Immunoglobulin G4-related disease manifesting as a sino-orbital mass: a comprehensive case report and review of literature

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Background: Immunoglobulin G4-related disease (IgG4-RD) is a rare autoimmune fibroinflammatory condition that can affect multiple organs. Sinonasal and orbital involvement is exceedingly uncommon and poses significant diagnostic challenges. These manifestations often mimic malignancy, chronic infection, or other inflammatory disorders, leading to delayed diagnosis and repeated investigations. This study aims to illustrate the diagnostic challenges and management of IgG4-RD and to increase awareness of this rare presentation among clinicians.

Case Description: We present a case of a 52-year-old male with a history of diabetes mellitus who developed a left-sided nasal mass extending into the orbit, resulting in nasal obstruction, epistaxis, proptosis, and diplopia. Imaging revealed a sinonasal mass with orbital extension through the lamina papyracea, raising suspicion for invasive or neoplastic pathology. Initial biopsies were inconclusive, demonstrating chronic inflammatory changes with no evidence of malignancy, granulomatous disease, or invasive fungal infection. Despite surgical debulking and close follow-up, symptoms persisted and recurred. Multiple subsequent biopsies were performed, which continued to show non-specific inflammatory findings. Further evaluation revealed elevated serum IgG4 levels, and repeat histopathological analysis demonstrated dense lymphoplasmacytic infiltration with an increased number of IgG4-positive plasma cells and an elevated IgG4/IgG ratio, consistent with IgG4-related disease. Correlation of clinical features, radiologic findings, laboratory results, and histopathology confirmed the diagnosis of IgG4-RD involving the sinonasal cavity and orbit.

Conclusions: This case emphasizes the importance of maintaining a high index of suspicion for IgG4-RD in patients presenting with sino-nasal-orbital masses, considering the disease's rarity and non-specific initial symptoms. Early diagnosis through repeated tissue sampling and comprehensive evaluation is essential to avoid misdiagnosis and unnecessary interventions.

Keywords: Immunoglobulin G4-related disease (IgG4-RD); sinonasal; orbital; rituximab; case report

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Introduction

Immunoglobulin G4-related disease (IgG4-RD) was first documented in 2001 in a study from Japan involving individuals with autoimmune pancreatitis who exhibited elevated serum IgG4 levels (1). Subsequent investigations revealed fibroinflammatory lesions affecting various organs beyond the pancreas in patients with autoimmune pancreatitis. These lesions manifested in the biliary tree, retroperitoneum, salivary glands, orbit, lymph nodes, kidneys, lungs, meninges, aorta, breast, prostate, thyroid, pericardium, and skin (2-4). IgG4-RD has been identified as the cause in 17–60% of cases categorized as idiopathic orbital inflammation (5-7). The incidence of acute inflammatory polyarthritis attributed to IgG4-RD is estimated at 0.28–1.08 cases per 100,000 individuals in Japan (8,9), with indications that the actual incidence might be higher due to potential under recognition and the omission of extra-pancreatic involvement from statistical analyses (10). No reported incidents have occurred in the USA, and epidemiological data regarding pediatric populations remain scarce, with only a few cases documented among children aged 5–15 years old, where ocular involvement was noted in 44% of pediatric patients (11).

Diagnosing IgG4-RD can pose challenges, as diagnostic approaches vary depending on the anatomical site affected. A comprehensive evaluation is essential to rule out numerous disorders that can mimic IgG4-RD. This spectrum of conditions includes malignancies, lymphoproliferative disorders, antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis, sarcoidosis, Sjögren's syndrome, Castleman's disease, and various other ailments (8,12). Sinonasal involvement in IgG4-RD is exceedingly rare, with only a few reported cases documented globally, and none reported in Saudi Arabia (13). In this report, we present an atypical case of IgG4-RD featuring nasal bleeding and a sino-orbital mass. We present this article in accordance with the CARE reporting checklist (available at https://acr.amegroups.com/article/view/10.21037/acr-2025-148/rc).

Highlight box

Key findings

- This case demonstrates a rare presentation of immunoglobulin G4-related disease (IgG4-RD) with sinonasal and orbital involvement in a diabetic patient, successfully managed with glucocorticoids and rituximab.

What is known and what is new?

- IgG4-RD is recognized for its multisystem involvement, but sinonasal manifestations are rare and often mimic malignancies or chronic infections.
- This report adds to the limited literature on sino-orbital presentations and underscores the importance of repeated biopsies and serum IgG4 evaluation.

What is the implication, and what should change now?

- Clinicians should consider IgG4-RD in the differential diagnosis of persistent sinonasal-orbital masses, especially in patients with risk factors such as diabetes. Early recognition and appropriate therapy, including glucocorticoid-sparing agents like rituximab, can improve long-term outcomes and reduce recurrence.

Case presentation

A 52-year-old male with a history of diabetes mellitus (DM) presented in July 2022 with chronic left nasal obstruction, discharge, bleeding, and subsequent left orbital symptoms, including proptosis, diplopia, and left eye pain that began two weeks earlier. Initial assessments revealed a granulomatous mass in the left nasal cavity extending into the orbit, which was tender to touch. Radiological imaging confirmed an enhancing left sinonasal mass involving the middle meatus components and extending to the left orbit (*Figure 1*). Intraoperative findings also showed that this was a single, continuous mass extending directly through the lamina papyracea into the orbital cavity. Surgical debulking of the mass was performed, and biopsy results showed inflammatory tissue infiltrated with lymphocytes, testing negative for fungal infection and malignancy. The patient was discharged and closely monitored. Partial improvement was noted with nasal glucocorticoid irrigations, which worsened upon cessation. Extensive laboratory workup revealed an erythrocyte sedimentation rate (ESR) of 50 mm/h, indicating ongoing inflammation, while normal C-reactive protein (CRP) levels ruled out acute inflammation. Negative ANCA results excluded ANCA-associated vasculitis, and a negative purified protein derivative (PPD) test ruled out tuberculosis at a 2-millimeter reading. The radiological examination showed improved outcomes (*Figure 2*).

Two months after surgery, the patient presented to the emergency department with progressively worsening left orbital symptoms, prompting further debulking and biopsy, which yielded inconclusive results. Despite postoperative improvement with nasal glucocorticoid irrigation, symptoms recurred. Given the persistence of symptoms and the recurrence of the left sino-orbital mass, a third biopsy was

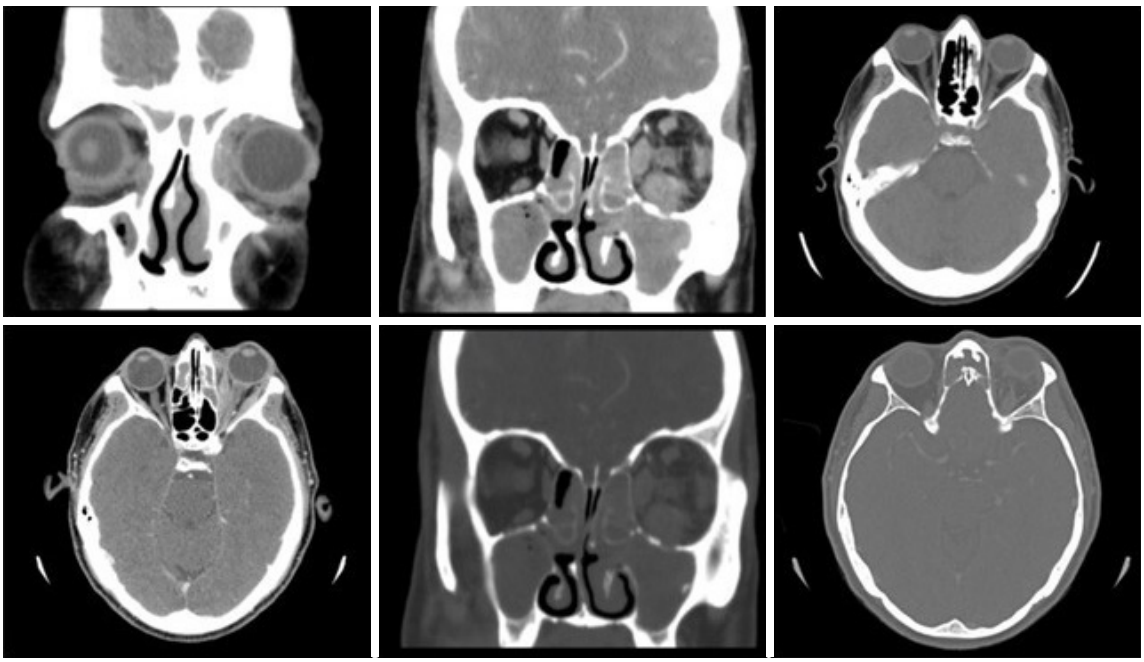

Figure 1 Pre-operative radiological imaging confirmed an enhancing left sinonasal mass involving the middle meatus components and extending to the left orbit.

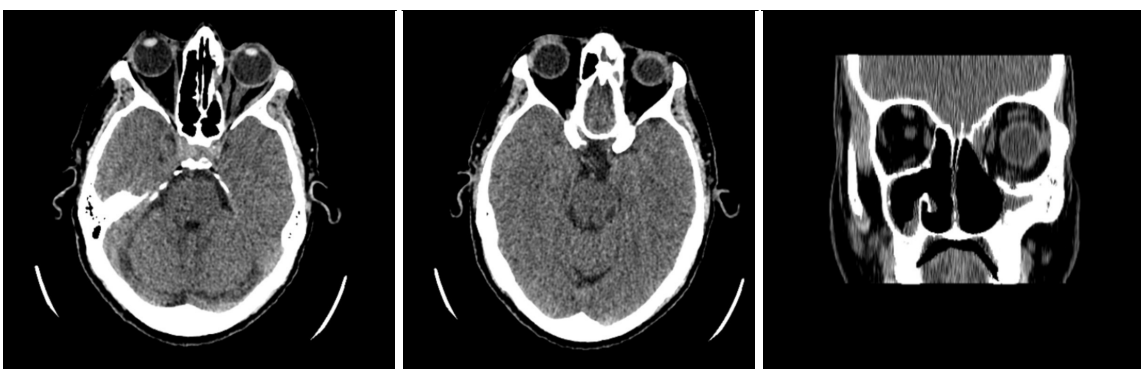

Figure 2 Post-operative radiological imaging.

performed for histopathological analysis and fungal culture to rule out lymphoma or invasive fungal infection; however, the findings remained inconclusive. After discussion with the pathologist and slide review, there was a raised suspicion of IgG4-RD.

Histopathological examination of the orbital wall specimen revealed areas of dense fibrosis and lymphoid aggregates with heavy lymphoplasmacytic infiltrates (*Figure 3*). Numerous plasma cells were identified, with more than 80 IgG4-positive plasma cells per high-power field and an IgG4/IgG ratio exceeding 40%. The infiltrate was diffusely distributed. Storiform fibrosis was observed, while obliterative phlebitis was not detected. Importantly, there were no granulomas, necrosis, or prominent eosinophilic or neutrophilic infiltrates, thus excluding eosinophilic chronic rhinosinusitis and other differential diagnoses. These findings, combined with elevated serum IgG4 levels, were diagnostic of IgG4-RD.

Differentials such as granulomatosis with polyangiitis (GPA), Erdheim-Chester disease, Rosai-Dorfman disease, xanthogranulomatous inflammation, and tumefactive fibroinflammatory lesions (TFIL) were excluded based on

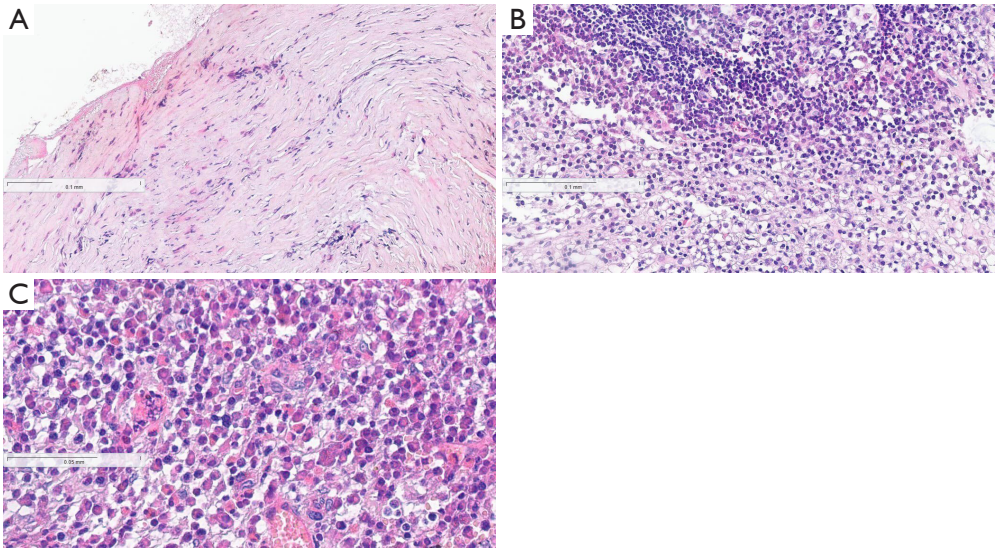

Figure 3 Histopathological features of the sino-orbital IgG4-related lesion. (A) H&E-stained section showing dense fibrosis. (B) H&E-stained section demonstrating lymphoid aggregates with heavy lymphoplasmacytic infiltrate. (C) Numerous plasma cells are seen (H&E-stained). H&E, hematoxylin and eosin; IgG4, immunoglobulin G4.

negative ANCA tests, absence of granulomas, necrosis, neutrophilic infiltration, and lack of emperipolesis or xanthomatous features. Histology revealed a dense lymphoplasmacytic infiltrate with storiform fibrosis and an IgG4/IgG ratio greater than 40%, supporting the diagnosis of IgG4-RD.

Serum IgG subclass levels were subsequently assessed, revealing elevated levels of serum IgG (2,679 mg/dL) and IgG4 (1,027 mg/dL), with an IgG4/IgG ratio >40%. A rheumatological consultation was sought, and the patient initiated treatment with prednisolone 60 mg, resulting in a significant improvement in left eye protrusion and periorbital edema, with no associated fever, cough, or chest pain. Additionally, rituximab was administered as part of the management plan. Regular follow-up over more than one year demonstrated complete remission of all sinonasal and orbital symptoms, with no recurrence. The patient received a second cycle of rituximab 6 months after the initial treatment, with no recurrence of sinonasal symptoms observed. All procedures performed in this case were in accordance with the ethical standards of the Jazan Health Ethics Committee, King Fahd Central Hospital (No. 2347), and with the Helsinki Declaration and its subsequent amendments. Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

Discussion

It is imperative to acknowledge that IgG4-RD represents a relatively recent medical entity, and consequently, much remains to be understood regarding its etiology, diagnostic modalities, and treatment strategies. Further research efforts are warranted to delineate optimal diagnostic criteria, therapeutic approaches, and long-term prognostic outcomes for affected individuals, with particular emphasis on longitudinal studies tracking disease trajectories.

IgG4-RD constitutes a rare form of autoimmunity that has garnered increased attention in recent years. Due to its elusive nature, diagnosis poses significant challenges as its symptoms can mimic those of other disorders, including infections and neoplastic diseases. Therefore, histological confirmation through tissue biopsy is imperative to rule out alternative pathological processes (8,12). Additionally, a comprehensive evaluation encompassing clinical presentation, imaging findings, and laboratory results is indispensable (8,12,14). Characteristically, IgG4-RD is typified by the infiltration of affected tissues by IgG4-positive plasma cells, culminating in fibrosis and immune-mediated tissue damage (14). While manifestations can occur in various organs such as the eyes, salivary glands, lacrimal glands, and pancreas (8,12), the most commonly

Table 1 Characteristics, treatment strategies, and outcomes of cases in the literature

References	Age, years	Sex	Organ manifestation	Serum IgG4, mg/dL	Therapy	Comments
Prabhu *et al.* [2014] (20)	15	Female	Nasal cavity, maxillary sinus, naso-lacrimal duct	206	Prednisolone	Follow-up
Prabhu *et al.* [2014] (20)	15	Female	Nasal cavity, orbit, pterygopalatine fossa, cavernous sinus	579	Rituximab	Inadequate response to prednisone
Song *et al.* [2015] (21)	72	Male	Sinonasal mass involving the maxillary and anterior ethmoid sinuses	94.5	Long-term oral glucocorticoids	Clinical improvement post-surgery; sustained resolution at 6 months
Hess *et al.* [2022] (22)	19	Male	Lateral orbital floor, erosion into maxillary sinus	24	Prednisone, IV methylprednisolone and RTX added after disease progressed	Symptomatic improvement at 8 months
Hess *et al.* [2022] (22)	66	Female	Maxillary sinus, midline septum and palate	56	Prednisone, RTX added after disease progressed	Symptomatic improvement at 4 years

IgG4, immunoglobulin G4; IV, intravenous; RTX, rituximab.

involved organs include the pancreas, biliary tract, salivary and lacrimal glands, kidneys, thyroid gland, lungs, and aorta (15).

Clinical presentations of IgG4-RD exhibit diversity contingent upon the affected organ. In orbital involvement, the predominant symptoms typically encompass painless periorbital and/or eyelid swelling, erythema, and proptosis, often accompanied by lacrimal gland involvement (6,16). Demographically, IgG4-RD displays a predilection for older males over females (ratio: 2.8–7.5:1) (17,18). However, orbital involvement appears to affect a relatively younger patient cohort, with a more balanced gender distribution (ratio: 1.3:1), often associated with salivary gland lesions and elevated serum IgG4 levels (19). The presented case involves a male patient with a history of DM presenting with a sinonasal mass extending into the orbit, representing a rare manifestation of IgG4-RD orbital involvement.

In our cases, the atypical presentation involving nasal bleeding and blockage in a patient with DM diverted attention away from this condition. As depicted in *Table 1*, reported cases of IgG4-RD encompass a spectrum of organ involvement. The majority of affected individuals were children, with the median age being 15 years, and only one elderly male case was aged 72 years. In most instances, serum IgG4 levels were elevated, ranging from 206 to 579 mg/dL. However, in our case, the IgG4 level was notably higher, at 1,027 mg/dL.

Treatment strategies for IgG4-RD aim to alleviate inflammation, achieve remission, and preserve organ function. Initial therapy typically entails oral glucocorticoids (e.g., prednisone 30–40 mg once daily), administered for 2–4 weeks and then gradually tapered over 2–3 months. Rituximab serves as a viable glucocorticoid-sparing alternative, particularly in patients with contraindications to glucocorticoid (e.g., uncontrolled diabetes), and can be employed to induce or sustain remission in individuals who are unable to tolerate glucocorticoid tapering or experience disease relapse within 12 months of glucocorticoid cessation. In most of the cases documented, glucocorticoids such as prednisone or prednisolone were employed, sometimes supplemented with immunosuppressive agents like mycophenolate mofetil, azathioprine, or rituximab in select scenarios. The addition of rituximab was warranted in cases demonstrating inadequate response to prednisolone, with ongoing follow-up to monitor treatment efficacy. Rituximab, a monoclonal antibody targeting CD20-positive B cells, has shown promising results in prospective studies, with 77% of patients achieving the primary outcome of response without relapse for up to 6 months (23). Additional trials have further supported the efficacy of rituximab in both initiating and sustaining remission in IgG4-RD patients refractory to prior immunosuppressive therapies (21,24). While some cases necessitated alternative interventions and maintenance therapy due to inadequate initial response, the majority exhibited favorable clinical outcomes or effective disease management with the selected therapeutic regimen.

In this particular instance, the clinical presentation of a sinonasal bleeding mass extending to the orbit initially

prompted considerations of chronic invasive fungal sinusitis or lymphoma rather than IgG4-RD. The patient's age >50 years and a documented history of DM factored into the evaluation of these potential differential diagnoses. Moreover, sinonasal-orbital involvement represents an exceedingly rare manifestation of IgG4-RD. Notably, presentation with a sinonasal mass extending to the orbit has been reported in only one documented case (20). In addition, two pediatric cases of orbital IgG4-RD have been documented, characterized by orbital masses with IgG4-positive plasma cell infiltration, managed successfully with surgical debulking or immunosuppressive therapy, and without evidence of systemic involvement (25). A meta-analysis of IgG4-RD cases revealed orbital involvement in merely 101 out of 3,034 patients (incidence: 3.33%) (26).

This case is strengthened by a comprehensive diagnostic approach, including serial biopsies, multidisciplinary team involvement, and radiologic follow-up, leading to accurate diagnosis and effective management. However, it is limited by the initial diagnostic ambiguity due to non-specific biopsy findings and the rarity of sinonasal-orbital IgG4-RD, which may delay recognition in similar cases.

Diagnosing IgG4-RD can be challenging, given that elevated plasma IgG4 levels lack both specificity and sensitivity for definitive diagnosis. Sekiguchi *et al.* [2016] reported that normal IgG4 levels were observed in approximately one-fourth of patients (27), and utilizing a cutoff level of 135 mg/dL for IgG4 yielded a positive predictive value for IgG4-RD of merely 34% (23). However, in our case, the diagnosis was substantiated by elevated serum IgG4 levels, an elevated ESR, and a positive histopathological report demonstrating IgG4-rich plasma cells with an IgG4/IgG ratio >40% in the left sinonasal orbital mass.

There has been a reported response rate of 97 percent for glucocorticoids when used alone as initial therapy for IgG4-RDs (26). Glucocorticoids remain the primary agents for initial therapy. In the aforementioned meta-analysis, glucocorticoids were the most frequently employed medication to initiate therapy (74% of reported cases), with a mean starting dose of 0.6 mg/kg (26). In our case, the patient received prednisolone according to a tapering schedule, resulting in complete improvement of both left sinonasal and left eye manifestations. However, recurrence may ensue upon discontinuation of glucocorticoids (28). It is prudent to consider adding a glucocorticoid-sparing agent, especially if prednisone cannot be tapered due to the presence of chronically active disease (12). Glucocorticoid-sparing medications such as methotrexate, azathioprine, mycophenolate, 6-mercaptopurine, and cyclophosphamide may be warranted (26). The use of rituximab as a monotherapy to achieve remission has demonstrated promising outcomes, with significant decreases in IgG4 levels and circulating plasma blasts observed post-treatment (21,24). In our case, the presence of DM precluded long-term glucocorticoid therapy, thus rituximab was chosen as the preferred option.

Presentation with a sinonasal mass extending to the orbit has now been described across multiple cohorts (Hess *et al.*, 2022; Yamamoto *et al.*, 2025), underscoring that this manifestation, although still rare, is more prevalent than initially (22,29).

This case highlights the importance of considering IgG4-RD in patients with recurrent sino-orbital masses, especially when routine treatments fail and histopathological findings are inconclusive. Enhanced awareness among otolaryngologists, ophthalmologists, and pathologists is essential to facilitate earlier diagnosis and intervention, potentially preventing irreversible complications and improving patient outcomes.

Conclusions

Diagnosing IgG4-RD presenting with sinonasal manifestations necessitates a high index of suspicion following comprehensive evaluation. Tissue sampling is imperative to exclude alternative disease processes. While glucocorticoids remain the primary initial therapeutic agents recommended for IgG4-RDs, glucocorticoid-sparing medications, such as rituximab, may be preferable in select cases and have shown favorable outcomes. Further research is warranted to determine the most effective diagnostic criteria and treatment strategies for IgG4-RD.

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Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at https://acr.amegroups.com/article/view/10.21037/acr-2025-148/rc

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Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this case were in accordance with the ethical standards of the Jazan Health Ethics Committee, King Fahd Central Hospital (No. 2347), and with the Helsinki Declaration and its subsequent amendments. Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

References

1. Hamano H, Kawa S, Horiuchi A, et al. High serum IgG4 concentrations in patients with sclerosing pancreatitis. N Engl J Med 2001;344:732-8.
2. McNab AA, McKelvie P. IgG4-related ophthalmic disease. Part I: background and pathology. Ophthalmic Plast Reconstr Surg 2015;31:83-8.
3. Divatia M, Kim SA, Ro JY. IgG4-related sclerosing disease, an emerging entity: a review of a multi-system disease. Yonsei Med J 2012;53:15-34.
4. Stone JH, Zen Y, Deshpande V. IgG4-related disease. N Engl J Med 2012;366:539-51.
5. Chen J, Zhang P, Ye H, et al. Clinical features and outcomes of IgG4-related idiopathic orbital inflammatory disease: from a large southern China-based cohort. Eye (Lond) 2021;35:1248-55.
6. Tsai CY, Kuo KT, Cheng AMS, et al. IgG4-related Ophthalmic Disease in Idiopathic Sclerosing and Non-Sclerosing Orbital Inflammation: A 25-Year Experience. Curr Eye Res 2019;44:1220-5.
7. Andron A, Hostovsky A, Nair AG, et al. The impact of IgG-4-ROD on the diagnosis of orbital tumors: A retrospective analysis. Orbit 2017;36:359-64.
8. Umehara H, Okazaki K, Nakamura T, et al. Current approach to the diagnosis of IgG4-related disease - Combination of comprehensive diagnostic and organ-specific criteria. Mod Rheumatol 2017;27:381-91.
9. Guma M, Firestein GS. IgG4-related diseases. Best Pract Res Clin Rheumatol 2012;26:425-38.
10. Wallace ZS, Deshpande V, Stone JH. Ophthalmic manifestations of IgG4-related disease: single-center experience and literature review. Semin Arthritis Rheum 2014;43:806-17.
11. Smerla RG, Rontogianni D, Fragoulis GE. Ocular manifestations of IgG4-related disease in children. More common than anticipated? Review of the literature and case report. Clin Rheumatol 2018;37:1721-7.
12. Khosroshahi A, Wallace ZS, Crowe JL, et al. International Consensus Guidance Statement on the Management and Treatment of IgG4-Related Disease. Arthritis Rheumatol 2015;67:1688-99.
13. Raina A, Yadav D, Krasinskas AM, et al. Evaluation and management of autoimmune pancreatitis: experience at a large US center. Am J Gastroenterol 2009;104:2295-306.
14. Wallace ZS, Mattoo H, Carruthers M, et al. Plasmablasts as a biomarker for IgG4-related disease, independent of serum IgG4 concentrations. Ann Rheum Dis 2015;74:190-5.
15. Brito-Zerón P, Ramos-Casals M, Bosch X, et al. The clinical spectrum of IgG4-related disease. Autoimmun Rev 2014;13:1203-10.
16. Harrison JD, Rodriguez-Justo M. Commentary on IgG4-related sialadenitis: Mikulicz's disease, Küttner's tumour, and eponymy. Histopathology 2011;58:1164-6.
17. Kim KP, Kim MH, Song MH, et al. Autoimmune chronic pancreatitis. Am J Gastroenterol 2004;99:1605-16.
18. Nishimori I, Tamakoshi A, Otsuki M, et al. Prevalence of autoimmune pancreatitis in Japan from a nationwide survey in 2002. J Gastroenterol 2007;42 Suppl 18:6-8.
19. Brito-Zerón P, Kostov B, Bosch X, et al. Therapeutic approach to IgG4-related disease: A systematic review. Medicine (Baltimore) 2016;95:e4002.
20. Prabhu SM, Yadav V, Irodi A, et al. IgG4-related disease with sinonasal involvement: A case series. Indian J Radiol Imaging 2014;24:117-20.

21. Song BH, Baiyee D, Liang J. A rare and emerging entity: Sinonasal IgG4-related sclerosing disease. Allergy Rhinol (Providence) 2015;6:151-7.
22. Hess AO, Lobo BC, Leon ME, et al. Sinonasal IgG4-related sclerosing disease: A rare entity and challenging diagnosis. Laryngoscope Investig Otolaryngol 2022;7:1725-32.
23. Naghibi M, Ahmed A, al Badri AM, et al. The successful treatment of IgG4-positive colitis with adalimumab in a patient with IgG4-related sclerosing disease--a new subtype of aggressive colitis?. J Crohns Colitis 2013;7:e81-e84.
24. Batu ED, Arici ZS, Orhan D, et al. Immunoglobulin G4-related orbital disease: report of two pediatric cases. Clin Exp Rheumatol 2015;33:409-10.
25. Griepentrog GJ, Vickers RW, Karesh JW, et al. A clinicopathologic case study of two patients with pediatric orbital IgG4-related disease. Orbit 2013;32:389-91.
26. Jariwala MP, Agarwal M, Mulay K, et al. IgG4-related orbital inflammation presenting as unilateral pseudotumor. Indian J Pediatr 2014;81:1108-10.
27. Sekiguchi H, Horie R, Kanai M, et al. IgG4-Related Disease: Retrospective Analysis of One Hundred Sixty-Six Patients. Arthritis Rheumatol 2016;68:2290-9.
28. Mittal R, Ganguly A, Rath S, et al. IgG4-related orbital inflammation presenting as bilateral proptosis in a child. Eye (Lond) 2014;28:1264-6.
29. Yamamoto M, Kanda M, Mizushima I, et al. Clinical profile of IgG4-related disease in Japan based on the rare disease data registry. Immunol Med 2025;48:192-202.

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