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Case Report

IgG4-related disease revealed by acute pancreatitis: A case report and literature review ☆,☆☆

*Salma El Aouadi**, *Kaoutar Imrani*, *Amine Naggar*, *Soukaina Bahha*, *Nabil Moatassim Billah*, *Ittimade Nassar*

Central Radiology Department, Ibn Sina University Hospital, Rabat, Morocco

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ABSTRACT

IgG4-related disease is a systemic autoimmune disorder characterized by multiorgan involvement, often presenting with pancreatic, renal, biliary, and salivary gland abnormalities. Diagnosis relies on clinical, serological, imaging, and occasionally histological findings. This report describes a 65-year-old male presenting with acute pancreatitis, bilateral renal lesions, and biliary strictures. Elevated serum IgG4 levels (3.76 g/L) confirmed the diagnosis using the 2019 ACR/EULAR and 2020 Comprehensive Diagnostic Criteria despite the lack of histological confirmation. Corticosteroid therapy led to rapid clinical and biochemical improvement, underscoring the importance of integrating multiple diagnostic modalities in managing IgG4-related disease and demonstrating the effectiveness of early intervention.

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Introduction

IgG4-related disease is a systemic autoimmune condition marked by chronic inflammation and fibrosis in various organs. Since its recognition in 2003, diagnostic frameworks have evolved to incorporate clinical, serological, radiological, and histological findings, with the 2019 ACR/EULAR and 2020 Comprehensive Diagnostic Criteria offering robust guidance. While the pancreas is commonly affected, extra-pancreatic involvement, including renal, biliary, and lymphatic systems, highlights the disease's multisystem nature. The pathogenesis, featuring autoimmune and allergic components, remains partially understood. This report discusses a case of IgG4-related disease manifesting as acute pancreatitis with extensive extra-pancreatic involvement, diagnosed without histology and managed successfully with corticosteroids.

Case report

A 65-year-old male with a history of recurrent pancreatic-like abdominal pain, previously unexplored, presented with a

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* Corresponding author.
E-mail address: salma.elaouadi@gmail.com (S.E. Aouadi).

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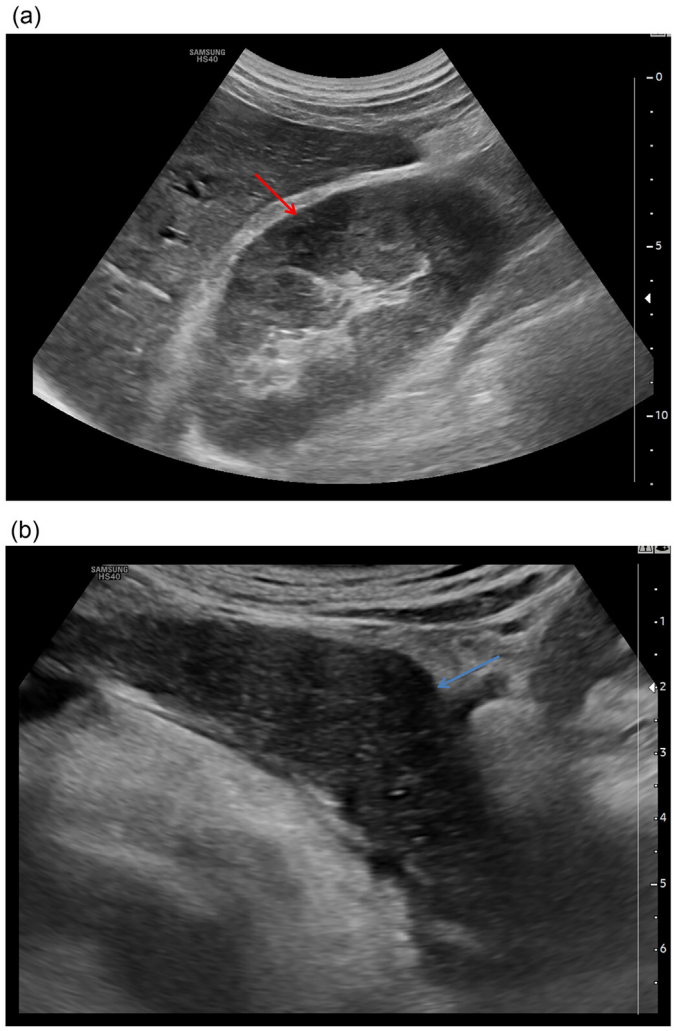

Fig. 1 – (A) Longitudinal US image shows renal cortical lesion, triangular and hypoechoic, consistent with nephritis (red arrow). (B) Transverse US image showing swollen, hypoechoic pancreas (blue arrow).

10-day history of worsening abdominal pain and vomiting. Initial blood tests revealed significantly elevated lipase levels at 357 IU/L, leukocytosis at 13,090/mm³ and an elevated C-reactive protein (CRP) of 80 mg/L, suggesting acute pancreatitis.

Imaging was performed to establish the diagnosis. Initial ultrasound (Fig. 1) revealed a swollen, hypoechoic pancreas and bilateral triangular, hypoechoic renal lesions consistent with nephritis. Contrast-enhanced CT (Figs. 2 and 3) confirmed pancreatic enlargement with loss of lobulations and a "sausage-like" appearance, along with a simple cystic lesion in the pancreatic tail. It also identified bilateral renal nephritis and enlarged lymph nodes in peripancreatic and mediastinal regions. MR cholangiopancreatography (Fig. 4) corroborated the findings from both ultrasound and CT, while providing additional insights into the biliary system, revealing stenosis and dilation of the right intrahepatic bile ducts, as well as circumferential thickening and filiform stenosis of the distal main bile duct. These imaging findings, encompassing pancreatic, renal, biliary, and lymph node abnormalities, were consistent with IgG4-related disease.

To confirm our diagnosis, additional laboratory tests were conducted. These revealed elevated serum IgG4 levels at 3.76 g/L, while renal and autoimmune markers, including antinuclear antibodies, extractable nuclear antigen antibodies, and ANCA, were normal, as were tumor markers (CA19-9, AFP, and CEA). Infectious causes, including HBV and HCV, were excluded. However, biopsy confirmation was not feasible due to the inaccessibility of the affected sites. Still, the diagnosis was established using the 2019 ACR/EULAR classification criteria and the Revised 2020 Comprehensive Diagnostic Criteria for IgG4-related disease. Key criteria met by the patient included clinical evidence of multiorgan involvement (pancreas, kidneys, biliary tract, and lymph nodes), characteristic imaging findings (sausage-like pancreas, bilateral nephritis, and biliary strictures), and elevated serum IgG4 levels (>1.35 g/L). Differential diagnoses, including malignancy, vasculitis, and infections, were excluded. The combination of clinical, serological,

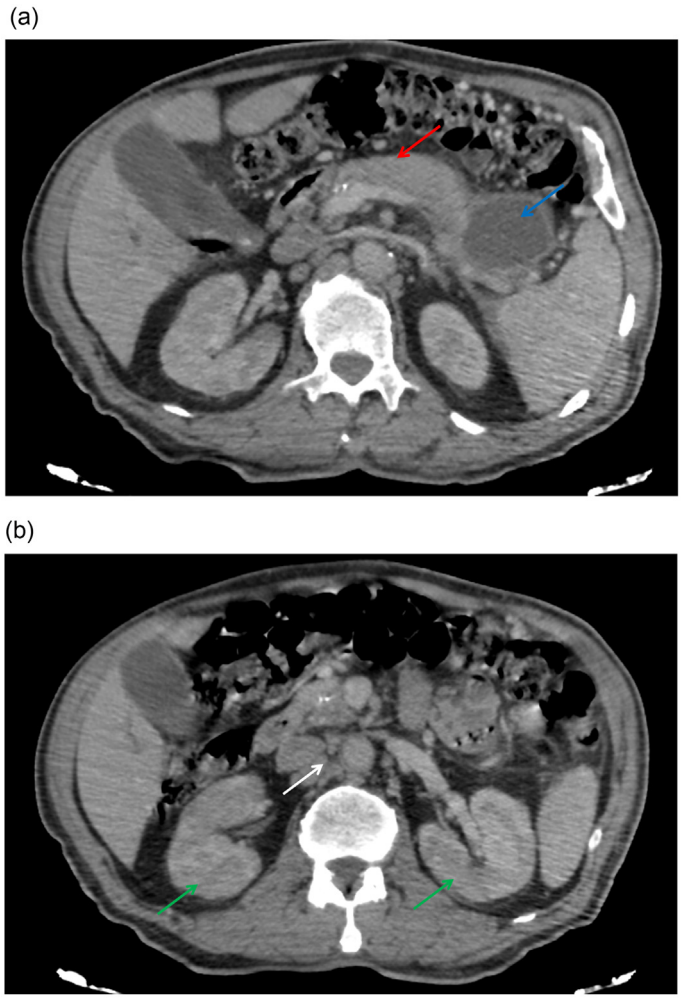

Fig. 2 – Axial contrast-enhanced CT images (A, B) showing: (A) swollen pancreas with loss of lobulations and a "sausage-like" appearance (red arrow), along with a simple cystic lesion in the pancreatic tail (blue arrow). (B) Bilateral peripheral renal lesions, triangular and hypodense (green arrows), and enlarged lymph nodes in the peripancreatic regions (withe arrow).

and radiological evidence strongly supports the diagnosis of IgG4-related disease, even in the absence of histological confirmation.

Corticosteroid therapy, the first-line treatment for IgG4-related disease, was initiated with oral prednisolone at 40 mg/day for 4 weeks, followed by a gradual tapering of 5 mg every 2 weeks to a maintenance dose of 5-10 mg/day over 3-6 months. The patient showed significant improvement, with abdominal pain resolving within 2 weeks. Biochemical follow-up demonstrated a decrease in serum IgG4 levels (from 3.76 g/L to 1.8 g/L at 3 months). Given the sustained response, steroid-sparing immunosuppressive therapy was not immediately necessary.

Discussion

First described as a distinct clinical entity in 2003 [1], IgG4-related disease is a systemic autoimmune disease marked by the infiltration of various organs by IgG4-positive plasma cells, leading to inflammation and fibrosis [2]. While the pancreas is the organ most frequently impacted, the disease is increasingly being recognized in extra-pancreatic organs as well, including the bile ducts, gallbladder, lymph nodes, retroperitoneum, mesentery, kidneys, lungs, breasts, prostate gland, and skin [3]. The head and neck region may also be involved, with the salivary and lacrimal glands, orbits, thyroid, pituitary gland, and meninges being the most commonly affected [4].

IgG4-related disease is a relatively rare condition, but its true prevalence and incidence are still being defined due to increasing recognition and diagnosis [5].In Japan, where the disease has been most extensively studied, the prevalence is estimated to be around 6-30 per 100,000 individuals [6]. It typically affects middle-aged and older adults, with the average age of onset being between 50 and 60 years [7]. Men are affected more often than women, with the possible exception of those with principally head and neck involvement where the gender distribution appears to be more balanced [8].

The pathogenesis of IgG4-related disease remains largely unclear, with evidence suggesting features of both autoim-

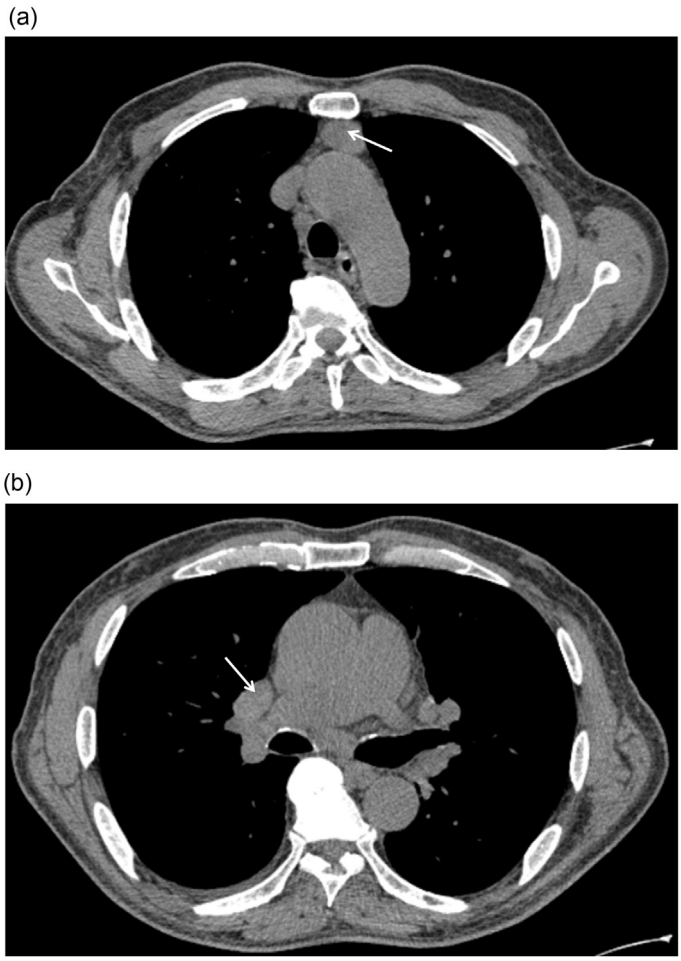

Fig. 3 – Axial nonenhanced thoracic CT images showing mediastinal lymphadenopathy (withe arrows).

mune and allergic conditions. IgG4, the least prevalent subclass of IgG, typically maintains stable serum levels [9]. Although IgG4 is commonly found in affected organs, its precise role in the development of the disease continues to raise numerous unanswered questions [10].

The clinical manifestations of IgG4-related disease can vary significantly based on the organ involved, typically presenting as subacute or chronic conditions. Often, the disease is discovered incidentally during clinical examinations, such as thyroid enlargement, swelling of salivary or lacrimal glands, or lymphadenopathy [11]. Constitutional symptoms like fever, malaise, and night sweats are infrequent [12]. Tumefaction may present as localized masses or nodules, affecting areas such as the orbit, kidney, and lung, or as diffuse enlargement of organs like the pancreas [13,14]. Severe complications can arise from obstruction or compression, including obstructive jaundice associated with autoimmune pancreatitis or IgG4 sclerosing cholangitis, visual disturbances due to IgG4-related dacryoadenitis, and hydronephrosis linked to IgG4-related retroperitoneal fibrosis [13,14]. Additionally, about 40% of patients may present symptoms related to asthma or allergies [15].

Hematological tests in IgG4-related disease are typically normal, though some patients may exhibit slightly elevated C-reactive protein, elevated ESR, anemia, thrombocytosis, and mild eosinophilia [5]. Serological findings may include elevated IgE, polyclonal hypergammaglobulinemia, hypocomplementemia, and the presence of antinuclear antibodies and rheumatoid factor [16,17]. Elevated serum IgG4, with a cut-off value of 1.35 g/L, shows high sensitivity (97.0%) but moderate specificity (79.6%) for diagnosing IgG4-related disease [18]. However, some patients with early or limited disease may not have high IgG4 levels. Additionally, elevated serum IgG4 can occur in non-IgG4-related disease conditions, reducing its specificity and positive predictive value [5].

Imaging plays a crucial role in the diagnosis of IgG4-related diseases, as these conditions can affect multiple organs with a variety of manifestations. The radiological features are often essential for identifying the extent of disease involvement and guiding further diagnostic procedures. However, the imaging characteristics vary significantly depending on the organ involved:

1- Pancreas: In IgG4-RD, pancreatic involvement often presents as autoimmune pancreatitis, characterized by 2 patterns: diffuse and focal [19,20]. While the diffuse form shows uniform pancreatic enlargement with a "sausage-like" appearance, the focal form, often involving the pancreatic head, can mimic malignancy [21]. A distinct feature observed on

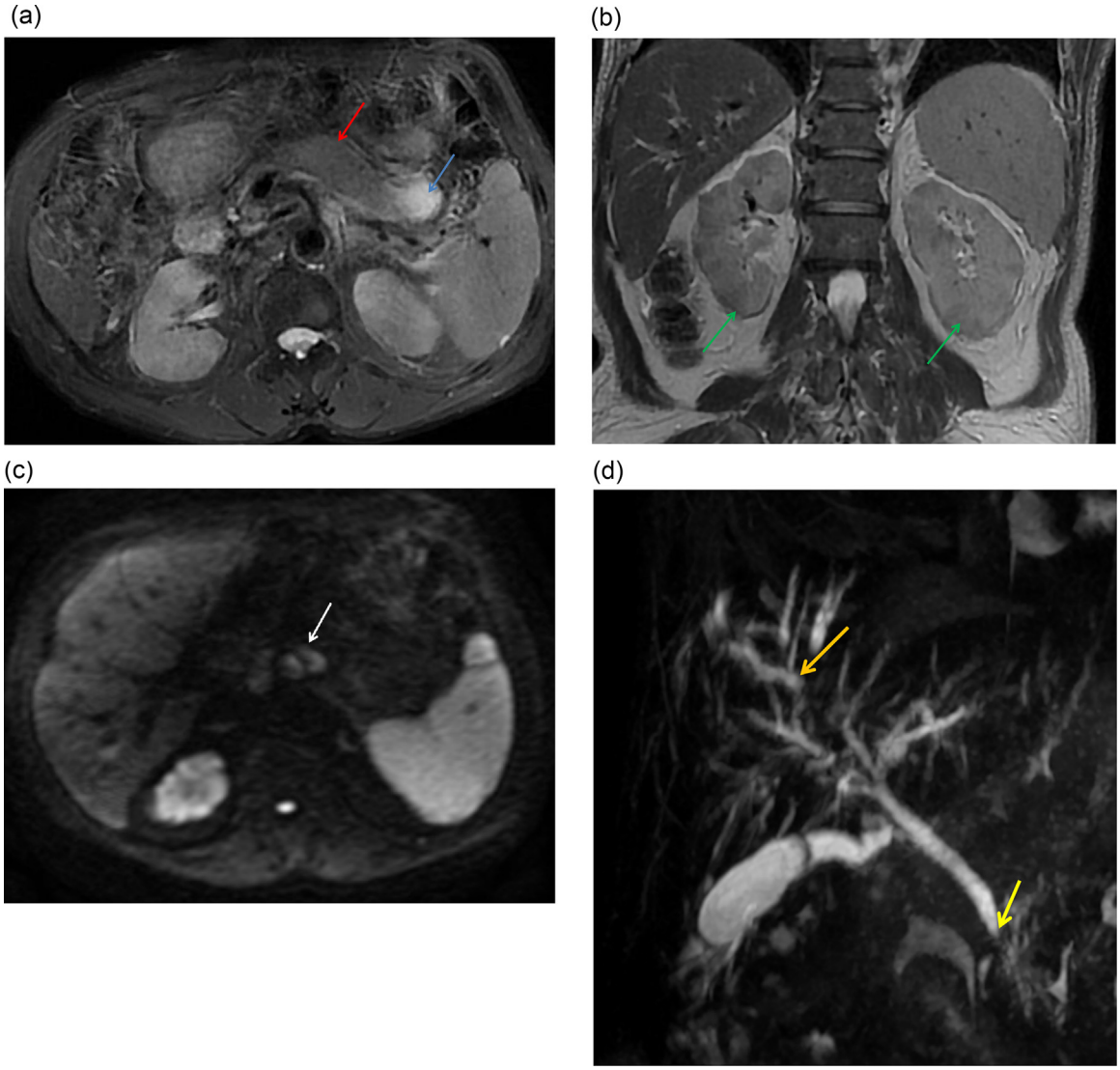

Fig. 4 – Magnetic resonance cholangiopancreatography: (A) Axial T2-weighted MR image shows enlargement of the pancreas with a "sausage-like" appearance (red arrow), and a thin-walled cyst in the tail, with homogeneous content (blue arrow). (B) Coronal T2-weighted MR image show hypointense peripheral renal lesions (green arrows). (C) Axial diffusion-weighted MR image (b = 800 sec/mm^2) revealing peripancreatic lymphadenopathy (withe arrows). (D) Coronal MR cholangiopancreatographic image shows several areas of stenosis and dilation of the right intrahepatic bile ducts (orange arrow), with filiform stenosis at the distal level of the main bile duct (yellow arrow).

imaging is the capsule-like rim surrounding the pancreas, especially evident on contrast-enhanced CT or MRI [22]. This finding, along with narrowing of the main pancreatic duct without significant upstream dilation, aids in differentiating IgG4-related pancreatitis from pancreatic cancer [22]. Additionally, IgG4-RD-related pancreatic lesions typically exhibit mild to moderate enhancement postcontrast, with potential perilesional inflammation, contrasting with the intense and heterogeneous enhancement seen in pancreatic cancer.

2- Bile Ducts Hepatobiliary involvement in IgG4-RD frequently presents with bile duct wall thickening and stenosis, often in the intrapancreatic segment of the common bile duct [22,23]. Smooth, symmetrical thickening around the bile ducts is a hallmark feature, contrasting with the irregularities seen in primary sclerosing cholangitis or cholangiocarcinoma [22]. MR cholangiopancreatography is particularly useful for evaluating these changes and for excluding other pathologies.

3- Kidneys: Renal involvement in autoimmune pancreatitis occurs in up to 35% of patients, displaying 5 distinct radiological patterns: bilateral round or wedge-shaped peripheral cortical, diffuse patchy involvement, a rim of soft tissue surrounding the kidney, bilateral nodules in the renal sinuses, and diffuse wall thickening of the renal pelvis [24]. Following contrast administration, these lesions are hypoattenuating compared to normal renal cortex in the arterial phase, becoming isoattenuating in later phases. MRI shows low signal intensity on both T1- and T2-weighted images, with mild enhancement on T1-weighted images after contrast [24]. The presence of renal lesions helps differentiate autoimmune pancreatitis from pancreatic cancer, as they strongly indicate the former [25].

4- Lacrimal and Salivary Glands: In cases involving the lacrimal and salivary glands, radiologic findings show glandular enlargement. On CT, there is uniform enhancement of the affected glands postcontrast. MRI reveals low to intermediate signal intensity on T2-weighted images and low signal intensity on T1-weighted images, with consistent enhancement after gadolinium [2]. Differential diagnoses include viral infections, Sjögren's syndrome, lymphoma, sarcoidosis, and granulomatosis with polyangiitis. If the involvement is unilateral, malignancy should be considered [2].

5- Orbits: Recent studies indicate that many cases of idiopathic orbital inflammation, known as orbital pseudotumors, are linked to IgG4-related disease [26]. These lesions can present unilaterally or bilaterally and may involve the entire orbit or specific structures such as the extraocular muscles, lacrimal system, and optic nerves [27]. On CT scans, orbital pseudotumors exhibit soft tissue attenuation with homogeneous enhancement, while MRI findings show low signal intensity on T2-weighted images relative to brain tissue and hyperintensity on diffusion-weighted images due to fibrosis [28].

6- Thyroid: IgG4-related disease can affect the thyroid in 2 main forms: Riedel thyroiditis and the fibrous variant of Hashimoto thyroiditis. Riedel thyroiditis presents as a firm, painless mass due to extensive fibrosis. On CT scans, it typically shows either focal or diffuse low attenuation with minimal contrast enhancement compared to normal thyroid tissue. In some Hashimoto thyroiditis cases, the histological appearance can mimic that of IgG4-related disease [22].

7- Lymphadenopathy: Concomitant lymphadenopathy is often seen in patients with IgG4-related disease, with hilar lymphadenopathy occurring in a significant proportion of patients with autoimmune pancreatitis [29]. This involvement typically extends to mediastinal, axillary, cervical, and intra-abdominal lymph nodes. Lymph nodes in IgG4-related disease are usually small (<2 cm), and patients do not present with fever or weight loss, which helps distinguish this disease from lymphoma and Castleman disease [22].

8- Retroperitoneal Fibrosis: Often seen in patients with autoimmune pancreatitis, it presents radiologically as a soft-tissue mass that envelops the abdominal aorta and its branches, potentially entrapping the ureters and causing hydronephrosis and hydroureter [22]. On cross-sectional imaging, IgG4-related retroperitoneal fibrosis shows homogeneous late contrast enhancement and features irregular margins, indicating periaortic lesions or IgG4-related periaortitis. These lesions are typically nonstenotic and may be associated with aortic dilation [30,31].

9- Mesentery: Sclerosing mesenteritis is a rare chronic disorder affecting the small bowel mesentery, often linked to other fibroinflammatory conditions [22]. Imaging typically shows a soft-tissue mass encasing the mesenteric vessels, which can resemble other mesenteric diseases [22]. However, a distinguishing feature on CT is the "fat ring sign," indicating preserved fat around the mesenteric vessels, which helps differentiate IgG4-related mesenteritis [32]. In some cases, partial or complete small intestine obstruction may be observed.

10- Lungs: IgG4-related pulmonary disease in autoimmune pancreatitis includes 4 main radiological patterns: solid nodular or masslike lesions, round ground-glass opacities, alveolar interstitial disease, and bronchovascular disease [33]. Solid nodules or masslike lesions may mimic lung cancer, while round ground-glass opacities can resemble bronchoalveolar carcinoma on CT [22]. The alveolar interstitial pattern, marked by ground-glass and honeycomb components, resembles nonspecific interstitial pneumonia [22]. Bronchovascular disease shows interlobular septal thickening and often enlarged hilar or mediastinal lymph nodes, similar to sarcoidosis [22]. Pleural thickening and subpleural lung involvement may also occur [22].

Other organ involvement in IgG4-related disease is rare but may show distinctive radiological signs. In the gastrointestinal tract, this includes gastric wall thickening and ulcer formation [34]. IgG4-related prostatitis presents radiologically with prostate enlargement, often accompanied by obstructive features on imaging [22]. Other rare sites, such as the testicles, breasts, pituitary gland, and meninges, may also display radiologic findings consistent with IgG4-related disease, characterized by tissue thickening and masses with inflammatory changes [35].

The role of imaging is crucial in differentiating IgG4-related disease (IgG4-RD) from malignancies and infections. Imaging techniques such as ultrasound, CT, and MRI provide key insights into the characteristic features of IgG4-RD, including diffuse or nodular tissue infiltration and fibro-inflammatory lesions. These features help distinguish IgG4-RD from other conditions, where malignancies typically present with focal masses or metabolic abnormalities, and infections show more homogeneous patterns.

The primary method for diagnosing IgG4-related disease is through identifying characteristic histological and immunohistochemical features, consistent across affected organs. It exhibits 3 main histopathological signs: (i) dense lymphoplasmacytic infiltrates, (ii) storiform-pattern fibrosis, and (iii) obliterative phlebitis [36]. Immunostaining for IgG4-positive plasma cells typically shows dense infiltration, with counts varying from 11 to over 200 cells per high-power field [36]. An IgG4 to IgG ratio above 40% is also essential, but these histological findings must be interpreted alongside clinical, radiological, and serological data, as histology alone cannot confirm the diagnosis [36].

However, histological confirmation may be infeasible in certain clinical scenarios. In cases where accessible biopsy sites are limited or when organ involvement is diffuse and involves multiple sites, obtaining histological confirmation may pose significant challenges. Additionally, in patients presenting with highly characteristic clinical, radiological, and serological findings—such as elevated serum IgG4 levels, typical imaging abnormalities, and the exclusion of differential diagnoses—histological confirmation may be deemed unnecessary.

The diagnostic criteria for IgG4-related disease have significantly evolved over time, transitioning from a reliance on histopathological findings to a comprehensive framework introduced by the ACR/EULAR in 2019 and refined in 2020 [37]. Initially focused on specific organ involvement and tissue biopsies, the understanding of IgG4-related disease has expanded to incorporate clinical, serological, and radiological data. The updated criteria include key components such as clinical findings indicating involvement of commonly affected organs (including the pancreas, salivary glands, lacrimal

glands, kidneys, or lungs) along with associated symptoms; serological evidence of elevated serum IgG4 levels, which supports diagnosis when combined with other criteria; and radiological findings from imaging studies (like CT or MRI) that reveal characteristic patterns of organ involvement, such as swelling and fibrosis [37]. While histological examination is not obligatory, if a biopsy is performed, it should demonstrate typical features like dense lymphoplasmacytic infiltration and an increased number of IgG4-positive plasma cells [37]. The criteria also emphasize the importance of excluding other conditions that can mimic IgG4-related disease, such as malignancies, other autoimmune diseases, and infections [37]. This comprehensive framework represents a pivotal shift by allowing for diagnosis without mandatory histological confirmation, facilitating timely identification and treatment, while recognizing the challenges of obtaining biopsies and emphasizing the integration of multiple diagnostic modalities.

This case aligns with the ACR/EULAR 2019 and 2020 criteria, the diagnosis of IgG4-related disease was supported by characteristic imaging features, elevated serum IgG4 levels, and the exclusion of differential diagnoses, even in the absence of histological confirmation. These criteria provided a robust framework for establishing the diagnosis, integrating clinical, radiological, and serological evidence to ensure a comprehensive assessment.

In the literature, several cases of IgG4-related disease have been reported, featuring variations in clinical and radiological presentations. Our case stands out due to several unique aspects, particularly its initial presentation without immediate histological evidence. Unlike many cases where the diagnostic approach relies heavily on histology, our diagnosis was primarily supported by clinical, biological, and radiological data, which is less common but highlights the evolving diagnostic strategies for this condition. Additionally, the rapid response to treatment further underscores the reliability of nonhistological diagnostic methods in managing IgG4-related disease.

However, nonhistological diagnosis poses challenges. While elevated IgG4 levels and imaging provide crucial information, they may not always be sufficient to capture the full complexity of the disease, especially in cases with overlapping symptoms or atypical presentations. This raises the risk of misdiagnosis or the need for additional invasive procedures. A balanced approach integrating clinical judgment, imaging, and serological data is essential to ensure accurate and comprehensive diagnosis.

The treatment of IgG4-related disease primarily focuses on reducing inflammation and managing symptoms, with corticosteroids being the first-line therapy. Prednisone is commonly used, and the dosage is tailored to the severity of the disease, often resulting in significant improvement within weeks [5]. Additionally, monitoring serum IgG4 levels serves as a valuable marker for assessing disease activity and response to treatment. A decline in IgG4 levels correlates with disease improvement, while persistently elevated levels may indicate ongoing disease activity or the need for further therapeutic adjustments.

For patients who do not respond adequately to corticosteroids or experience relapses, additional immunosuppressive agents like azathioprine, mycophenolate mofetil, and methotrexate may be considered [5]. Rituximab, a monoclonal antibody targeting CD20-positive B cells, is also effective for refractory cases [5]. Regular monitoring is essential to assess treatment response and manage side effects, with long-term follow-up necessary due to the possibility of relapses. Treatment plans should be individualized based on the patient's clinical presentation and response to therapy.

Conclusion

This case highlights the diagnostic challenges of IgG4-related disease, particularly when histological confirmation is not feasible. By integrating clinical, serological, and imaging findings, a robust diagnosis was achieved, and corticosteroid therapy led to significant improvement. The case underscores the value of comprehensive diagnostic criteria and the need for continued research into noninvasive diagnostic tools and long-term management strategies.

Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

Author contributions

All authors contributed to this work. All authors have read and approved the final version of the manuscript.

Patient consent

Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

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