Images in Clinical Medicine

Pancreatic carcinoma disguised as type 1 autoimmune pancreatitis with a mass-forming appearance: a clinical dilemma

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In December 2022, a 69-year-old male was presented to the local hospital due to upper abdominal discomfort, gradually developing jaundice with itching, dark urine, and pale-colored stools, with symptoms first appearing ten months prior. Laboratory examinations revealed alanine aminotransferase (ALT) at 507 U/L, aspartate aminotransferase (AST) at 528 U/L, total bilirubin (TBIL) at 226.5 μmol/L, and direct bilirubin (DBIL) at 135.2 μmol/L. Significantly, IgG4 was found at 22,390 mg/L, and CA19-9 at 2,736 U/mL, both values exceeding the upper limit of normal; other serum tumor biomarkers like CEA were normal with around 4.2 ng/mL of <5.0 ng/mL. A chest-abdomen-pelvis contrast-enhanced computed tomography (CECT) displayed a 26-mm-diameter soft-tissue mass in the uncinate process of the pancreas with delayed enhancement, accompanied by dilation in both pancreatic ducts. Additionally, both intra- and extra-hepatic bile ducts and the common bile duct (CBD) were dilated. Percutaneous transhepatic cholangial drainage (PTCD) was performed to alleviate discomfort and bile pressure. Endoscopic ultrasound (EUS) confirmed the aforementioned findings, revealing a hypoechoic lesion with irregular margins and bile duct thickening. Fine-needle aspiration (FNA) also demonstrated that histiocytes were the predominant components with a small number of lymphocytes, with no atypical cells in either the smear or biopsy. Immunohistochemical staining indicated that CK was positive and Ki-67 was negative; regrettably, IgG and IgG4 were not performed. Consequently, the patient was diagnosed with a mass-forming type of type 1 autoimmune pancreatitis (AIP), and a steroid trial of prednisone at 35 mg/day was administered for further clarification. This treatment not only partially alleviated the clinical symptoms and signs but also reduced the IgG4 levels progressively from 18,407 to 11,903 mg/L. However, the imaging reassessment in March 2023 indicated that the pancreatic mass remained largely unchanged without any significant alterations.

The patient was transferred to our hospital, and the laboratory examinations remained unremarkable, except for elevated IgG4 at 11,682 mg/L and CA19-9 at 1,229 U/mL. A repeated abdominal CECT showed results nearly identical to the previous examination, demonstrating that the mass was hypovascular, with heterogeneous and mild enhancement in the portal and delayed phases and significant thickening of the gallbladder wall (*Figure 1A*,*1B*). Fluorodeoxyglucose (FDG)-positron emission tomography (PET)/CT revealed increased radioactive uptake of the mass, with an SUVmax of 4.2, surrounded by numerous

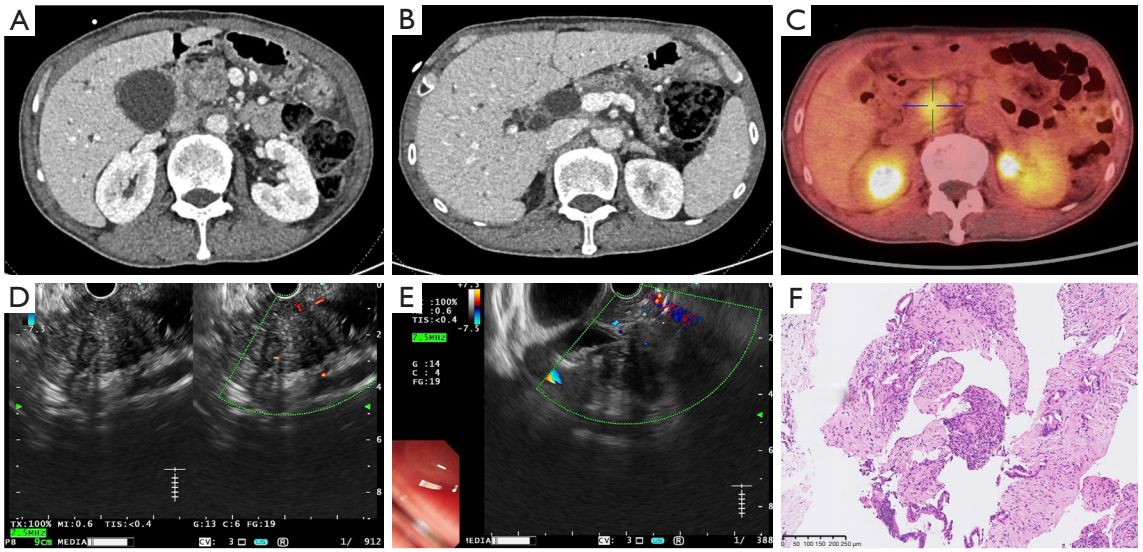

Figure 1 Preoperative imaging and histopathological examinations of biopsy. (A,B) Abdominal CECT-scan axial view; (C) FDG-PET/CT view; (D,E) EUS view; (F) histopathological examination of FNB specimen with hematoxylin-eosin staining. CECT, contrasted-enhanced computed tomography; FDG-PET/CT, fluorodeoxyglucose positron emission tomography/computed tomography; EUS, endoscopic ultrasound; FNB, fine-needle biopsy.

small lymph nodes (*Figure 1C*). Some lymph nodes distributed in the mediastinum, hilus of the lung, and right cardio-phrenic angle were plump and enlarged, with slightly higher radioactive uptake. Paradoxically, a repeated EUS displayed an uneven hypoechoic mass with scattered, flaky hyper-echoes; the upstream pancreatic duct was severely dilated, up to 10 mm. Interestingly, both the gallbladder wall and the CBD were diffusely thickened (nearly 2 mm) and exhibited an onion skin appearance; the upstream and downstream regions of the bile duct were dilated (up to 13 mm) and narrow, respectively (*Figure 1D*,*1E*). Unexpectedly, the histopathology of the specimens obtained from fine-needle biopsy (FNB) via 22G Acquire™ (Boston Scientific, Marlborough, MA, USA) revealed moderately differentiated adenocarcinoma and masses of plasmacytic infiltration with fibrosis; the immunohistochemical staining was positive for both IgG and IgG4 (*Figure 1F*).

Laparoscopic radical pancreaticoduodenectomy was performed on the patient (*Figure 2A*). The histopathological examination revealed moderate to poorly differentiated adenocarcinoma, accompanied by manifestations of chronic pancreatitis, including parenchymal atrophy and lymphoid and fibrous hyperplasia. Furthermore, signs of both chronic cholecystitis and chronic inflammation of the bile duct wall were evident (*Figure 2B*). Immunohistochemical staining demonstrated CD138 positivity and a ratio of IgG4 to IgG greater than 40% (*Figure 2C*,*2D*). Ultimately, the patient was diagnosed with pancreatic carcinoma. Following the completion of molecular pathological examinations, chemoradiation and immunotherapy will be administered.

The widely accepted viewpoint is that chronic pancreatitis is a risk factor for pancreatic carcinoma. However, when considering AIP, especially type 1, which represents a distinct form of pancreatitis with autoimmunity as the predominant mechanism, the relationship between AIP and pancreatic carcinoma remains unclear. This relationship needs further clarification, notwithstanding some published case reports. The urgent requirement to explore the pathophysiological basis of these two diseases, especially their coexistence, calls for in-depth study. Type 1 AIP and pancreatic carcinoma can often present with similar characteristics, and while EUS, accompanied by biopsy, provides localized examination, only a surgical specimen pathology can definitively determine whether it represents a mimicry or a complication. Such determination is crucial for prognostic evaluation and subsequent treatment strategies.

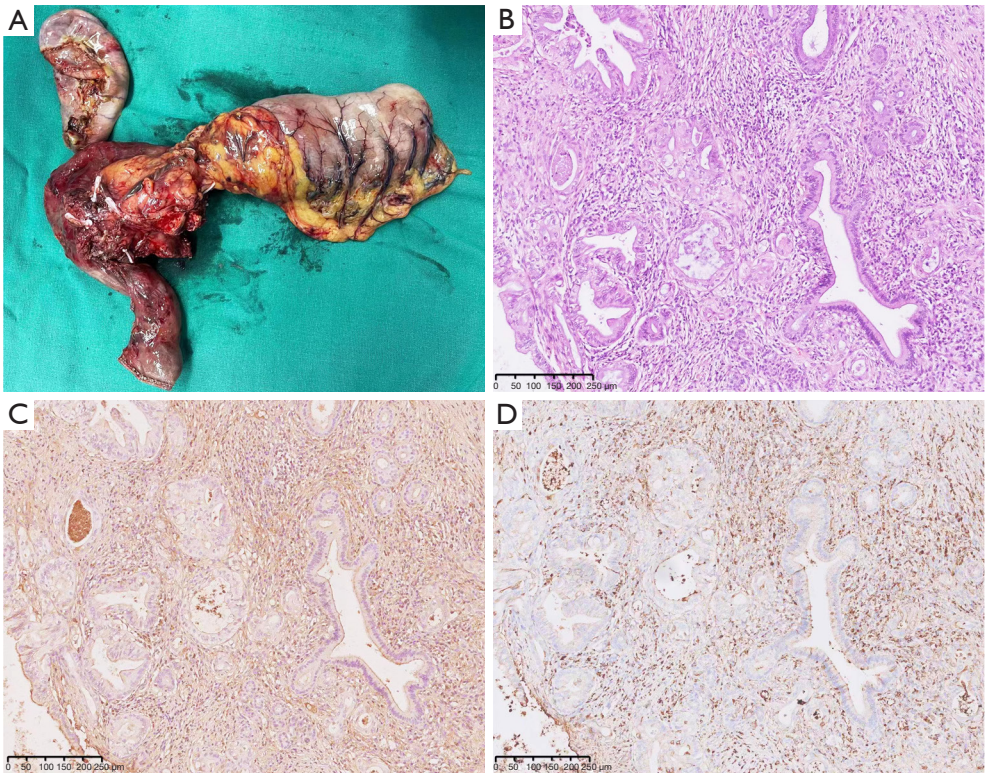

Figure 2 Gross specimen of laparoscopic radical pancreaticoduodenectomy and histopathological examinations. Operative specimen picture (A) and its histopathological examination with hematoxylin-eosin staining (B), IgG (C), and IgG4 (D).

Consequently, even for patients diagnosed with type 1 AIP, heightened vigilance and rigorous follow-up are essential when any signs suggestive of malignancy emerge to prevent potential misdiagnoses.

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Footnote

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Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. Written informed consent was obtained from the patient for publication of this manuscript and any accompanying image resources.

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