Annals of Case Reports and Clinical Studies
Case Report (ISSN: 2834-5673)

Case Report: IgG4-Related Pancreatic and Bile Duct Masses Mimicking Malignancy: A Diagnostic Trap Uncovered

Anthony Albayeh[#*], Alexandre Khoury[#], Laura El Halabi[#] and Bassam Mattar

Kansas University Medical Center, Internal Medicine Department, Kansas St, Wichita, KS 67214, USA

[#]authors contributed equally to this work and shared co-first authorship.

Citation: *Anthony Albayeh, Alexandre Khoury, Laura El Halabi, Bassam Mattar. Case Report: IgG4-Related Pancreatic and Bile Duct Masses Mimicking Malignancy: A Diagnostic Trap Uncovered. Ann Case Rep Clin Stud. 2026;5(2):1-15.*

Received Date: 23 January, 2026; **Accepted Date:** 28 January, 2026; **Published Date:** 02 February, 2026

[*]**Corresponding author:** Anthony Albayeh, Kansas University Medical Center, Internal Medicine Department, Kansas St, Wichita, KS 67214, USA

ABSTRACT

Background: IgG4-Related Disease (IgG4-RD) is a systemic fibroinflammatory condition that can involve the pancreas and biliary tree, often mimicking malignancy.

Case Presentation: A 78-year-old male with coronary artery disease, type 2 diabetes mellitus, and prior colon polyps presented with jaundice, significant weight loss, and abnormal liver function tests. Imaging revealed a pancreatic tail mass and proximal common bile duct stricture, initially raising concern for malignancy. Endoscopic ultrasound-guided biopsies demonstrated storiform fibrosis, obliterative phlebitis, and increased IgG4-positive plasma cells, confirming IgG4-RD. The patient was treated with corticosteroids with initial improvement, but relapse occurred after stent removal 70 days later. He was subsequently started on rituximab with plans for repeat dosing at six months.

Keywords: IgG4-Related Disease; Pancreas; Coronary artery

INTRODUCTION

IgG4-Related Disease (IgG4-RD) is a systemic fibroinflammatory condition that can affect a single organ or involve multiple organ systems throughout the body. It is a rare immune-mediated disorder characterized by fibrosis and tumor-like mass formation. The most commonly affected organs include the orbital adnexal structures, salivary glands, pancreas, kidneys, and retroperitoneum [1]. The pathology of IgG4-related disease, also referred to as IgG4 syndrome, is consistent across all involved organs. The histological criteria that strongly suggest IgG4-RD require at least two of the following characteristic features: 1- a dense lymphoplasmacytic infiltrate, 2- fibrosis with a storiform pattern, and 3- obliterative phlebitis [2]. The digestive system is the most frequently affected site in IgG4-related disease, and the clinical presentation typically corresponds to the specific organ involved [3]. In this paper, we present a rare case of IgG4-related disease involving both the pancreas and the hepatic system, mimicking pancreatic cancer, cholangiocarcinoma or other biliary tract

cancers. We also review the characteristics of IgG4-related disease within the digestive tract, with particular emphasis on the pancreas and hepatic system. Furthermore, we discuss the common presenting symptoms, imaging modalities, pathological and histological findings, and current treatment approaches. Finally, we highlight the importance of early and accurate diagnosis and summarize recent advances in the understanding and management of this complex disease.

CASE PRESENTATION

A 78-year-old male with a past medical history significant for colon polyps, coronary artery disease status post stent placement (2017), diverticulosis, hypothyroidism, inguinal hernia, nephrolithiasis, rotator cuff syndrome, spinal stenosis, and type 2 diabetes mellitus presented for his annual wellness examination on April 15, 2025. His past surgical history included cardiac catheterization with stent placement and cataract extraction. Current medications were aspirin 81 mg daily, atorvastatin 40 mg daily, levothyroxine 112 mcg daily, metformin XR 1000 mg daily, mealtime insulin (Novolog), and a multivitamin. He reported no known drug allergies, denied alcohol consumption, and endorsed cigar smoking.

Initial laboratory testing revealed elevated liver enzymes (ALP 199 U/L, ALT 171 U/L, AST 115 U/L). Physical examination demonstrated mild hepatosplenomegaly. Cross-sectional CT imaging of the abdomen and pelvis showed intrahepatic biliary ductal dilatation with an abrupt transition at the porta hepatis, soft tissue thickening of the extrahepatic bile duct measuring 1.6 cm × 1.3 cm, and a hypo enhancing lesion in the pancreatic tail with minimal peripancreatic stranding, as well as a 1.6 cm simple pancreatic cyst (Figure 1). These findings raised suspicion for cholangiocarcinoma or pancreatic malignancy. Repeat laboratory studies showed persistently elevated liver enzymes and elevations in amylase and lipase.

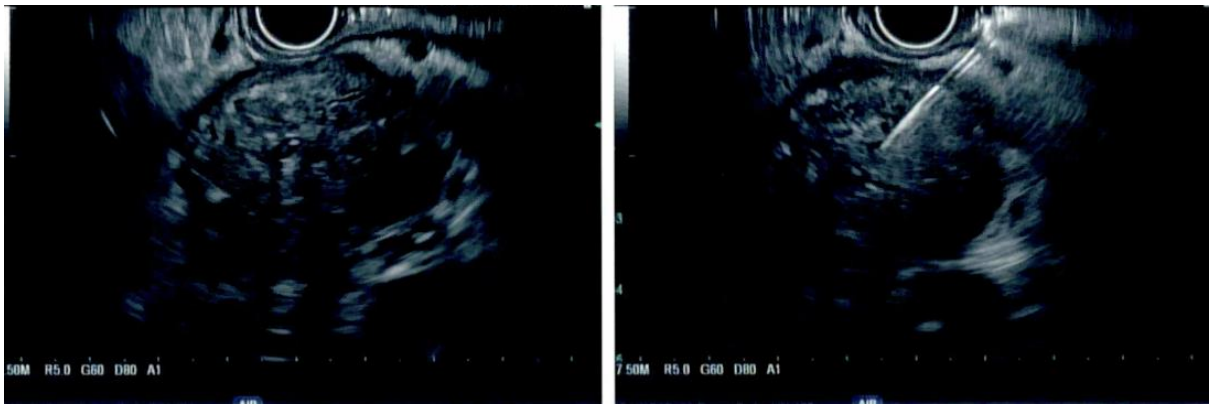

Figure 1: Pancreas body mass left image, Pancreas Body mass FNBx right image.

Endoscopic evaluation in June 2025 included Endoscopic Ultrasound (EUS), which identified a 16 mm × 14 mm hypoechoic mass in the proximal common bile duct with upstream intrahepatic ductal dilatation, as well as a 33 mm × 22 mm mass in the pancreatic body partially encasing the splenic vein. Endoscopic Retrograde Cholangiopancreatography (ERCP) demonstrated a 2 cm proximal CBD stricture consistent with a Bismuth–Corlette type I lesion, and a 7 Fr × 15 cm plastic biliary stent was successfully placed (Figure 2).

Histopathologic examination of biopsies from both the pancreatic and hilar regions revealed storiform fibrosis, obliterative phlebitis, and increased IgG4-positive plasma cells, while cytology was negative for malignancy.

Follow-up laboratory evaluation showed a persistently mild elevation in ALP (176 U/L), elevated serum IgG4 (131.8 mg/dL), positive ANA at 1:80, and an HbA1c of 11.8%. Other liver chemistries normalized with treatment. Repeat imaging demonstrated regression of the pancreatic mass from 6.2 cm to 4.2 cm, a 1.9 cm hepatic cyst in the left lobe, and mild heterogeneous hepatic steatosis.

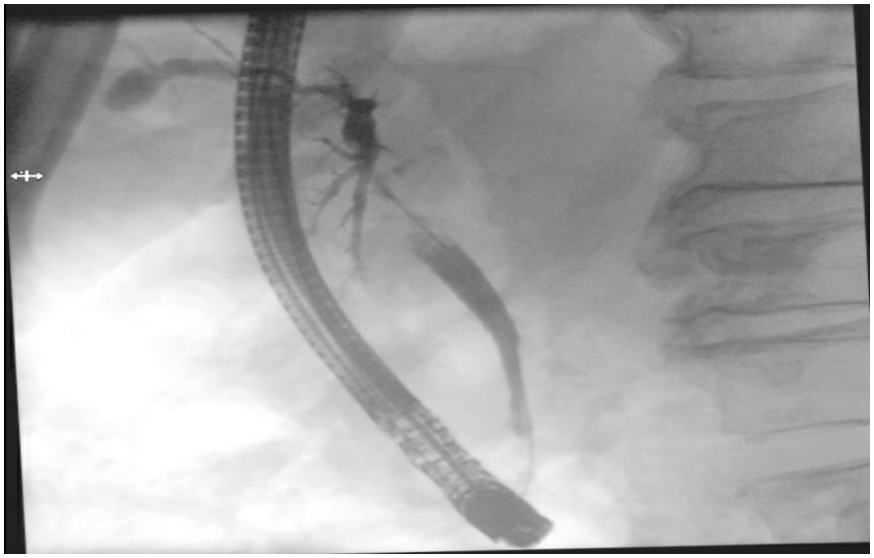

Figure 2: Cholangiogram with initial ERCP showing Biliary Stricture).

Based on clinical, radiologic, and histopathologic findings, the patient was diagnosed with recurrent IgG4-related sclerosing cholangiopathy with type 1 autoimmune pancreatitis. Initial therapy with prednisone 40 mg daily followed by a weekly taper resulted in clinical and biochemical improvement. Biliary stent was removed on August 14, 2025 (Figure 3) and ERCP showed complete resolution of the stricture. Afterwards, the patient developed recurrent biliary obstruction and was initiated on rituximab 1000 mg IV on days 1 and 15, with repeat dosing planned at six months. Alternative immunomodulators (azathioprine, mycophenolate, inebilizumab) were considered but deferred due to his age and favorable prior response to corticosteroids.

On subsequent follow-up, the patient remained clinically stable with near-normal liver function tests and good tolerance of rituximab therapy. He continues to undergo surveillance for biliary obstruction and management of diabetes, with long-term immunotherapy planned to maintain disease remission.

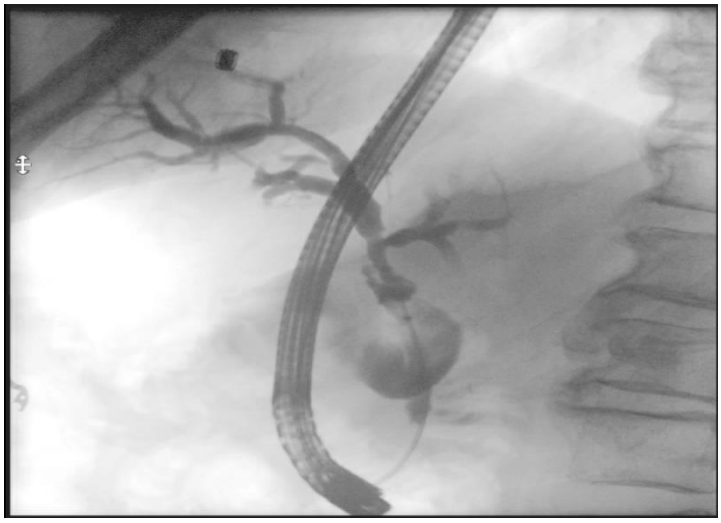

Figure 3: Cholangiogram at repeat ERCP post stent removal showing resolution of the stricture.

DISCUSSION

IgG4-Related Diseases (IgG4-RD) are a group of chronic immune-mediated disorders with evidence of genetic predisposition. A study conducted in a Japanese population identified several genetic loci associated with IgG4-RD, including the HLA-DRB104:05* allele [4]. Environmental factors may also contribute to disease susceptibility. An interesting study published in 2021 reported that individuals engaged in blue-collar occupations, particularly those exposed to biological dust, mineral dust, gas fumes, and other environmental toxins, have an increased risk of developing IgG4-RD involving the biliary tract and pancreas. This observation may, in part, explain the higher prevalence of the disease among middle-aged men [5].

The pathophysiology of IgG4-RD is complex and involves the interplay of multiple immune cell types. One of the earliest histopathological findings in this disorder is involvement of tertiary lymphoid structures, ectopic lymphoid tissues that arise within non-lymphoid organs [6]. Among the key immune players are T follicular helper (Tfh) cells, which promote the differentiation of B cells into IgG4-producing plasma cells [7]. A recent study published in 2023 identified a novel subtype of T cells expressing TOP2A at sites of IgG4-related inflammation. These TOP2A$^+$ T cells appear to be unique to IgG4-RD and may play a specific role in its immunopathogenesis [8].

Because this disease lacks specific or pathognomonic symptoms, its clinical presentation is frequently mistaken for more serious conditions such as malignancies. Diagnosis is often challenging due to the absence of definitive biomarkers, laboratory findings, or imaging features unique to IgG4-RD. To address the issues of misdiagnosis and underdiagnosis, the European League Against Rheumatism (EULAR) and the American College of Rheumatology (ACR) have proposed classification criteria to guide the diagnosis of IgG4-related disease involving the digestive system [9,10]. The first diagnostic criterion is the presence of elevated serum and tissue IgG4 levels. A serum IgG4 concentration above 1.35 g/L is generally accepted as the diagnostic cutoff. However, since elevated IgG4 levels are not specific to IgG4-RD and may also be seen in other hepatobiliary or pancreatic disorders, higher thresholds have been proposed for differentiation. One study suggested that a serum IgG4 level greater than 2.5 g/L helps distinguish IgG4-related sclerosing cholangitis from primary sclerosing cholangitis, while another study identified 5.6 g/L as a critical cutoff value to differentiate IgG4-related

sclerosing cholangitis from both primary sclerosing cholangitis and bile duct carcinoma [11,12]. The second and most definitive diagnostic criterion, often considered the gold standard, is histopathological examination of the affected organ. The characteristic histopathological features of IgG4-related disease within the digestive tract are consistent across different sites and include: a dense lymphoplasmacytic infiltrate, widespread storiform (swirling) fibrosis, and obliterative phlebitis [13]. Immunohistochemical staining should demonstrate 10–50 IgG4-positive plasma cells per high-power field to support the diagnosis.

Our paper presents a case of IgG4-related disease involving two digestive organs: the pancreas and the biliary system. Autoimmune Pancreatitis (AIP), a key manifestation of IgG4-RD, is classified into two distinct types, type 1 and type 2. Type 1 AIP, the most common pancreatic presentation of IgG4-RD, is characterized by dense lymphoplasmacytic infiltration, storiform fibrosis, and a strong male predominance in older adults. In contrast, type 2 AIP features granulocytic epithelial lesions within the pancreatic duct and is often associated with inflammatory bowel disease [14]. Since only type 1 AIP is related to IgG4-RD, our discussion will focus on this subtype. Differentiating between the two types of AIP is challenging because they share similar clinical manifestations, including obstructive jaundice, abdominal pain, and both endocrine and exocrine pancreatic insufficiency. Symptoms such as steatorrhea, postprandial bloating, increased stool frequency, and weight loss are commonly observed in both forms. The International Consensus Diagnostic Criteria for autoimmune pancreatitis, established by the International Association of Pancreatology, can be summarized by the acronym "HISORt": Histology, Imaging, Serology, Other organ involvement, and Response to treatment [15].

Histologically, microscopic examination typically reveals lobule-centric lymphoplasmacytic infiltration rich in IgG4-positive plasma cells, accompanied by storiform fibrosis. Concerning imaging, contrast-enhanced CT scans often demonstrate diffuse pancreatic enlargement with delayed enhancement, a capsule-like low-density rim, and main pancreatic duct strictures. Endoscopic ultrasound can further aid diagnosis by identifying diffuse hypoechogenic enlargement of the pancreas and thickening of the biliary tree. EUS-guided core biopsy also enables histopathological confirmation [16]. Although serum biomarkers are not diagnostic, they can support AIP diagnosis in the appropriate clinical context. Inflammatory markers are typically normal or only mildly elevated in both forms of AIP. Elevated serum IgG4 levels are found in approximately 60% to 70% of type 1 AIP cases and correlate with the extent of multi-organ involvement, whereas they are usually normal in type 2 AIP [16]. Other organ involvement is particularly useful for distinguishing type 1 from type 2 AIP. Extra-pancreatic manifestations occur in about 61–95% of type 1 AIP cases [15-17]. A favorable response to corticosteroids further supports the diagnosis. Prednisone at a dose of 0.6–1 mg/kg/day typically results in clinical and radiologic improvement in suspected IgG4-related AIP [18].

Most patients with hepatobiliary IgG4-RD have concomitant AIP; therefore, the presence of both a hepatic hilar mass and pancreatic involvement in our patient is consistent with IgG4-RD, though this presentation remains uncommon. Hepatobiliary involvement is estimated to occur in 4% to 10% of all IgG4-RD cases [18]. IgG4-RD of the liver can exhibit overlapping clinical and pathological features with Autoimmune Hepatitis (AIH). However, unlike AIH, which is characterized by hepatocellular injury and positive autoantibodies such as anti–smooth muscle and anti–liver/kidney microsomal antibodies, IgG4-RD primarily targets the bile ducts and portal regions [19]. Importantly, IgG4-RD may form tumor-like inflammatory masses that can closely mimic malignancy. These lesions must be carefully distinguished from true neoplasms and from other inflammatory

masses, as IgG4-RD lesions are characterized histologically by prominent lymphoplasmacytic infiltration with abundant IgG4-positive plasma cells, whereas non-IgG4 inflammatory lesions typically show a predominance of histiocytic infiltration and fewer IgG4-positive plasma cells [20].

Autoimmune Pancreatitis (AIP) can have overlapping characteristics in clinical presentation or imaging workup with pancreatic adenocarcinoma or even cholangiocarcinoma. Abdominal pain, elevated lipase, painless obstructive jaundice, endocrine or exocrine pancreatic failure are some of the manifestations found in both, making it difficult to differentiate clinically between the two entities [21,22].

An elevated IgG4 level alone is also insufficient to rule out cancerous etiology as cancer can also elevate the IgG4 numbers. However, levels greater than 135 mg/dL have been shown to better correlate with the diagnosis of an IgG4 related disease rather than pancreatic cancer where levels are usually lower. Considering a much more elevated cut-off of more than 280 mg/dL was even more decisive in regards of ruling out cancer [21-23].

Radiologically, typical findings of AIP can be found on Computed Tomography (CT) imaging. A sausage-like appearance is rarely observed on CT in cases of pancreatic cancer, pointing out towards AIP. Along with that, delayed-phase dynamic enhancement of pancreatic enlargement on CT and Magnetic Resonance Imaging (MRI) is characteristic of AIP. Also, on MRI, Apparent Diffusion Coefficient (ADC) values were found to be lower in AIP patients. Finally, a rim shaped like a capsule is specific to AIP as demonstrated by different studies [22,24-26].

On Endoscopic Retrograde Cholangiopancreatography (ERCP), the irregular narrowing of the main pancreatic duct is more suggestive of AIP, and on Magnetic Resonance Cholangiopancreatography (MRCP) a less upstream dilatation of the main pancreatic duct favors AIP diagnosis. The role of MRCP cannot be undermined in detecting the response to steroid therapy [27,28].

As for 18F-Fluorodeoxyglucose (FDG) Positron Emission Tomography (PET)/CT moderate to intense FDG intake of an enlarged pancreas without pancreatic duct obstruction points toward AIP and can be useful in determining any other site of the disease [29].

A histopathological diagnosis is highly desirable in some cases since it can help clear any confusion. AIP has a panoply of histopathological features including:

- Lymphoplasmacytic infiltrate of IgG4-positive plasma cells and CD4 T-lymphocytes
- A storiform pattern of fibrosis
- Obliterative phlebitis

On immunohistochemistry, an infiltrate of 50 or more IgG4-positive plasma cells with/without a higher than 40% ratio of IgG4 plasma cells of all IgG plasma cells supports the diagnosis [30].

Finally, the involvement of other organs histologically can help diagnose IgG4-related diseases. Samples from gallbladder and salivary gland if obtained can sometimes be characteristic of the disease preventing unnecessary complementary workups [31].

The goal of IgG4 related disease treatment is to reduce its activity and prevent irreversible damage. The response to the multiple regimens depends on the extent of disease (organs involvement) and its duration based mainly on the degree of fibrosis [32].

Glucocorticoids are the first-line therapy for IgG4-related diseases. 0.6-0.8 mg/kg for 2weeks to 4 weeks of Prednisone is the mainstay starting dose. Remission was obtained in 95 to 100% of patients. The optimal regimen and duration of treatment are still debated [33-36].

However, the risk of relapsing cannot be eliminated along with the risk related to glucocorticoid use, especially in the frail population. On top of that comes the uncertainty of the efficacy of glucocorticoid medication in remission maintenance. Here comes the inevitable role of immunosuppressive medication (Disease-Modifying Antirheumatic Drugs (DMARDs) including Azathioprine, Mycophenolate mofetil, Methotrexate, Leflunomide, Tacrolimus, Ciclosporin A, Cyclophosphamide…). Used on top of low-dose glucocorticoid medication, they prevent relapses, improve remission rates and prevent glucocorticoids' undesired effects [36-38].

Targeting B cells in IgG4 related disease is also an important pillar of the treatment in patients with multiple relapses, intolerance, and resistance to glucocorticoid therapy. Rituximab used at a dose of 375 mg/m^2 every week for 4 weeks or 1000 mg on day 1 and day 15 leads to high remission even when used as monotherapy and is effective in treating disease flares [36,37].

Other regimens are being currently developed, notably Belimumab, Bruton kinase inhibitors, CD19 targeting regimens, and a monoclonal antibody targeting SLAM (Elotuzumab) [37].

- Monitoring for recurrence and multi-organ involvement.

Monitoring disease relapses is crucial to check for the treatment's successful induction and detect any treatment-needing recurrence. This can be done using lab tests (serum IgG4 levels, IgE concentrations, complement levels, eosinophils count). Organ-specific markers like urinary protein, bilirubin, and alkaline phosphatase levels can be obtained to detect multi-organ involvement or relapse [32].

Radiological observation can be used along with lab tests, especially when the disease is more apparent on imaging. An assessment done every 3 months to 6 months in the absence of disease manifestations is adequate [32].

A study by Lanzillotta et al. predicted prognosis of IgG4 related disease patients based on differences in clinical phenotypes and can give a clue about the prognosis of the disease in specific populations of patients thus helping in predicting disease outcomes [19]. Moreover, another study by Zongfei et al. showed the role of elevated serum IL-2 receptor and tumor necrosis factor alpha (TNF alpha) levels in detecting risk for refractory and relapsed IgG4-related disease [39].

We present in the table below similar case reports found in the literature detailing patient demographics, presentation, diagnostic method, treatment received, and outcome (Table 1).

Table 1: Similar case reports found in the literature.

Author	Year	Age (years)	Sex	Presentation	Diagnostic method	Treatment	Outcome
Suliemman et al. [40]	2018	57	Male	Presented with a pancreatic head mass and obstructive jaundice	Physical examination, cervical ultrasound, fine needle aspiration cytology (FNAC), IgG level, AST and ALT levels, abdominal ultrasound, MRI/MRCP, liver function tests (LFTs), complete blood count (CBC), serology for hepatitis B virus (HBV) and hepatitis C virus (HCV), AFP, CA19-9, CA15-3, CEA, total IgG, complement C3 and C4, endoscopic ultrasound, FDG PET/CT, lymph node biopsy, histopathological examination, immunohistochemistry	Intravenous pulsed steroids (Methylprednisolone 500 mg daily) for three days, followed by oral prednisolone 40 mg daily	Eight months after the onset of treatment, the patient was asymptomatic, with complete resolution of the lymphadenopathy, jaundice, skin lesions, allergies and conjunctival manifestations, and the obstructive urinary symptoms
Zhang et al. [41]	2022	52	Male	Presented with yellow urine and abdominal distension for more than one month and yellowish skin and sclera for 20 days	Physical examination, LFTs, IgG4 levels, CRP, procalcitonin, tumor markers, CBC, comprehensive metabolic panel (CMP), thyroid test, coagulation test, serum protein electrophoresis, immunoglobulin levels, doppler ultrasound of liver gallbladder and spleen, abdominal CR, MRCP, MRI, color	Methylprednisolone 40 mg intravenously once a day followed by oral metoprolol 32 mg with gradual tapering; silibinin capsules, acid suppression and calcitriol supplementation	After 3 months of treatment, abdominal distension, yellow urine, and yellowish skin and sclera were relieved

					Doppler ultrasound of the submandibular gland, gastroscopy, needle biopsy of the left submandibular gland and lymph node, gastric antrum mucosal tissue biopsy, pathology, immunohistochemistry, endoscopic histology for H. pylori		
Zhang et al. [42]	2022	65	Male	Presented with fatigue and edema of both lower extremities for three months	LFTs, IgG4 levels, lymph node ultrasound, enhanced CT abdomen, gastrointestinal endoscopy, pathology, immunohistochemistry	Ursodeoxycholic acid and bicyclol	Symptoms improved significantly and liver function indicators gradually improved
Rebollo et al. [42]	2025	58	Woman	Consulted for pruritus and liver panel alterations	AST, ALT, GGT, alkaline phosphatase, CA19-9, MRI, endoscopic ultrasound-guided FNA, histopathological study, IgG levels, ANA, endoscopic ultrasound, liver biopsy	Corticosteroids associated with azathioprine	Good response
Hsu et al. [43]	2018	52	Male	Visited because of one month history of intermittent epigastralgia	LFTs, CBC with differential, cholesterol profile, electrolytes, renal function parameters, tumor markers (AFP, CA19-9, CEA), esophagogastroduodenoscopy, abdominal	Three months of corticosteroid treatment	After 48 months of follow-up, patient remained in good condition without relapse

					ultrasound, abdominal CT, CT-guided biopsy, immunohistochemistry, FDG-PET/CT, IgG and IgG4 levels, IgA, IgM		
Geary et al. [44]	2018	72	Male	Presented with obstructive jaundice	LFTs, MRI abdomen, ERCP, CT abdomen, right hepatectomy and left jejunostomy, histopathology, immunohistochemistry, IgG4 and IgG levels	Steroid therapy (stress dose)	Patient worsened and passed away during admission three months later
Harada et al. [45]	2022	82	Male	Presented with abnormal liver function and gallbladder wall thickening	CBC, serum chemical studies, CEA, CA19-9, dynamic CT, MRI, contrast-enhanced dynamic MRI, MRCP, EUS, gallbladder resection specimen, pathological examination, IgG and IgG4	Laparoscopic whole layer cholecystectomy	Remained relapse-free for one and a half years after surgery
Xiao et al. [46]	2017	58	Female	Presented with one-month history of jaundice, right upper quadrant discomfort, pruritus, pale stools and dark urine	LFTs, CA19-9, contrast-enhanced CT, laparotomy, pathological examination, immunohistochemistry, IgG4	Surgical resection	No recurrence after nine months of follow-up
Cao et al. [47]	2015	40	Male	Presented with upper abdominal	LFTs, CA19-9, CEA, abdominal ultrasound, CT scan, PET/CT, EUS-	Pancreatico-duodenectomy	No discomfort after seven months of

				pain and jaundice	guided biopsy, pancreatico-duodenectomy, pathology		follow-up
Sharbin et al. [48]	2025	51	Female	Presented with three to four days of abdominal pain	WBCs, lipase, liver and metabolic panels, CA19-9, CEA, CT abdomen and pelvis, MRI, MRCP, EUS with FNB, histopathology, serum immunoglobulins, IgG4	Forty mg oral prednisone taper for two weeks	Imaging findings consistent with resolving autoimmune pancreatitis
Nguyen-tat et al. [49]	2012	79	Male	Presented with painless jaundice and abdominal discomfort	Laboratory examinations, endoscopic cholangiography, CT imaging, left hemi hepatectomy, histopathology, GGT, bilirubin, MRI, abdominal ultrasound, IgG and IgG4 levels	Eight weeks of steroid therapy followed by azathioprine and prednisolone taper	Complete clinical remission during six months of follow-up

CONCLUSION

IgG4-related disease is an uncommon but clinically important mimic of pancreatic and biliary malignancies. This case highlights how its mass-forming presentation, particularly when involving multiple digestive organs, can closely resemble pancreatic cancer, cholangiocarcinoma, or other hepatobiliary tumors, often leading to delayed diagnosis or unnecessary surgical exploration. Early recognition of its characteristic clinical, serologic, radiologic, and histopathologic features is therefore essential. Measurement of serum IgG4 levels, careful review of imaging findings, and, most importantly, tissue diagnosis demonstrating storiform fibrosis, obliterative phlebitis, and IgG4-positive plasma cell infiltration are critical steps before considering operative management.

Our patient's marked clinical and biochemical improvement following corticosteroid therapy, and subsequent stabilization on rituximab, reinforces the excellent steroid responsiveness of IgG4-RD and underscores the importance of immunomodulatory therapy in recurrent or refractory disease. Timely diagnosis not only prevents irreversible organ injury but also avoids unnecessary surgical resections in patients with benign, immune-mediated disease. Clinicians should maintain a high index of suspicion for IgG4-related disease in any patient.

CONFLICT OF INTEREST

The authors declare that they have no conflicts of interest and received no financial support for the research, authorship, or publication of this article.

REFERENCES

1. Wallace ZS, Stone JH. An update on IgG4-related disease. Curr Opin Rheumatol. 2015;27(1):83-90.
2. Deshpande V, Zen Y, Chan JK, Yi EE, Sato Y, Yoshino T, et al. Consensus statement on the pathology of IgG4-related disease. Mod Pathol. 2012;25(9):1181-92.
3. Della-Torre E, Mancuso G, Lanzillotta M, Ramirez G, Arcidiacono P, Capurso G, et al. Urgent manifestations of immunoglobulin G4 -related disease. Scand J Rheumatol. 2021;50(1):48-51.
4. Terao C, Ota M, Iwasaki T, Shiokawa M, Kawaguchi S, Kuriyama K, et al. IgG4-related disease in the Japanese population: a genome-wide association study. Lancet Rheumatol. 2019;1(1):e14-22.
5. Hubers LM, Schuurman AR, Buijs J, Mostafavi N, Bruno MJ, Vermeulen RCH, et al. Blue-collar work is a risk factor for developing IgG4-related disease of the biliary tract and pancreas. JHEP Rep. 2021;3(6):100385.
6. Maehara T, Moriyama M, Nakashima H, Miyake K, Hayashida JN, Tanaka A, et al. Interleukin-21 contributes to germinal centre formation and immunoglobulin G4 production in IgG4-related dacryoadenitis and sialoadenitis, so-called Mikulicz's disease. Ann Rheum Dis. 2012;71(12):2011-9.
7. Ueno H. T follicular helper cells in human autoimmunity. Curr Opin Immunol. 2016;43:24-31.
8. Li Y, Wang Z, Han F, Zhang M, Yang T, Chen M, et al. Single-cell transcriptome analysis profiles cellular and molecular alterations in submandibular gland and blood in IgG4-related disease. Ann Rheum Dis. 2023;82(10):1348-58.
9. Wallace ZS, Naden RP, Chari S, Choi H, Della-Torre E, Dicaire J, et al. The 2019 American College of Rheumatology/European league against rheumatism classification criteria for IgG4-related disease. Arthritis Rheumatol. 2020;72(1):7-19.
10. Umehara H, Okazaki K, Kawa S, Takahashi H, Goto H, Matsui S, et al. The 2020 revised comprehensive diagnostic (RCD) criteria for IgG4-RD. Mod Rheumatol. 2021;31(3):529-33.
11. Boonstra K, Culver EL, De Buy Wenniger LM, Van Heerde MJ, Van Erpecum KJ, Poen AC, et al. Serum immunoglobulin G4 and immunoglobulin G1 for distinguishing immunoglobulin G4-associated cholangitis from primary sclerosing cholangitis. Hepatology. 2014;59(5):1954-63.
12. Oseini AM, Chaiteerakij R, Shire AM, Ghazale A, Kaiya J, Moser CD, et al. Utility of serum immunoglobulin G4 in distinguishing immunoglobulin G4-associated cholangitis from cholangiocarcinoma. Hepatology. 2011;54(3):940-8.
13. Khosroshahi A, Wallace ZS, Crowe JL, Akamizu T, Azumi A, Carruthers MN, et al. International consensus guidance statement on the management and treatment of igg4-related disease. Arthritis Rheumatol. 2015;67(7):1688-99.
14. Basyal B, Sharma M, Goosenberg E. Autoimmune pancreatitis. In: StatPearls. Treasure Island (FL): StatPearls Publishing. 2025.

15. 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):352-8.
16. Kamisawa T, Chari ST, Giday SA, Kim MH, Chung JB, Lee KT, et al. Clinical profile of autoimmune pancreatitis and its histological subtypes: an international multicenter survey. Pancreas. 2011;40(6):809-14.
17. Naitoh I, Nakazawa T, Ohara H, Ando T, Hayashi K, Tanaka H, et al. Clinical significance of extrapancreatic lesions in autoimmune pancreatitis. Pancreas. 2010;39(1):e1-5.
18. Löhr J, Beuers U, Vujasinovic M, Alvaro D, Frøkjær JB, Buttgereit F, et al. European Guideline on IgG4-related digestive disease – UEG and SGF evidence-based recommendations. United Eur Gastroenterol J. 2020;8(6):637-66.
19. Onofrio FQ, Hirschfield GM, Gulamhusein AF. A practical review of primary biliary cholangitis for the gastroenterologist. Gastroenterol Hepatol. 2019;15(3):145-54.
20. Itazaki Y, Einama T, Konno F, Fujinuma I, Takihata Y, Iwasaki T, et al. IgG4-related hepatic inflammatory pseudotumor mimicking cholangiolocellular carcinoma. Clin J Gastroenterol. 2021;14(6):1733-9.
21. Yakout A, Fluxa D, Nassar A, Gomez V. The overlap of autoimmune pancreatitis and pancreatic cancer: a clinical challenge. ACG Case Rep J. 2025;12(7):e01773.
22. Takuma K, Kamisawa T, Gopalakrishna R, Hara S, Tabata T, Inaba Y, et al. Strategy to differentiate autoimmune pancreatitis from pancreas cancer. World J Gastroenterol. 2012;18(10):1015-20.
23. Ghazale A, Chari ST, Smyrk TC, Levy MJ, Topazian MD, Takahashi N, et al. Value of serum IgG4 in the diagnosis of autoimmune pancreatitis and in distinguishing it from pancreatic cancer. Am J Gastroenterol. 2007;102(8):1646-53.
24. Kamisawa T, Egawa N, Nakajima H, Tsuruta K, Okamoto A, Kamata N. Clinical difficulties in the differentiation of autoimmune pancreatitis and pancreatic carcinoma. Am J Gastroenterol. 2003;98(12):2694-9.
25. Bodily KD, Takahashi N, Fletcher JG, Fidler JL, Hough DM, Kawashima A, et al. Autoimmune pancreatitis: pancreatic and extrapancreatic imaging findings. Am J Roentgenol. 2009;192(2):431-7.
26. Sahani DV, Kalva SP, Farrell J, Maher MM, Saini S, Mueller PR, et al. Autoimmune pancreatitis: imaging features. Radiology. 2004;233(2):345-52.
27. Takuma K, Kamisawa T, Tabata T, Inaba Y, Egawa N, Igarashi Y. Utility of pancreatography for diagnosing autoimmune pancreatitis. World J Gastroenterol. 2011;17(18):2332.
28. Kamisawa T, Chen PY, Tu Y, Nakajima H, Egawa N, Tsuruta K, et al. MRCP and MRI findings in 9 patients with autoimmune pancreatitis. World J Gastroenterol. 2006;12(18):2919-22.
29. Zheng Y, Elsayes KM, Waranch C, Abdelaziz A, Menias CO, Sandrasegaran K, et al. IgG4-related disease in the abdomen and pelvis: atypical findings, pitfalls, and mimics. Abdom Radiol. 2020;45(8):2485-99.
30. Parums DV. IgG4-related disease: A review of persistent challenges in the pathogenesis, diagnosis, and approaches to treatment. Med Sci Monit. 2025;31:e950212.

31. Deshpande V. IgG4-related disease of the gastrointestinal tract: a 21st century chameleon. Arch Pathol Lab Med. 2015;139(6):742-9.
32. Wallace ZS, Katz G, Hernandez-Barco YG, Baker MC. Current and future advances in practice: IgG4-related disease. Rheumatol Adv Pract. 2024;8(2):rkae020.
33. Masamune A, Nishimori I, Kikuta K, Tsuji I, Mizuno N, Iiyama T, et al. Randomised controlled trial of long-term maintenance corticosteroid therapy in patients with autoimmune pancreatitis. Gut. 2017;66(3):487-94.
34. Hart PA, Kamisawa T, Brugge WR, Chung JB, Culver EL, Czakó L, et al. Long-term outcomes of autoimmune pancreatitis: a multicentre, international analysis. Gut. 2013;62(12):1771-6.
35. Hart PA, Topazian MD, Witzig TE, Clain JE, Gleeson FC, Klebig RR, et al. Treatment of relapsing autoimmune pancreatitis with immunomodulators and rituximab: the Mayo Clinic experience. Gut. 2013;62(11):1607-15.
36. Lanzillotta M, Vujasinovic M, Löhr JM, Della Torre E. Update on autoimmune pancreatitis and IgG4-related disease. United Eur Gastroenterol J. 2025;13(1):107-15.
37. Katz G, Stone JH. Clinical perspectives on IgG4-related disease and its classification. Annu Rev Med. 2022;73(1):545-62.
38. Kamisawa T, Zen Y, Pillai S, Stone JH. IgG4-related disease. The Lancet. 2015;385(9976):1460-71.
39. Zongfei J, Lingying M, Lijuan Z, Ying S, Rongyi C, Dongmei L, et al. Prognostic factors in IgG4-related disease: a long-term monocentric Chinese cohort study. Clin Rheumatol. 2021;40(6):2293-300.
40. Sulieman I, Mahfouz A, AlKuwari E, Szabados L, Elmoghazy W, Elaffandi A, et al. IgG4-related disease mimicking pancreatic cancer: Case report and review of the literature. Int J Surg Case Rep. 2018;50:100-5.
41. Zhang X, Jin X, Guan L, Lin X, Li X, Li Y. IgG4-related disease with gastrointestinal involvement: case reports and literature review. Front Immunol. 2022;13:816830.
42. Ruiz Rebollo ML, Álvarez-Quiñones-Sanz M, Fuertes Alija JJ, Izquierdo Santervás S. IgG4-related disease mimicking pancreatic tumour. Gastroenterol Hepatol Engl Ed. 2021;44(9):660-1.
43. Hsu WL, Chang SM, Wu PY, Chang CC. Localized autoimmune pancreatitis mimicking pancreatic cancer: Case report and literature review. J Int Med Res. 2018;46(4):1657-65.
44. Geary K, Yazici C, Seibold A, Guzman G. IgG4-related cholangiopathy and its mimickers: a case report and review highlighting the importance of early diagnosis. Int J Surg Pathol. 2018;26(2):165-73.
45. Harada Y, Mihara K, Amemiya R, Nakagawa M, Hanada R, Inoue K, et al. Isolated IgG4-related cholecystitis with localized gallbladder wall thickening mimicking gallbladder cancer: a case report and literature review. BMC Gastroenterol. 2022;22(1):99.
46. Xiao J, Li G, Yang G, Jia C, Li B. Case report: A female case of isolated IgG4-related sclerosing cholangitis mimicking cholangiocarcinoma. Medicine (Baltimore). 2017;96(16):e6542.
47. Cao Z, Tian R, Zhang T, Zhao Y. Localized autoimmune pancreatitis: report of a case clinically mimicking pancreatic cancer and a literature review. Medicine (Baltimore). 2015;94(42):e1656.
48. Sharbin GK, Edeson ME, Burg E, Hosseini HK, Hosseini DK, Patel S, et al. Autoimmune pancreatitis masquerading as a pancreatic neoplasm: A case report. Medicine (Baltimore). 2025;104(28):e43359.

49. Nguyen-tat M, Gamstätter T, Marquardt J, Geißinger E, Schadmand-Fischer S, Lang H, et al. IgG4-related sclerosing cholangitis mimicking cholangiocarcinoma. Z Für Gastroenterol. 2012;50(09):1008-12.