CASE REPORT - OPEN ACCESS

ISSN: 3067-2368 American Journal of Case Reports and Clinical Images

Contents lists available at https://amejcaserepclinimag.com/

American Journal of Case Reports and Clinical Images

Type 1 Autoimmune Pancreatitis Mimicking Pancreatic Malignancy: A Clinical Case

Dr. Diego Salinas Rodríguez, Dr. Guillermo Elizondo Riojas

Department of Radiology and Imaging, Hospital Universitario "Dr. José Eleuterio González", Universidad Autónoma de Nuevo León, Monterrey, México

ARTICLE INFO

Article history:
Received: 25-08-2025
Revised: 03-09-2025
Accepted: 07-09-2025
Published: 12-09-2025

KEYWORDS:
Autoimmune Pancreatitis
Pancreatic Pseudotumor
IgG4-related Disease

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.

Copyrights © 2025, Dr. Diego Salinas Rodríguez. This article is licensed under the Creative Commons Attribution-Non-Commercial-4.0-International-License-(CCBY-NC) (https://amejcaserepclinimag.com//blogpage/copyright-policy). Usage and distribution for commercial purposes require written permission.

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].

* Corresponding author.
Dr. Diego Salinas Rodríguez, Department of Radiology and Imaging, Hospital Universitario "Dr. José Eleuterio González" Universidad Autónoma de Nuevo León, Monterrey, México

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 hypoechoic, 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.

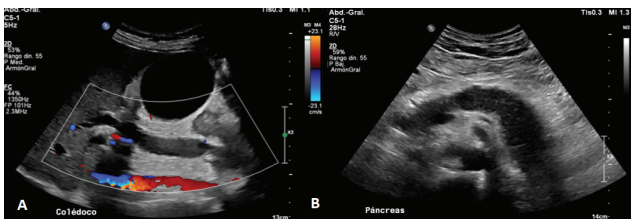

Figure 1: Abdominal US showing a dilatated extrahepatic biliary duct without any filling defects (A). Hypoecoic pancreas with diffuse enlargement and ecogenic peri-pancreatic fat stranding (B).

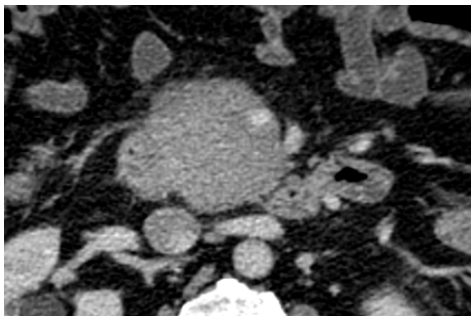

Figure 2: Venous phase CECT showing an homogeneous pancreatic head pseudomass encasing superior mesenteric vein.

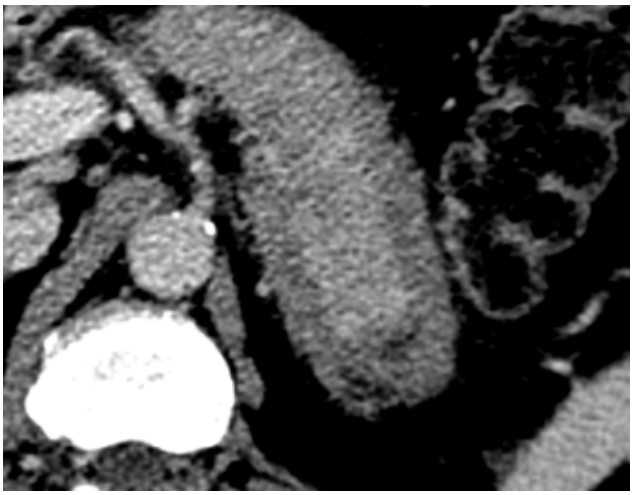

Figure 3: Pancreatic 'sausage-like' enlargement with a non-visible pancreatic duct.

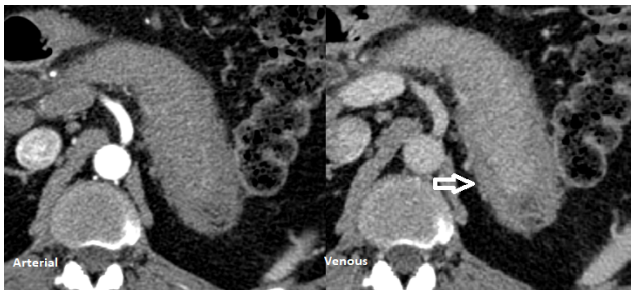

Figure 4: Arterial and venous phase CECT demostrating an hypoenhancement pancreas with late venous reinforcement. Peri-pancreatic capsule-like rim of low attenuation (arrow).

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 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).

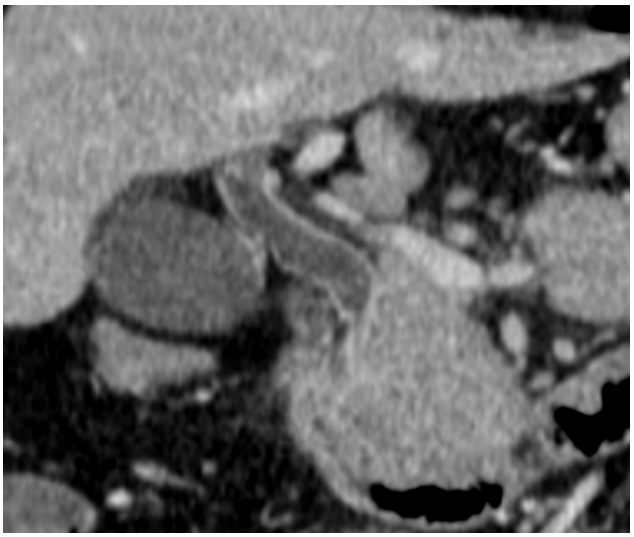

Figure 5: Venous phase CECT showing a biliary tract wall thickening.

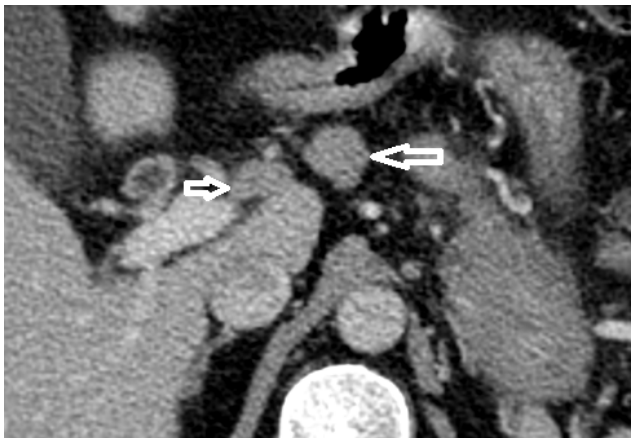

Figure 6: Markedly enlarged peri-pancreatic lymph nodes (arrows).

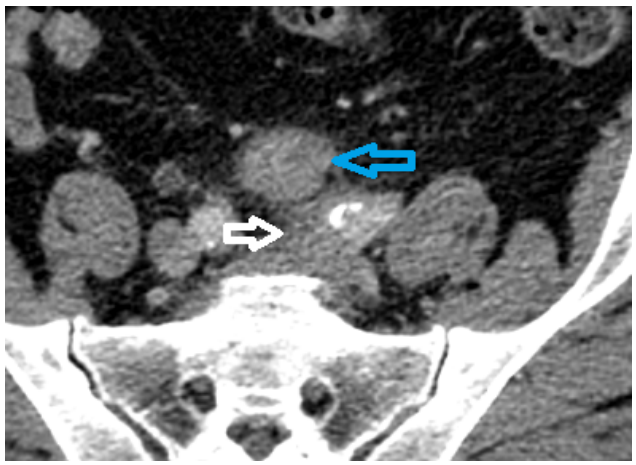

Figure 7: Retroperitoneal perivascular fat stranding (white arrow) associated with mesenteric pseudotumor (blue arrow).

Diego Salinas Rodríguez, Guillermo Elizondo Riojas (2025). Type 1 Autoimmune Pancreatitis Mimicking Pancreatic Malignancy: A Clinical Case. American J Case Rep Clin Imag. 2025; September, 2(2),01-04.

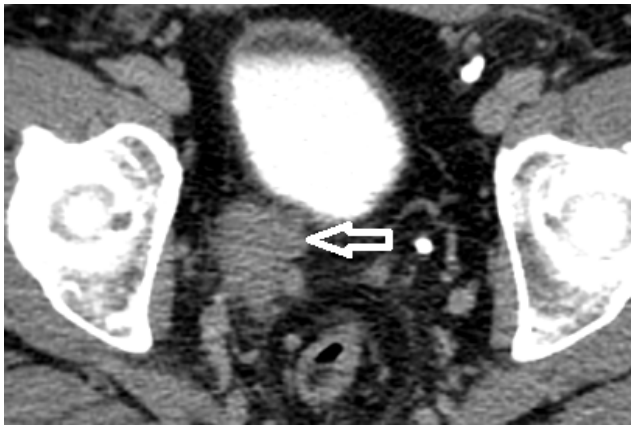

Figure 8: Elimination phase CECT showing right distal periureter soft tissue mass (arrow).

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.

References

1. Kawaguchi K, Koike M, Tsuruta K, Okamoto A, Tabata I, Fujita N. Lymphoplasmacytic sclerosing pancreatitis with cholangitis: a variant of primary sclerosing cholangitis extensively involving pancreas. Hum Pathol. 1991 Apr;22(4):387-95. doi: 10.1016/0046-8177(91)90087-6.
2. Yoshida K, Toki F, Takeuchi T, Watanabe S, Shiratori K, Hayashi N. Chronic pancreatitis caused by an autoimmune abnormality. Proposal of the concept of autoimmune pancreatitis. Dig Dis Sci. 1995 Jul;40(7):1561-8. doi: 10.1007/BF02285209.
3. Hamano H, Kawa S, Horiuchi A, Unno H, Furuya N, Akamatsu T, Fukushima M, Nikaido T, Nakayama K, Usuda N, Kiyosawa K. High serum IgG4 concentrations in patients with sclerosing pancreatitis. N Engl J Med. 2001 Mar 8;344(10):732-8. doi: 10.1056/NEJM200103083441005.
4. Notohara K, Burgart LJ, Yadav D, Chari S, Smyrk TC. Idiopathic chronic pancreatitis with periductal lymphoplasmacytic infiltration: clinicopathologic features of 35 cases. Am J Surg Pathol. 2003 Aug;27(8):1119-27. doi: 10.1097/00000478-200308000-00009.
5. Shimosegawa T, Chari ST, Frulloni L, Kamisawa T, Kawa S, Mino-Kenudson M, Kim MH, Klöppel G, Lerch MM, Löhr M, Notohara K, Okazaki K, Schneider A, Zhang L; International Association of Pancreatology. International consensus diagnostic criteria for autoimmune pancreatitis: guidelines of the International Association of Pancreatology. Pancreas. 2011 Apr;40(3):352-8. doi: 10.1097/MPA.0b013e3182142fd2.
6. Kawa S, Kamisawa T, Notohara K, Fujinaga Y, Inoue D, Koyama T, Okazaki K. Japanese Clinical Diagnostic Criteria for Autoimmune Pancreatitis, 2018: Revision of Japanese Clinical Diagnostic Criteria for Autoimmune Pancreatitis, 2011. Pancreas. 2020 Jan;49(1):e13-e14. doi: 10.1097/MPA.0000000000001443.
7. Chari ST, Smyrk TC, Levy MJ, Topazian MD, Takahashi N, Zhang L, Clain JE, Pearson RK, Petersen BT, Vege SS, Farnell MB. Diagnosis of autoimmune pancreatitis: the Mayo Clinic experience. Clin Gastroenterol Hepatol. 2006 Aug;4(8):1010-6; quiz 934. doi: 10.1016/j.cgh.2006.05.017.
8. Takahashi N, Fletcher JG, Hough DM, Fidler JL, Kawashima A, Mandrekar JN, Chari ST. Autoimmune pancreatitis: differentiation from pancreatic carcinoma and normal pancreas on the basis of enhancement characteristics at dual-phase CT. AJR Am J Roentgenol. 2009 Aug;193(2):479-84. doi: 10.2214/AJR.08.1883.
9. Irie H, Honda H, Baba S, Kuroiwa T, Yoshimitsu K, Tajima T, Jimi M, Sumii T, Masuda K. Autoimmune pancreatitis: CT and MR characteristics. AJR Am J Roentgenol. 1998 May;170(5):1323-7. doi: 10.2214/ajr.170.5.9574610.
10. Yoon SB, Jeon TY, Moon SH, Shin DW, Lee SM, Choi MH, Min JH, Kim MJ. Differentiation of autoimmune pancreatitis from pancreatic adenocarcinoma using CT characteristics: a systematic review and meta-analysis. Eur Radiol. 2023 Dec;33(12):9010-9021. doi: 10.1007/s00330-023-09959-5.
11. Ichikawa T, Sou H, Araki T, Arbab AS, Yoshikawa T, Ishigame K, Haradome H, Hachiya J. Duct-penetrating sign at MRCP: usefulness for differentiating inflammatory pancreatic mass from pancreatic

Diego Salinas Rodríguez, Guillermo Elizondo Riojas (2025). Type 1 Autoimmune Pancreatitis Mimicking Pancreatic Malignancy: A Clinical Case. American J Case Rep Clin Imag. 2025; September, 2(2),01-04.

carcinomas. Radiology. 2001 Oct;221(1):107-16. doi: 10.1148/radiol.2211001157.

12. Nishino T, Toki F, Oyama H, Oi I, Kobayashi M, Takasaki K, Shiratori K. Biliary tract involvement in autoimmune pancreatitis. Pancreas. 2005 Jan;30(1):76-82. PMID: 15632703.

13. Takahashi N, Kawashima A, Fletcher JG, Chari ST. Renal involvement in patients with autoimmune pancreatitis: CT and MR imaging findings. Radiology. 2007 Mar;242(3):791-801. doi: 10.1148/radiol.2423060003

14. Manfredi R, Graziani R, Cicero C, Frulloni L, Carbognin G, Mantovani W, Mucelli RP. Autoimmune pancreatitis: CT patterns and their changes after steroid treatment. Radiology. 2008 May;247(2):435-43. doi: 10.1148/radiol.2472070598.

15. Kozoriz MG, Chandler TM, Patel R, Zwirewich CV, Harris AC. Pancreatic and Extrapancreatic Features in Autoimmune Pancreatitis. Can Assoc Radiol J. 2015 Aug;66(3):252-8. doi: 10.1016/j.carj.2014.10.001.

16. Sahani DV, Kalva SP, Farrell J, Maher MM, Saini S, Mueller PR, Lauwers GY, Fernandez CD, Warshaw AL, Simeone JF. Autoimmune pancreatitis: imaging features. Radiology. 2004 Nov;233(2):345-52. doi: 10.1148/radiol.2332031436.

INTERNATIONAL STANDARD SERIAL NUMBER

Crossref doi

CC BY

Submit your manuscript to the

American Journal of Case Reports and Clinical Images
CODEN: USA

and benefit from:

- Convenient online submission
- Rigorous peer review
- Immediate publication on acceptance
- Open access: articles freely available online
- High visibility within the field
- Retaining the copyright to your article

Submit your manuscript at

https://amejcaserepclinimag.com/

&

wjcasereports@gmail.com; submission@amejcaserepclinimag.com