ACG CASE REPORTS JOURNAL

CASE REPORT | PANCREAS

Diffuse-Type Pancreatic Ductal Adenocarcinoma Mimicking Autoimmune Pancreatitis

Kenneth W. Chow, MD[1], Steve Hu, MD[1], Cameron Sikavi, MD[1], Matthew T. Bell, MD[1], Brandon Gisi, MD[1], Richard Chiu, DO[2], Caroline G. Yap, MD[2], Viktor Eysselein, MD[1], and Sofiya Reicher, MD[1]

[1]*Division of Gastroenterology & Hepatology, Harbor-UCLA Medical Center, Torrance, CA*
[2]*Department of Pathology, Harbor-UCLA Medical Center, Torrance, CA*

ABSTRACT

Pancreatic ductal adenocarcinoma (PDAC) classically presents as a solitary mass on cross-sectional imaging. Diffuse-type PDAC is an unusual variant that accounts for 1%–5% of PDACs. Owing to its rarity, there are no established radiographic or endosonographic definitions. We report a unique case of diffuse-type PDAC presenting with imaging findings of 2 distinct masses in the pancreatic head and tail and with endoscopic ultrasound findings of diffuse gland enlargement mimicking autoimmune pancreatitis. The case illustrates the importance of sampling several areas of the pancreas when diffuse enlargement is present on endoscopic ultrasound and multiple masses are seen on cross-sectional imaging.

KEYWORDS: pancreatic; adenocarcinoma; autoimmune; pancreatitis

INTRODUCTION

Pancreatic ductal adenocarcinoma (PDAC) is the fourth-leading cause of cancer death in the United States.[1] It classically presents as a solitary pancreatic mass on cross-sectional imaging. PDAC that presents with multiple pancreatic masses is rare, although cases of synchronous multifocal PDAC have been reported.[2,3] Diffuse-type PDAC (DTP) is an unusual variant that accounts for an estimated 1%–5% of PDACs.[4] Owing to its rarity, data on characteristic radiographic, endosonographic, or histologic findings of DTP are limited. Previous case reports describe presentations of DTP as either a solitary pancreatic mass or diffuse pancreatic enlargement without masses.[4,5] We report a unique case of DTP that presented with computed tomography (CT) findings of 2 distinct masses of the pancreatic head and tail and with endoscopic ultrasound (EUS) findings of diffuse gland enlargement without distinct masses mimicking autoimmune pancreatitis (AIP).

CASE REPORT

A 70-year-old woman with a history of papillary thyroid carcinoma, type 2 diabetes mellitus, gastrointestinal reflux disease, and lifelong tobacco use presented to her primary care physician for 2 weeks of severe epigastric pain. She was unsuccessfully trialed on a proton-pump inhibitor and soon developed night sweats and unintentional weight loss. Abdominal-pelvic CT showed 2 distinct lesions of the pancreatic tail and head measuring 2.3 × 1.6 cm and 1.6 × 1.0 cm, respectively, and a pancreas that was borderline prominent in size (Figure 1). A EUS demonstrated a hypoechoic sausage-shaped diffusely enlarged pancreas with duct dilation of 2.5 mm in the body; no discrete lesions were seen (Figures 2 and 3). Workup for AIP was negative with normal IgG4 levels, and EUS-guided fine-needle biopsy of the pancreatic head showed infiltrating adenocarcinoma (Figure 4). Her liver function tests were within normal limits, and tumor markers were notable for a CA 19-9 level of 8 U/mL, a carcinoembryonic antigen level of 21 ng/mL, and a chromogranin-A level of 607 ng/mL. A repeat CT scan showed interval enlargement of the pancreatic tail mass, and a positron emission tomography scan showed increased F-fluorodeoxyglucose activity in the corresponding regions of the pancreatic head and tail. A subsequent EUS-FNB of the pancreas tail returned with moderately differentiated invasive adenocarcinoma (Figure 5). Endoscopic retrograde cholangiopancreatography performed for

ACG Case Rep J 2023;10:e01070. doi:10.14309/crj.0000000000001070. Published online: June 10, 2023

Correspondence: Sofiya Reicher, MD (SReicher@dhs.lacounty.gov).

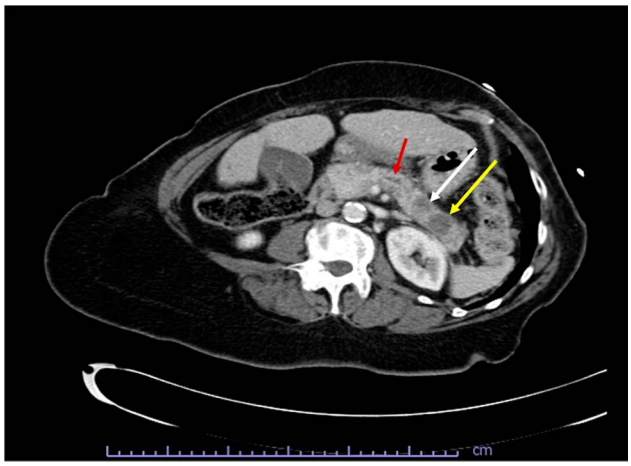

Figure 1. Axial-oblique abdominal-pelvic computed tomography demonstrates a low-density lesion in the pancreatic tail (yellow arrow) measuring 2.3 × 1.6 cm. An adjacent ill-defined low-density lesion (white arrow) measures 1.6 × 1.0 cm. The pancreas is borderline in size, and the pancreatic duct is prominent (red arrow).

worsening liver function tests demonstrated a 3 cm stricture of the distal common bile duct, which was managed with a fully covered metal stent. A multidisciplinary tumor board panel deemed the neoplasm to be borderline-resectable diffuse-type PDAC, and the patient was started on a neoadjuvant chemotherapy regimen of gemcitabine and abraxane in preparation for total pancreatectomy.

DISCUSSION

Multiple pancreatic masses are a rare disease presentation, seen in just 1.5% of all patients who underwent a dynamic enhanced CT pancreas protocol.[6] The differential for multiple pancreatic masses is broad and includes multifocal autoimmune pancreatitis, secondary metastases, and pancreatic neuroendocrine neoplasms. DTP is a rare manifestation of PDAC, and little is known about its presentation and endoscopic findings. A retrospective study by Choi et al examined 14 cases of DTP and described radiologic features of a peripheral capsule-like structure, the absence of intratumor pancreatic duct dilatation, and lack of pancreatic parenchymal atrophy.[7] CT showed a solitary tumor in 79% of patients and a tumor involving virtually the entire pancreas in the remaining 21% of patients. No patients presented with multiple pancreatic masses. The pathogenesis of DTP remains unclear; it could stem from progression of a focal PDAC or could result from synchronous multifocal tumor development. Our case report provides support to the latter hypothesis given the distinct masses seen on cross-sectional imaging, suggesting a multifocal etiology. In addition, there are no documented cases that demonstrate progression of a focal PDAC to DTP, suggesting that DTP is not simply a late presentation of classic PDAC. Interestingly, in our case, there were no distinct masses seen on EUS and biopsies were sampled from the general region of the pancreatic head and tail.

Diffuse pancreatic enlargement is a much more specific finding and is classically associated with diffuse-type AIP, although pancreatic neuroendocrine tumors can rarely present with diffuse glandular enlargement.[8] Autoimmune pancreatitis is classified into 2 radiologic subtypes: diffuse-type and focal-type AIP.[9] Diffuse-type AIP is more common and is associated with sausage-like pancreatic enlargement with loss of the pancreatic lobular contours and clefts. Focal-type AIP typically presents as a distinct mass, most commonly in the pancreatic head, and can resemble PDAC on imaging.[10] Although rare, clinicians should be aware of DTP and differentiate it from autoimmune pancreatitis, which entails a drastically different treatment course and prognosis. Biopsy should be performed to rule out malignancy in cases of suspected autoimmune pancreatitis before initiating treatment, even if no masses are seen.

Our case of a patient with DTP was a unique presentation with features of multiple distinct masses in one imaging modality and diffuse gland enlargement without masses in another,

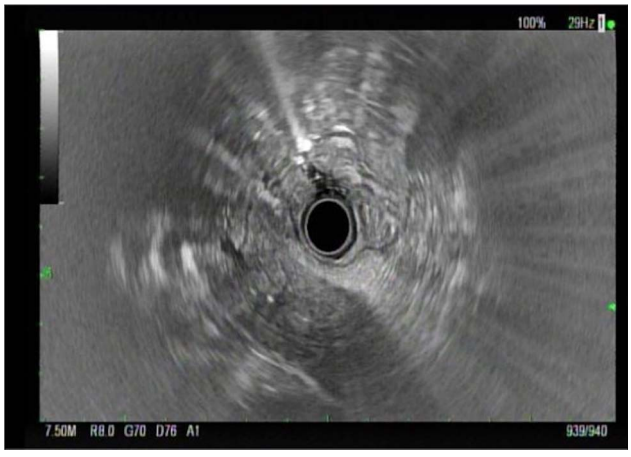

Figure 2. Endoscopic ultrasound demonstrates a diffusely enlarged pancreas, with the head, body, and tail displaying a diffusely hypoechoic sausage-like appearance.

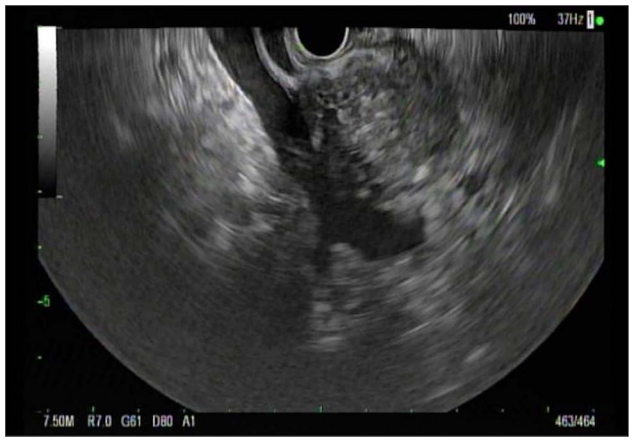

Figure 3. Endoscopic ultrasound demonstrates focal prominence of the pancreatic head with loss of interface with the portal vein-superior mesenteric vein confluence.

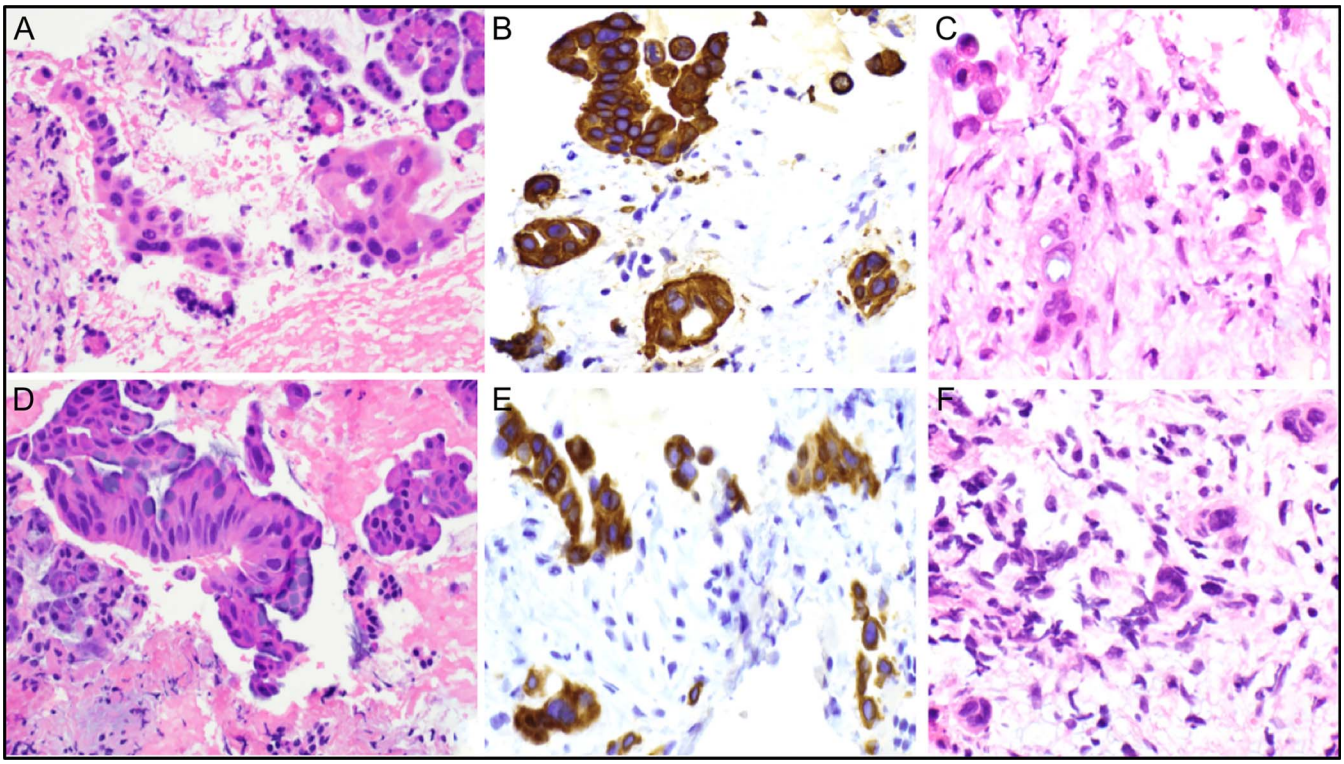

Figure 4. Fine-needle biopsy of the pancreatic head mass. Infiltrative singly dispersed and well-formed to cribriforming acini are irregularly embedded within the desmoplastic stroma (A, C, D, F). The tumor cells show positive staining for CK19 (B) and IMP3 (E).

although variances in imaging findings have been described in the literature. DTP is a rare phenomenon and likely under-reported in the literature. The entire pancreas must be examined during evaluation for PDAC, especially if there is discordance between imaging modalities. The case illustrates the importance of sampling several areas of the pancreas when diffuse enlargement is present on EUS and multiple pancreatic masses are seen on cross-sectional imaging.

DISCLOSURES

Author contributions: Study concept and design (S. Reicher); acquisition of data (KW Chow); analysis and interpretation of data (KW Chow, S. Hu, C. Sikavi, MT Bell, B. Gisi, R. Chiu, CG Yap, S. Reicher); drafting of the manuscript (KW Chow); critical revision of the manuscript for important intellectual content (all); administrative, technical, or material support (S. Reicher and V. Eysselein); study supervision (S. Reicher). All authors approved the final draft submitted. S. Reicher is the article guarantor.

Financial disclosure: None to report. All authors report no conflict of interest.

Prior presentation: American College of Gastroenterology Annual Meeting 2022; November 4, 2022; Charlotte, North Carolina.

Informed consent was obtained for this case report.

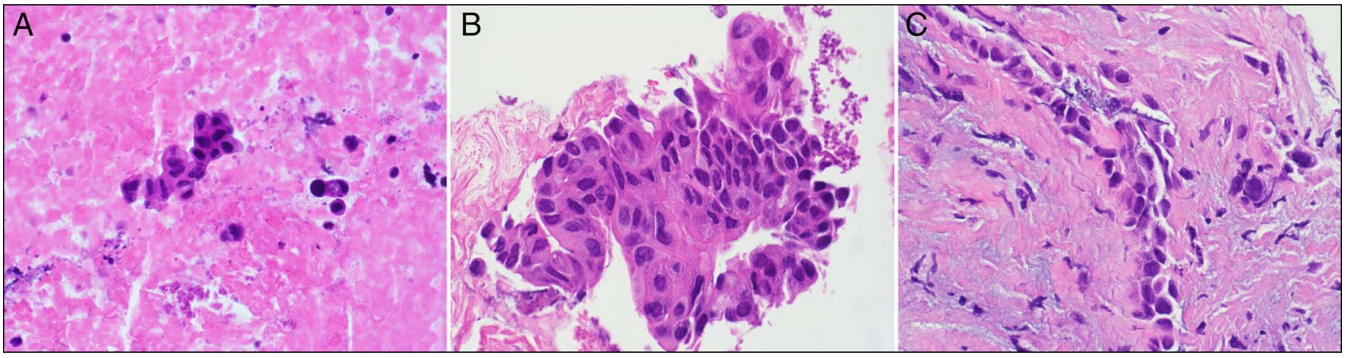

Figure 5. Fine-needle biopsy of the pancreatic tail mass reveals adenocarcinoma cells occurring singly or in small clusters (A) or in glandular formation (B) and infiltrating in the desmoplastic stroma (C).

Received November 13, 2022; Accepted May 10, 2023

REFERENCES

1. Puckett Y, Garfield K. *Pancreatic Cancer*. StatPearls Publishing: Orlando, FL, 2022.
2. Goong HJ, Moon JH, Choi HJ, et al. Synchronous pancreatic ductal adenocarcinomas diagnosed by endoscopic ultrasound-guided fine needle biopsy. *Gut Liver*. 2015;9(5):685–8.
3. Izumi S, Nakamura S, Mano S, Suzuka I. Resection of four synchronous invasive ductal carcinomas in the pancreas head and body associated with pancreatic intraepithelial neoplasia: Report of a case. *Surg Today*. 2009;39(12):1091–7.
4. Miyoshi H, Kano M, Kobayashi S, et al. Diffuse pancreatic cancer mimicking autoimmune pancreatitis. *Intern Med*. 2019;58(17):2523–7.
5. Nguyen HQ, Pham NTT, Hoang VT, Van HAT, Huynh C, Hoang DT. Diffuse pancreatic carcinoma with hepatic metastases. In: Cives M (ed). *Case Reports in Oncological Medicine*, Vol 2020. Hindawi Limited: London, 2020, pp 1–5.
6. Zhu L, Dai Mh, Wang St, et al. Multiple solid pancreatic lesions: Prevalence and features of non-malignancies on dynamic enhanced CT. *Eur J Radiol*. 2018;105:8–14.
7. Choi YJ, Byun JH, Kim JY, et al. Diffuse pancreatic ductal adenocarcinoma: Characteristic imaging features. *Eur J Radiol*. 2008;67(2):321–8.
8. Santes O, Morales-Maza J, Domínguez-Rosado I. Diffuse enlargement of the pancreas: An unusual radiologic presentation of a pancreatic neuroendocrine tumor. *Clin Gastroenterol Hepatol*. 2017;15(11):e165–e166.
9. Tabata T, Kamisawa T, Takuma K, Hara S, Kuruma S, Inaba Y. Differences between diffuse and focal autoimmune pancreatitis. *World J Gastroenterol*. 2012;18(17):2099–104.
10. Wakabayashi T, Kawaura Y, Satomura Y, et al. Clinical and imaging features of autoimmune pancreatitis with focal pancreatic swelling or mass formation: Comparison with so-called tumor-forming pancreatitis and pancreatic carcinoma. *Am J Gastroenterol*. 2003;98(12):2679–87.