease, standard chemotherapeutic regimens have not yet been established. Gemcitabine combined with albumin paclitaxel chemotherapy was effective in the cases, we reported.

CASE REPORT

A 39-year woman came to our hospital with complaints of abdominal pain for two years and jaundice for two months. She provided written informed consent for reporting this case. The patient was physically healthy and had no known comorbids. The physical examination revealed visible yellow staining of skin, mucosa and sclera, tenderness under the xiphoid process, and a palpably enlarged gallbladder and pancreas. Blood tests revealed serum amylase, lipase, and IgG4 levels within normal range. Abdominal ultrasound showed diffuse enlargement of pancreas and substantial echogenicity. The endoscopic ultrasound revealed diffuse enlargement of pancreas and bile duct dilatation. The CT showed diffuse enlargement of pancreas, with a sausage-like appearance, and an increased delay in heterogeneity after angiography (Figures 1A, B). The MRI showed diffuse enlargement of pancreas, with a change in homogeneity after angiography. The pancreatic segment of the common bile duct was compressed and narrowed, the upper and lower bile ducts were dilated, the main pancreatic duct was dilated, and the gallbladder was enlarged (Figures 2A, B). 18 F-FDG PET/CT showed diffuse enlargement of pancreas and increased glucose uptake with the maximum standard uptake value (SUVmax) of 16.8 (Figures 3A–D). The patient then

Correspondence to: Qian Li, Department of Gastroenterology, Xiangya Hospital, Central South University, China
E-mail: qianli0816@csu.edu.cn
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accepted endoscopic ultrasound-guided pancreatic fine-needle aspiration (EUS-FNA) biopsy.

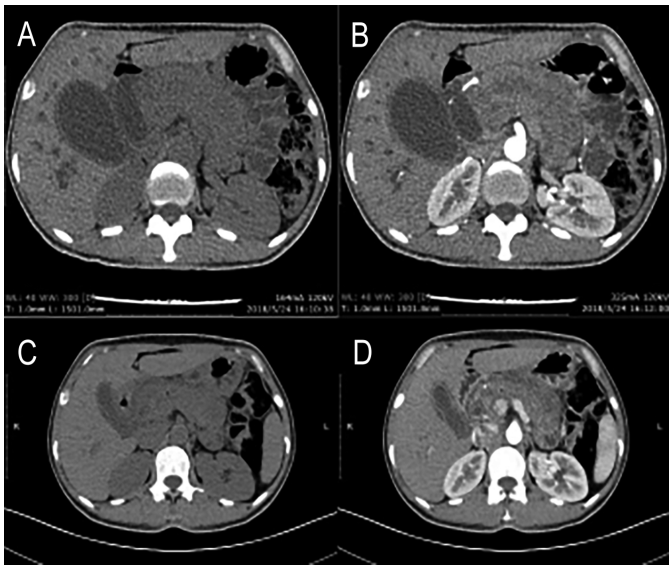

Figure 1: (A, B) CT showing diffuse enlargement of the pancreas, with a sausage-like appearance, with a longer delay in heterogeneity after angiography. (C, D) After 6 cycles of chemotherapy, the diffuse enlargement of CT pancreas had decreased significantly.

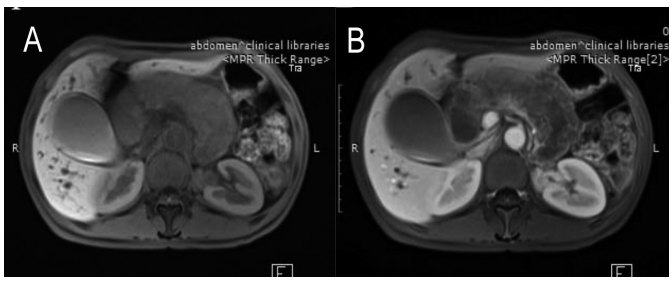

Figure 2: (A, B) MRI showed diffuse enlargement of the pancreas, which showed a change in homogeneity after angiography.

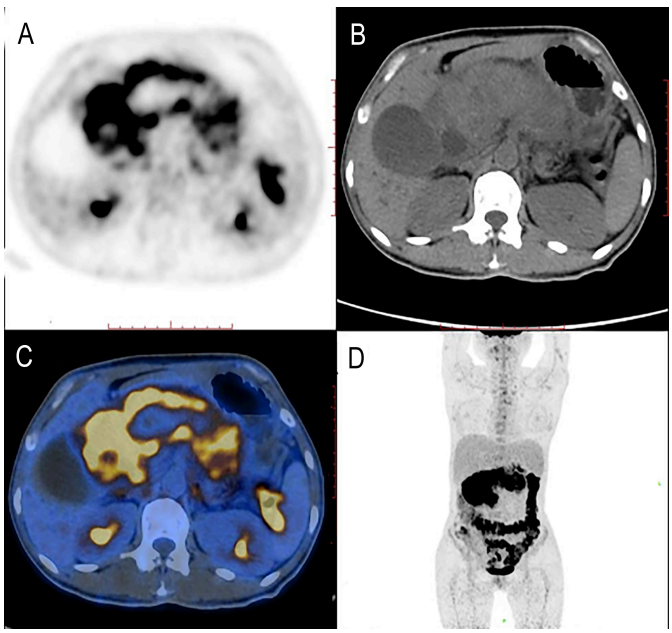

Figure 3 (A-D): 18 F-FDG PET/CT shows diffuse enlargement of the pancreas and increased glucose uptake.

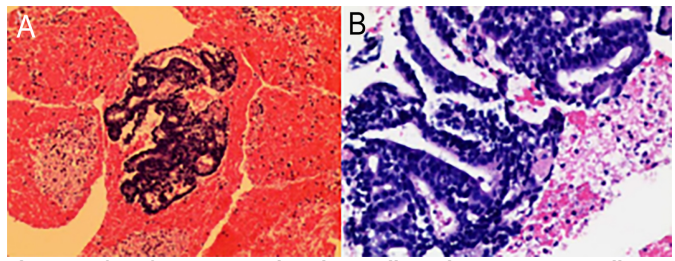

Figure 4: (A, B) In pancreatic acinar cell carcinoma, cancer cells are arranged as cohesive groups of cells with lumen formation, forming an adenoid structure, with little stromal reaction.

Histological examination confirmed PACC. Immunohistochemistry showed that the tumor cells were positive for α1-antitrypsin (AAT), α1-antichymotrypsin (AACT), and β-catenin, negative for chromogranin (CgA), synaptophysin (Syn), CD56 and CD10, and the ki-67 index was 50% (Figure 4A, B). After diagnosis of PACC, the patient was treated with gemcitabine combined with albumin paclitaxel. For her own reasons, the patient refused surgical treatment. After six cycles of chemotherapy, which lasted for three months, a re-evaluation *via* CT showed significant decrease of the diffuse pancreatic enlargement (Figures 1C, D).

DISCUSSION

Pancreas is mainly composed of acinar cells, which produce digestive enzymes such as trypsin, chymotrypsin, and pancreatic amylase. Most cases of pancreatic cancer are ductal-derived adenocarcinomas. PACCs are very rare pancreatic tumors, accounting for only 1% of all pancreatic malignancies.[1] PACC mainly present with non-specific symptoms, such as abdominal pain, diarrhea, weight loss, nausea and vomiting.[1-3] Biliary obstruction and jaundice can also occur, but are rarer than in ductal adenocarcinoma.[1] In rare cases, lipase hypersecretion syndrome occurs in patients with PACC, with an incidence of 10% to 15%, characterised by excessive lipase production, subcutaneous fat necrosis, and polyarthralgia.[3] In our case, the patient presented with abdominal pain and weight loss. Obstructive jaundice was also present, and it was caused by PACC pressing into the lower part of the common bile duct, as was confirmed by imaging and dilatation of the upper and lower bile ducts, and the gallbladder.

PACC may occur anywhere in the pancreas, and tumors are usually large in size, with an average diameter of 8–10 cm.[1,2] From an imaging point of view, in CT and MRI, PACC usually presents as a well-defined circular or elliptical mass.[4,5] In our case, the tumor cells had infiltrated the entire pancreas, resulting in diffuse enlargement of the pancreas and formation of a sausage-like structure.[6,7]

Histopathologically, PACC has a lobular architecture with little fibrous septation.[2,3] Several different growth patterns have been reported, the most common being the solid and acinar patterns, which can occur together in a single tumor.[2,3] In addition to the typical acinar structure, its microscopic appearance is indistinguishable from that of other pancreatic tumors, such as solid pseudopapillary tumor (SPT), pancreatic neuroen-

docrine tumors, and ductal adenocarcinoma. In this context, immunohistochemistry is very important for distinguishing between PACC and other pancreatic tumors. We usually use digestive enzymes secreted by the pancreas, such as trypsin, lipase, and amylase, as diagnostic immunological markers.[8] Recent studies have shown that monoclonal antibodies against the Bcl-10 protein are useful for detecting PACCs because of their high specificity and sensitivity.[8] In our case, trypsin and chymotrypsin were positive. CD56 and CD10, which have been found to be associated with SPT, were negative, as were the Syn and CgA, two typical neuroendocrine tumor immunological markers.

Prognosis of PACC tends to be better than that of ductal adenocarcinoma but worse than that of pancreatic endocrine tumors.[1,5] Surgery is still the best treatment for early local tumors. Radiotherapy and chemotherapy are generally used in patients with advanced and metastatic disease. There are no standard chemotherapeutic regimens for PACC, and data on response to chemotherapy are sourced from case reports.

There are case reports that FOLFOX, FOLFIRINOX, LV5FU2-Gemcitabine, LV5FU2-cisplatin, weekly paclitaxel, GEMOX, and other chemotherapeutic regimens show considerable efficacy against PACC.[9,10] In our case, gemcitabine plus albumin paclitaxel was combined with chemotherapy. After 6 cycles of chemotherapy, diffuse enlargement of the pancreas, as viewed under CT, was significantly smaller than before chemotherapy, and the common bile duct dilatation and gallbladder enlargement were also less pronounced than before. We conclude that gemcitabine plus albumin paclitaxel combined with chemotherapy is effective in PACC.

In conclusion, we report a case of PACC with diffuse enlargement of the pancreas. This rare and unique imaging manifestation may serve as a marker for clinical recognition of PACC. We also found that the chemotherapeutic regimen of gemcitabine combined with albumin paclitaxel is effective for PACC.

PATIENT'S CONSENT:
The patient provided written consent.

CONFLICT OF INTEREST:
The authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:
HL: Drafted this manuscript.
YW, XX, JZ: Data collection, collation, and analysis.
QL: Supervised and modified the article.

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