CASE REPORT

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Postoperative Hepatic Inflammatory Pseudotumor in Autoimmune Pancreatitis: Hepatic Involvement of IgG4-related Disease

Hepatik Enflamatuvar Psödotümör Gelişen Opere Otoimmün Pankreatit Olgusu: IgG4 İlişkili Hastalığın Karaciğer Tutulumu

Ali Uludağ[1], Aylin Altan Kuş[2]

[1]İstanbul Aydın University WM Medical Park Florya Hospital, Department of Radiology, İstanbul, Turkey
[2]Acıbadem Atakent Hospital, Clinic of Radiology, İstanbul, Turkey

Abstract

Autoimmune pancreatitis (AIP) is a rare subtype of chronic pancreatitis with IgG4-positive lymphoplasmacytic infiltration and storiform fibrosis, often mimicking pancreatic cancer. A 69-year-old male presented with abdominal pain, jaundice, elevation of cholestatic enzymes, and elevated CA 19-9. Magnetic resonance imaging (MRI) and magnetic resonance cholangiopancreatography demonstrated diffuse pancreatic enlargement, T1 hypointensity, and delayed enhancement. Malignancy could not be excluded; pancreaticoduodenectomy confirmed Type I IgG4-related AIP. Two months postoperatively, MRI detected a cystic-cavitary hepatic lesion. Infectious causes were excluded, and corticosteroid therapy for presumed hepatic inflammatory pseudotumor (IPT) achieved complete resolution. AIP may closely resemble pancreatic adenocarcinoma clinically and radiologically, particularly in focal disease. Accurate diagnosis requires integrated evaluation of MRI morphology, ductal configuration, and enhancement behavior. Although uncommon, hepatic IPT is a key extrapancreatic manifestation of IgG4-related disease and should be recognized as a benign, steroid-responsive entity to avoid unnecessary surgical intervention.

Keywords: Autoimmune pancreatitis, hepatic inflammatory pseudotumor, IgG4-related disease, pancreatic mass

Öz

Otoimmün pankreatit (OİP), kronik pankreatitin nadir bir alt tipidir. Bu olgu sunumunda, Tip I otoimmün pankreatitli bir hastada Whipple operasyonu sonrası gelişen hepatik enflamatuvar psödotümörün radyolojik ve klinik özellikleri tartışılmıştır. Altmış dokuz yaşında erkek hasta, karın ağrısı ve sarılık şikayetleri ile başvurdu. Laboratuvar testlerinde karaciğer fonksiyon testlerinde bozulma ve CA 19-9 yüksekliği saptandı. Manyetik rezonans görüntüleme (MRG) ve manyetik rezonans kolanjiyopankreatografide pankreasta difüz boyut artışı ve konturlarda düzleşme, geç fazda yoğun kontrast tutulumu izlendi. Klinik değerlendirme sonucunda Whipple prosedürü uygulandı. Patoloji Tip I IgG4 ilişkili OİP ile uyumlu bulundu. Postoperatif ikinci ayda karaciğerde kistik-kaviter lezyon gelişti. Enfeksiyon dışlandı ve hepatik enflamatuvar psödotümör (EPT) ön tanısıyla steroid tedavisi başlandı. Takip MRG incelemelerinde lezyon tamamen geriledi. OİP, klinik ve radyolojik olarak pankreatik adenokarsinomu taklit edebilir. Özellikle fokal tutulum durumlarında ayırıcı tanıda MRG bulguları, pankreatik kanal morfolojisi ve kontrast tutulum paternlerinin dikkatle değerlendirilmesi gereklidir. Hepatik EPT, nadir olmakla birlikte, IgG4 ilişkili hastalık spektrumunda düşünülmelidir.

Anahtar kelimeler: Hepatik enflamatuvar psödotümör, IgG4 ilişkili hastalık, otoimmün pankreatit, pankreatik kitle

Address for Correspondence: Ali Uludağ, İstanbul Aydın University WM Medical Park Florya Hospital, Department of Radiology, İstanbul, Turkey
E-mail: uludaga@gmail.com **ORCID:** orcid.org/0000-0002-3717-5467
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Introduction

Autoimmune pancreatitis (AIP) is a rare subtype of chronic pancreatitis classified into two distinct types (1). Type I is more frequently observed in men and is associated with IgG4 and with antinuclear, anti-smooth muscle, anti-lactoferrin, and anti-carbonic anhydrase antibodies (2). Extrapancreatic inflammatory findings and the coexistence of other autoimmune diseases are commonly encountered in Type I AIP. This entity is defined as IgG4-related systemic disease. Type II, however, is not associated with specific autoantibodies or extrapancreatic manifestations. Serum IgG4 levels above 135 mg/dL are useful in differentiating AIP pancreatitis from other pancreatic disorders; however, such elevation may also occur in pancreatic carcinoma and chronic pancreatitis (2).

AIP may coexist with other autoimmune diseases, such as ulcerative colitis or autoimmune hepatitis. Radiological findings of AIP may include a focal mass, a normal-sized pancreas, or diffuse, uniform enlargement with loss of lobulated contours (3). Mild localized lymphadenopathy is frequently observed, most commonly involving the pancreatic head. In pancreatic carcinoma, abrupt ductal narrowing with distal smooth dilatation may help differentiate the two entities (4). Diagnostic criteria for AIP have been developed, incorporating radiological, laboratory, and histopathological assessments (5-7). Within the spectrum of IgG4-related AIP, hepatic inflammatory pseudotumor (IPT) subtypes characterized by fibrohistocytic and lymphoplasmacytic inflammation have also been described (6). This case report aims to highlight the radiological diagnosis of AIP and present a case of hepatic IPT that developed after Whipple surgery.

Case Report

A 69-year-old male presented to the general surgery outpatient clinic with abdominal pain. On physical examination, a midline surgical scar was present, but no palpable mass was detected. The sclerae were icteric. Laboratory analysis revealed elevated aspartate aminotransferase, alanine aminotransferase, gamma-glutamyl transferase, and alkaline phosphatase levels; mildly increased CA 19-9; direct hyperbilirubinemia; and elevated C-reactive protein.

Upper abdominal magnetic resonance imaging (MRI) and magnetic resonance cholangiopancreatography (MRCP) demonstrated a hydropic gallbladder and diffuse dilatation of the intrahepatic bile ducts. The common hepatic duct and proximal common bile duct measured up to 17 mm in diameter, with an abrupt narrowing of the intrahepatic portion of the common bile duct (Figure 1d). On contrast-enhanced MRI, the pancreas appeared diffusely enlarged with smooth contours, hypointense on T1-weighted images, and encapsulated. Diffusion restriction and intense delayed-phase enhancement were also observed (Figures 1a, 1b, 1c). These findings were highly suggestive of AIP; however, malignancy could not be excluded, and the patient underwent a Whipple procedure. Pathological examination confirmed Type I IgG4-related AIP.

Follow-up and Treatment Response

On follow-up MRI at two months, a cystic-cavitary lesion with peripheral enhancement was observed in hepatic segment 4b, raising suspicion for hepatic IPT. Infectious markers were negative, and corticosteroid therapy was initiated (Figure 2). At three months, the lesion appeared more solid and slightly enlarged, prompting an adjustment to the steroid regimen (Figure 3).

By eight months, MRI demonstrated complete resolution of the hepatic lesion, confirming a favorable response to corticosteroid therapy and supporting the diagnosis of hepatic IPT.

Discussion

Type I AIP is widely recognized as the pancreatic manifestation of IgG4-related disease. In contrast, Type II is characterized by unique histopathological and clinical features and typically lacks elevated serum IgG4 levels or associated autoantibodies (2). The IgG4-associated form of AIP is defined by dense periductal infiltration of IgG4-positive plasma cells accompanied by fibrosis. Imaging or clinical evaluation may reveal either diffuse or localized pancreatic enlargement with irregular ductal narrowing. Progressive acinar atrophy and fibrosis lead to the loss of normal lobular architecture (2,8).

IgG4-related AIP occurs in approximately 2-8% of patients with chronic pancreatitis (3). It predominantly affects middle-aged and elderly men, with a male-to-female ratio of 3-7:1 (3). Although patients with AIP do not present with specific symptoms, some may experience abdominal pain, obstructive jaundice, weight loss, new-onset diabetes, pancreatic enlargement, or, rarely, extrapancreatic lesions (3). MRI plays a critical role in diagnosis. In cases of diffuse involvement, homogeneous enlargement of the entire

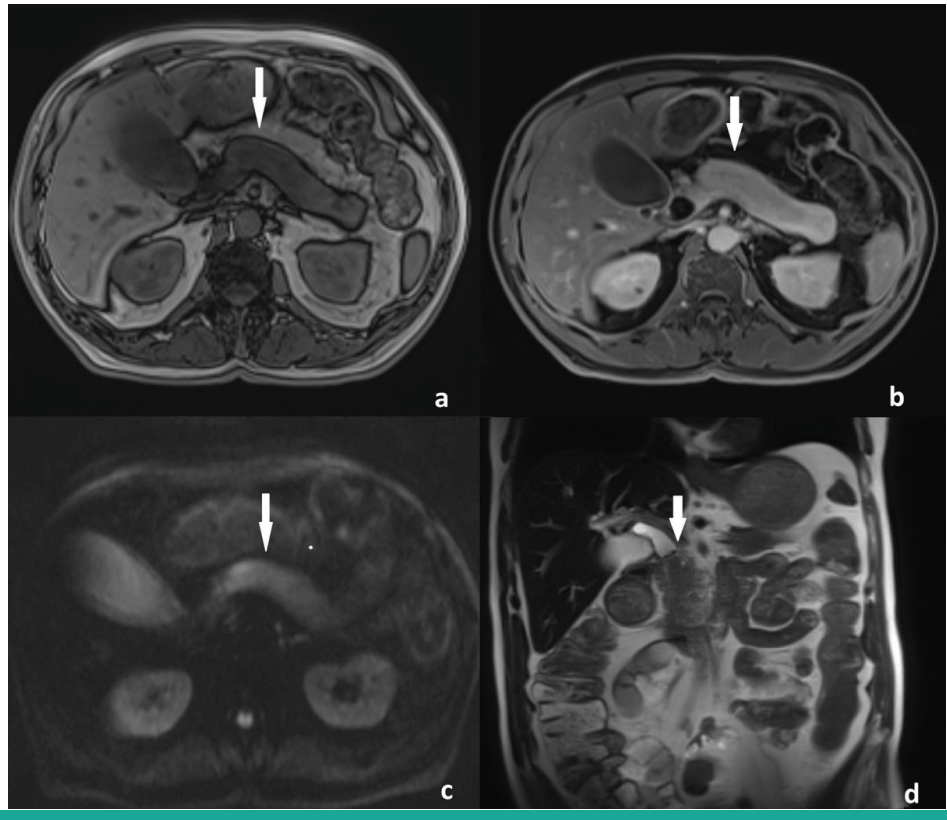

Figure 1. Axial T1 out of phase image (1a) demonstrates diffuse enlargement of the pancreas with smooth contours and decreased T1 signal intensity (arrow). Post-contrast axial T1-weighted image (1b) shows homogeneous enhancement of the pancreas (arrow). The axial diffusion-weighted image (b=1500 s/mm^2) (1c) reveals a uniform diffusion hyperintensity consistent with inflammatory activity (arrow). Coronal T2-weighted image (1d) demonstrates upstream dilatation of the common bile duct proximal to its intrapancreatic segment (arrow)

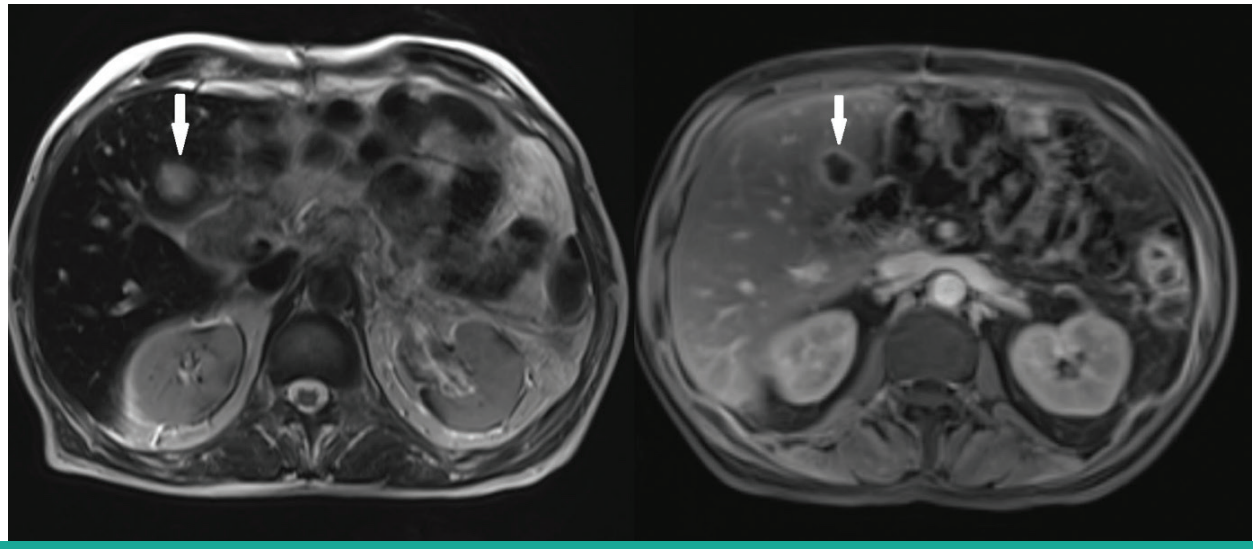

Figure 2. Contrast-enhanced MRI obtained two months after surgery. Axial T2-weighted image (left) shows a thick-walled, cystic, cavitary lesion in hepatic segment 4B (arrow). Axial post-contrast T1-weighted image (right) demonstrates peripheral wall enhancement with a non-enhancing cystic center (arrow). No diffusion restriction was observed (not shown), a finding consistent with hepatic inflammatory pseudotumor

MRI: Magnetic resonance imaging

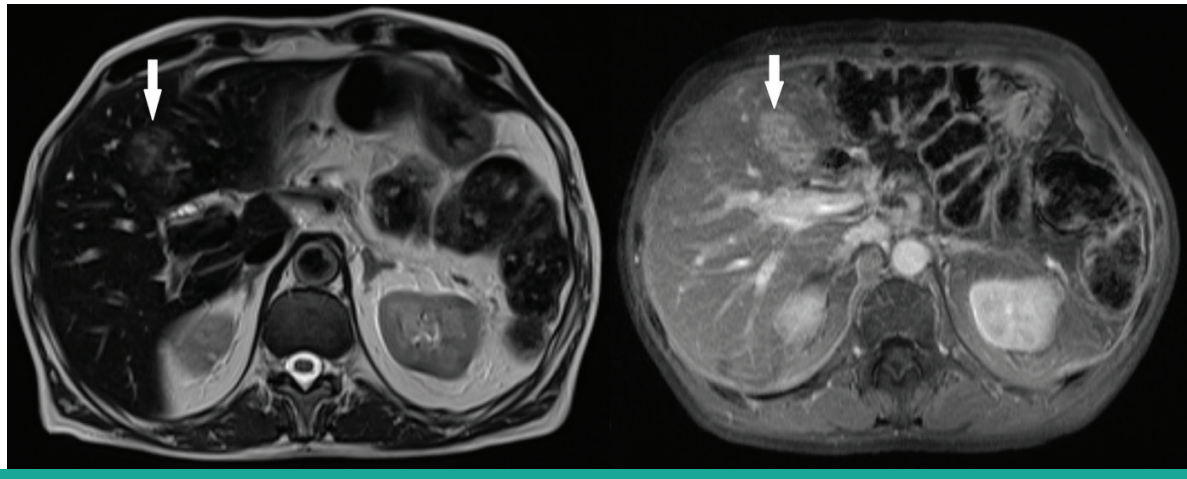

Figure 3. On the follow-up MRI obtained three months after surgery. Axial T2-weighted image (left) demonstrates that the cystic component of the lesion has transformed into a solid component (arrow). On axial post-contrast T1-weighted images (right), the lesion shows diffuse, homogeneous enhancement, confirming its solid nature (arrow)

MRI: Magnetic resonance imaging

gland is observed, with decreased signal intensity on T1-weighted images and increased signal intensity on T2-weighted sequences. On contrast-enhanced MRI, the delayed phase shows striking, homogeneous, intense enhancement. Together with findings such as "diffuse enlargement" and a "capsule-like rim", this appearance strongly supports the diagnosis of AIP (6). With focal involvement, a mass-like lesion is observed in a limited portion of the pancreas, most commonly in the pancreatic head. This appearance may mimic pancreatic adenocarcinoma, particularly if associated with target-like peripheral enhancement. AIP lesions typically demonstrate more homogeneous and more delayed enhancement than adenocarcinoma. The Desmoplastic reaction is less pronounced. Furthermore, MRCP may reveal multiple sequential ductal narrowings and dilatations or the so-called "penetrating duct sign", in which the duct continues through the lesion, gradually tapering. These findings suggest an inflammatory process and help differentiate it from malignancy (6-8). In AIP, the main pancreatic duct typically appears long, irregular, and narrowed, whereas in adenocarcinoma, the narrowing is usually abrupt, short, and accompanied by marked distal dilatation. Therefore, the presence of a focal mass is not a direct indicator of malignancy; rather, signal characteristics, ductal morphology, and enhancement patterns must be assessed together (6-8).

The diagnosis of AIP remains challenging, as a subset of cases closely mimics pancreatic adenocarcinoma. Approximately 3-9% of patients undergoing resection for suspected carcinoma are ultimately diagnosed with AIP (4). Various diagnostic criteria have been proposed internationally. The Japanese Pancreas Society first introduced diagnostic criteria including imaging findings, serology (elevated IgG4), and characteristic histopathological features (5). Subsequently, additional diagnostic parameters were proposed (7). In 2011, the International Association of Pancreatology published the International Consensus Diagnostic Criteria for Type I AIP, which defined five key features: Pancreatic imaging (parenchymal and ductal), serology (IgG4), histopathology and immunostaining, involvement of other organs, and response to steroid therapy (5,7,8).

Differentiating hepatic IPT from metastasis or abscess can be challenging due to overlapping imaging characteristics. Nevertheless, certain radiological and clinical clues may assist in making the distinction. IPTs are typically solitary lesions with delayed homogeneous enhancement and mild diffusion restriction, corresponding to their fibroinflammatory histology. In contrast, hepatic abscesses commonly demonstrate central diffusion restriction with peripheral rim enhancement and are usually accompanied by fever and marked leukocytosis. Metastatic lesions are often multiple, show heterogeneous enhancement with surrounding edema, and frequently exhibit early arterial enhancement followed by washout in delayed phases (9).

Although hepatic involvement in Type I AIP is rare, hepatic IPTs may present as an important extrapancreatic manifestation. Limited case series have described histological features of dense IgG4-positive plasma cell

infiltration, fibroinflammation, and obliterative phlebitis. These lesions frequently mimic malignancy; however, their rapid response to corticosteroid therapy confirms the diagnosis. For instance, Kamisawa et al. (10) reported two cases in which both AIP and hepatic pseudotumor responded to steroids. These findings suggest that hepatic inflammatory lesions represent a benign, treatment-responsive entity within the systemic spectrum of IgG4-related AIP.

Conclusion

This case represents a pathologically confirmed case of Type I AIP, consistent with the literature in terms of age, sex, laboratory findings, and radiological findings. The hepatic lesion that subsequently developed was diagnosed as an IPT, a rare manifestation. Cross-sectional imaging not only plays a pivotal role in diagnosing pancreatic involvement but also enables early detection of multi-organ manifestations, such as hepatic IPT.

Ethics

Informed Consent: Informed consent was obtained from the patient for sharing the clinical and radiological findings included in this case report. The patient understood that all data presented would be anonymized and that no personally identifiable information would be disclosed. Written consent was documented and archived in accordance with institutional and ethical guidelines.

Footnotes

Authorship Contributions

Surgical and Medical Practices: A.U., A.A.K., Concept: A.U., Design: A.U., Data Collection or Processing: A.U., A.A.K., Analysis or Interpretation: A.U., A.A.K., Literature Search: A.U., A.A.K., Writing: A.U.

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References

1. Gallo C, Dispinzieri G, Zucchini N, Invernizzi P, Massironi S. Autoimmune pancreatitis: cornerstones and future perspectives. World J Gastroenterol. 2024;30(8):817-832.
2. Wu S. A comprehensive review of IgG4-related pancreatitis: pathogenesis, diagnosis, and therapeutic advances. Front Immunol. 2025;16:1590902.
3. Zhang XD, Li M, Chen H, Wang L, Xie X, Ma Z, et al. Autoimmune pancreatitis: a bibliometric analysis from 2002 to 2022. Front Immunol. 2023;13:1135096.
4. Sharbin GK, Khan B, Al-Hilli S, Al-Anazi S. Autoimmune pancreatitis masquerading as a pancreatic malignancy: a case report and literature review. Medicine (Baltimore). 2025;104(28):e42481.
5. Shimosegawa T, Chari ST, Frulloni L, Kamisawa T, Kawa S, Mino-Kenudson M, et al. International consensus diagnostic criteria for autoimmune pancreatitis: Guidelines of the International Association of Pancreatology. Pancreas. 2011;40(3):352-358.
6. Yoon SB, Jeon TY, Moon SH, Shin DW, Lee SM, Choi MH, et al. Systematic review and meta-analysis of MRI features for differentiating autoimmune pancreatitis from pancreatic adenocarcinoma. Eur Radiol. 2022;32(10):6691-6701.
7. Ha J, Choi SH, Kim KW, Kim JH, Kim HJ, et al. MRI features for differentiation of autoimmune pancreatitis from pancreatic ductal adenocarcinoma: a systematic review and meta-analysis. Dig Liver Dis. 2022;54(7):849-856.
8. Choi SY, Kim SH, Kang TW, Song KD, Park HJ, Choi YH. Differentiating mass-forming autoimmune pancreatitis from pancreatic ductal adenocarcinoma on the basis of contrast-enhanced MRI and DWI findings. AJR Am J Roentgenol. 2016;206(2):291-300.
9. Calistri L, Marchionni N, Zoppi M, et al. Magnetic resonance imaging of inflammatory pseudotumor of the liver: a 2021 systematic literature update and series presentation. Abdom Radiol (NY). 2022;47(8):2795-2810.
10. Kamisawa T, Egawa N, Nakajima H, Tsuruta K, Okamoto A, Kamata N, et al. A case of autoimmune pancreatitis associated with hepatic inflammatory pseudotumor. J Gastroenterol. 2006;41(7):653-656.