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Type 1 Autoimmune Pancreatitis Mimicking Pancreatic Malignancy: A Clinical Case

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ABSTRACT

Autoimmune pancreatitis is a rare form of chronic pancreatitis that can mimic pancreatic malignancy. We report the case of a 55-year-old male presenting with obstructive jaundice and abdominal discomfort. Imaging studies, including ultrasonography and contrast-enhanced computed tomography, revealed a prominent pseudomass in the pancreatic head. However, a narrowed pancreatic duct and diffusely enlarged pancreas were observed, which contrast with typical findings of malignancy. Additionally, the pancreas exhibited a capsule-like rim and delayed contrast enhancement. Notably, extrapancreatic findings, such as biliary ductal thickening, peripancreatic lymphadenopathy, and retroperitoneal fibrosis, were pivotal in steering the diagnosis toward type 1 autoimmune pancreatitis. The diagnosis was made based on the constellation of imaging findings, the favorable clinical response to corticosteroid therapy, and elevated serum IgG4 levels. This case highlights the importance of recognizing both pancreatic and extrapancreatic manifestations of autoimmune pancreatitis to facilitate early and accurate diagnosis.

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Introduction

The concept of autoimmune pancreatitis (AIP) has evolved significantly since its first mention in 1961. In 1991, Kawaguchi et al. described lymphoplasmacytic sclerosing pancreatitis (LPSP), which became the pathological basis for AIP, characterized by lymphoplasmacytic infiltration, storiform fibrosis, and obliterative phlebitis [1]. In 1995, Yoshida et al. formally proposed the concept of AIP [2], and in 2001, Hamano et al. identified elevated serum IgG4 levels as a key marker in Japanese patients, leading to the recognition of IgG4-related disease (IgG4-RD) [3].

Parallel to these developments, Western countries reported a distinct subtype of AIP, characterized by idiopathic duct-centric pancreatitis (IDCP) or AIP with granulocyte epithelial lesions (AIP with GEL) [4]. The 2011 International Consensus Diagnostic Criteria (ICDC) formalized the classification of AIP into two subtypes: type 1 (LPSP) and type 2 (IDCP) [5]. The Japan Pancreas Society (JPS) adapted these criteria for broader use, resulting in the 2011 Japanese Clinical Diagnostic Criteria for AIP, which were later revised in 2018 to incorporate advancements in imaging and simplify the diagnostic process [6]. The Mayo Clinic HISORt Criteria [7] provides a more clinical approach, with flexible criteria allowing diagnosis based on treatment response and clinical presentation, making it particularly useful in the absence of biopsies or definitive markers. These revisions continue to refine the approach to diagnosing and managing AIP worldwide.

The ICDC identifies five cardinal features for diagnosing type 1 AIP. One of these is pancreatic parenchymal imaging using computed tomography (CT) or magnetic resonance (MR), where typical findings include diffuse pancreatic enlargement with delayed enhancement and a characteristic "capsule-like" rim. Imaging of the pancreatic duct through endoscopic retrograde cholangiopancreatography (ERCP) or magnetic resonance cholangiopancreatography (MRCP) is also a key feature, often revealing long or multifocal strictures of the duct without upstream dilation [5].

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Case Report:

A 55-year-old male with no significant past medical history was referred to the radiology department for abdominal ultrasonography (US) following elevated bilirubin levels (total bilirubin: 5.8 mg/dL, direct bilirubin: 4.2 mg/dL) and a two-month history of intermittent abdominal pain and mild fatigue. The patient reported no significant weight loss, fever, or changes in bowel habits. Physical examination revealed mild epigastric tenderness, and laboratory tests showed mildly elevated alkaline phosphatase and transaminase levels. Serum amylase and lipase levels were within normal limits.

Imaging Findings:

Abdominal ultrasonography (US) revealed a dilated intrahepatic and extrahepatic biliary tract without evidence of stones or biliary tumors, findings suggestive of distal biliary obstruction (Figure 1A). Additionally, the pancreas appeared diffusely enlarged and hypochoic, with surrounding peripancreatic echogenic fat stranding (Figure 1B).

Based on the US findings, a contrast-enhanced computed tomography (CECT) was performed. The CECT demonstrated a prominent pancreatic head pseudomass encasing the superior mesenteric vein and displaying homogeneous high density in the venous phase (Figure 2). No upstream pancreatic duct dilation was observed, and the pancreas exhibited a diffusely enlarged, "sausage-like" appearance (Figure 3). Additionally, the pancreas displayed a characteristic capsule-like rim and arterial hypoenhancement with delayed reinforcement in the venous phase (Figure 4), which are typical imaging features of type 1 autoimmune pancreatitis (AIP).

Further extrapancreatic findings were observed, often indicative of systemic involvement in type 1 AIP. These included diffuse thickening of the biliary tract wall (Figure 5), markedly enlarged peripancreatic lymph nodes (Figure 6), circumferential perivascular fat stranding in the retroperitoneum, and a mesenteric pseudotumor (Figure 7). In the pelvis, a right periureteral soft tissue mass caused mild dilation of the ureter

(Figure 8), a feature consistent with retroperitoneal fibrosis, commonly associated with type 1 AIP.

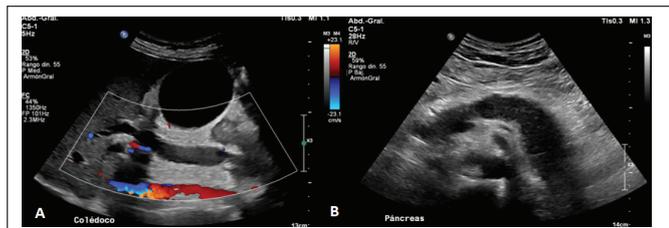


Figure 1: Abdominal US showing a dilated extrahepatic biliary duct without any filling defects (A). Hypoechoic pancreas with diffuse enlargement and echogenic peri-pancreatic fat stranding (B).

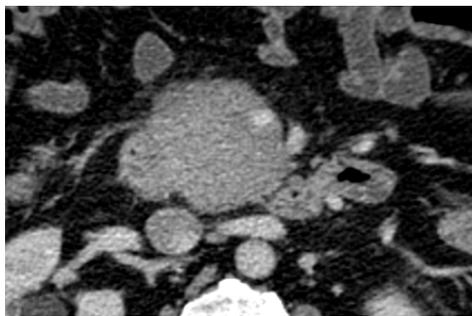


Figure 2: Venous phase CECT showing an homogeneous pancreatic head pseudomass encasing superior mesenteric vein.

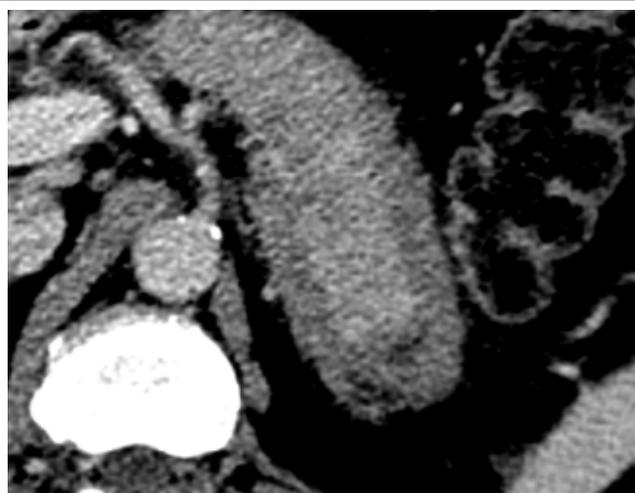


Figure 3: Pancreatic 'sausage-like' enlargement with a non-visible pancreatic duct.

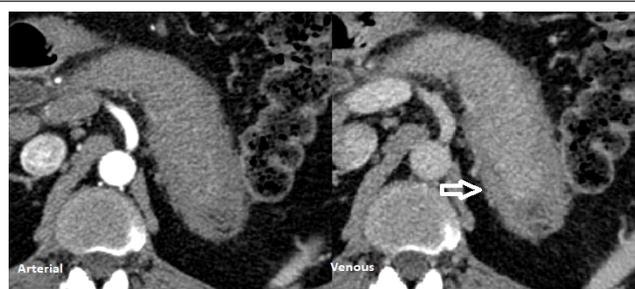


Figure 4: Arterial and venous phase CECT demonstrating a hypoenhancement pancreas with late venous reinforcement. Peri-pancreatic capsule-like rim of low attenuation (arrow).

Outcome:

The patient was initiated on a 2-week course of oral corticosteroid therapy with prednisone. This treatment resulted in marked clinical improvement, including resolution of abdominal pain and jaundice, as well as normalization of liver function tests, specifically bilirubin

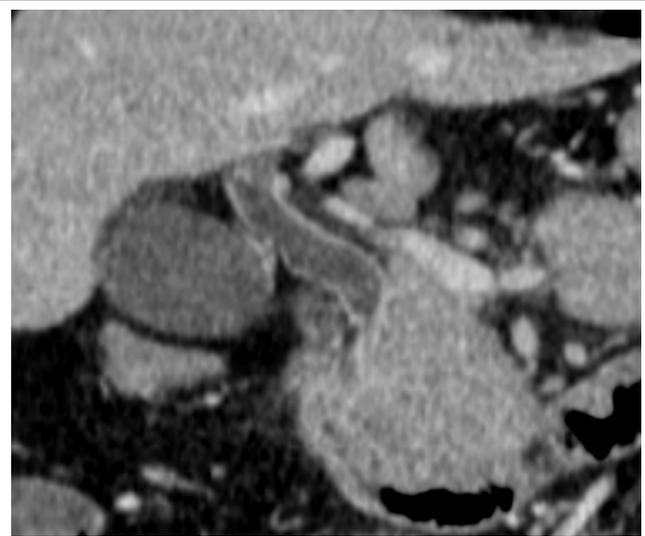


Figure 5: Venous phase CECT showing a biliary tract wall thickening.



Figure 6: Markedly enlarged peri-pancreatic lymph nodes (arrows).

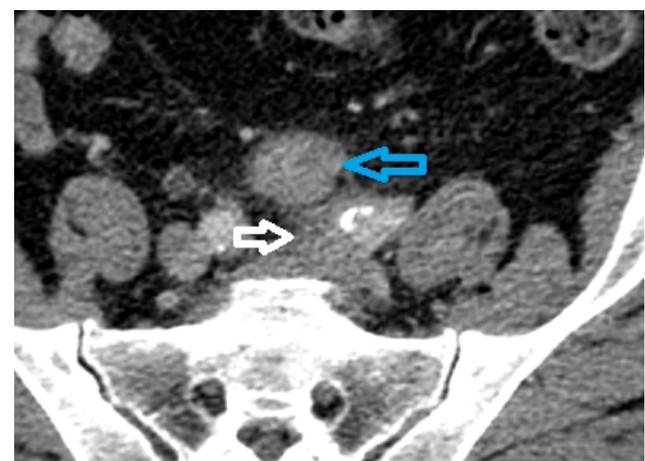


Figure 7: Retroperitoneal perivascular fat stranding (white arrow) associated with mesenteric pseudotumor (blue arrow).

and alkaline phosphatase levels. Initial laboratory evaluation revealed significantly elevated serum IgG4 levels at 280 mg/dL (reference range: <135 mg/dL). Following corticosteroid therapy, these levels markedly decreased to 90 mg/dL, reflecting a positive treatment response. A follow-up ultrasound after treatment showed substantial reduction in pancreatic and biliary abnormalities, including decreased dilation of both intrahepatic and extrahepatic biliary tracts. The combination of clinical, laboratory, and imaging improvements strongly supported the diagnosis of type 1 autoimmune pancreatitis (AIP).

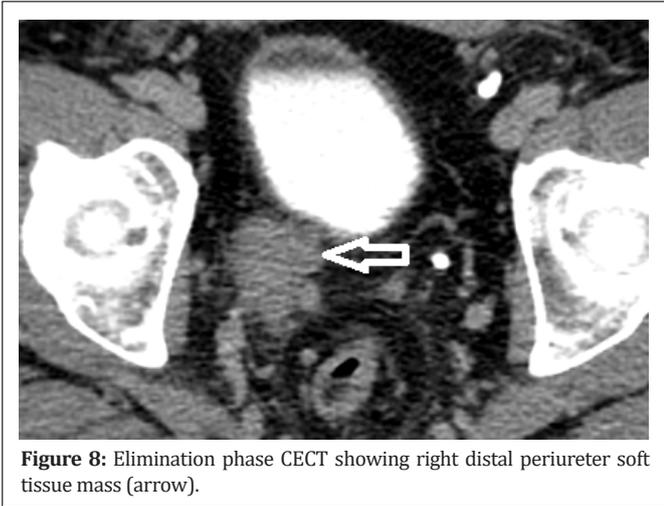


Figure 8: Elimination phase CECT showing right distal periureter soft tissue mass (arrow).

Discussion:

According to the criteria outlined by the International Consensus Diagnostic Criteria (ICDC), pancreatic findings in type 1 autoimmune pancreatitis (AIP) can be divided into typical findings (Level 1) and indeterminate findings (Level 2). Our case exemplifies typical findings for type 1 AIP, including a “capsule-like” rim, delayed enhancement, and diffuse enlargement. In such scenarios, the ICDC suggests collateral evidence to support a definitive diagnosis. This can be achieved through histology, serology, steroid responsiveness, or evidence of other organ involvement (OOI). OOI is particularly relevant as it can be confirmed through histology, imaging, or clinical findings [5].

Level 1 radiologic findings for OOI include segmental or multifocal strictures of the distal or proximal bile duct and retroperitoneal fibrosis. Level 2 findings include symmetric enlargement of the salivary/lacrimal glands and radiologic involvement of the kidneys [5]. In this case, Level 1 findings were identified in both the pancreas and other organs, strongly supporting the diagnosis and avoiding the need for invasive procedures. It is worth noting that although the pancreatic duct was not considered a diagnostic criterion in this case and was not visualized through ERCP or MRCP, the absence of ductal dilation is a supportive finding for the diagnosis.

Additionally, type 2 AIP was excluded as a differential diagnosis, given the absence of extra-pancreatic involvement and its common association with inflammatory bowel disease.

Imaging

The enhancement patterns of AIP provide critical diagnostic clues. During the pancreatic phase, the mean CT attenuation of AIP focal masses (71 HU) is comparable to pancreatic carcinoma (59 HU), making differentiation challenging in this phase. However, in the hepatic phase, AIP masses show significantly higher attenuation (90 HU) compared to carcinoma (64 HU) [8]. This delayed enhancement in AIP reflects its unique inflammatory characteristics and contrasts with the hypovascular nature of carcinoma.

A capsule-like rim of low attenuation surrounding the pancreas is another distinguishing feature of autoimmune pancreatitis, observed in approximately 12%–40% of cases [9]. This hypoattenuating rim is noted for its high specificity, with reported values ranging between 93% and 99% [10].

Regarding pancreatic duct imaging in AIP, the characteristic “duct-penetrating” sign is defined by a narrowed pancreatic duct passing through a pancreatic mass without total obstruction or upstream dilation [11].

Type 1 AIP is frequently associated with extrapancreatic involvement, which can manifest in various organs. The most commonly affected site is the biliary tract, with approximately 88% of patients showing biliary involvement [12]. Other frequently involved organs include the salivary glands, retroperitoneum, and kidneys. Retroperitoneal fibrosis occurs in about 20% of AIP cases and presents as soft tissue encasing the abdominal aorta or ureters, which can lead to ureteral obstruction and hydronephrosis [13]. Retroperitoneal involvement is steroid-responsive, and imaging post-treatment often shows a reduction in the fibrotic tissue [14].

CECT frequently demonstrates peripancreatic lymphadenopathy in AIP cases [15]. This is an important distinguishing factor, as pancreatic ductal adenocarcinoma can also present with peripancreatic lymph node involvement, but the nodes are more likely to exhibit necrosis or irregular enhancement patterns [16].

Conclusion

Type 1 autoimmune pancreatitis (AIP) is a systemic condition that affects both the pancreas and multiple extrapancreatic organs. Its characteristic imaging features, such as diffuse pancreatic enlargement and the presence of a capsule-like rim, along with steroid responsiveness, are key to differentiating AIP from other pathologies. Extrapancreatic manifestations, particularly biliary and retroperitoneal involvement, further support the diagnosis of AIP and help avoid unnecessary surgeries or misdiagnosis.

Conflict of Interest: The authors declare no conflicts of interest.

Ethical Consideration: Not Required.

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