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Case-based Review

# Immunoglobulin G4-Associated Rosai–Dorfman Disease: Report of 3 Cases

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## Abstract

Rosai–Dorfman disease is characterized by dilated lymph node sinuses filled with lymphocytes, plasma cells, and histiocytes. Many of these histiocytes classically exhibit emperipolesis of lymphocytes and plasma cells. Abundant immunoglobulin G4+ plasma cells occur in some cases, and a potential relationship with immunoglobulin G4-related disease has been suggested. Here, we report 3 cases of immunoglobulin G4-associated Rosai–Dorfman disease. Immunoglobulin G4-related disease was suspected based on immunoglobulin G4+ plasma cell infiltration, but the final diagnosis was immunoglobulin G4-associated Rosai–Dorfman disease. At present, the evidence does not support a link between immunoglobulin G4-associated Rosai–Dorfman disease and immunoglobulin G4-related disease, and one condition should not be considered part of the spectrum of the other. We believe it is of paramount importance to increase the awareness of immunoglobulin G4-associated Rosai–Dorfman disease for pathologists who interpret the biopsies and clinicians who integrate the diagnosis and treat such patients to not overdiagnose immunoglobulin G4-related disease.

**Keywords:** Immunoglobulin G4-related disease, Rosai–Dorfman disease, sinus histiocytosis, lymphadenopathy, pachymeningitis

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## Introduction

Rosai–Dorfman disease (RDD) is part of the non-Langerhans cell histiocytosis.[1] Rosai–Dorfman disease may display an increased number of immunoglobulin G4 (IgG4)-positive plasma cells and an elevated IgG4/IgG ratio in the involved organs.[2] Furthermore, RDD may exhibit storiform fibrosis and obliterative phlebitis.[2,3] As such, the entity IgG4-associated RDD has been included as part of this group of histiocytosis.[4] Herein, we report 3 cases of IgG4-associated RDD and a review of the literature.

## Case Presentation

### Patient 1

A 52-year-old man presented with a 6-month history of lymphadenopathy in the neck, axillae, mediastinal and inguinal regions, parotid and submandibular gland enlargement, fever, and unintentional weight loss.

On examination, he was febrile, and the cervical, axillary, and inguinal lymph nodes were enlarged (Figure 1A). Laboratory tests showed normochromic normocytic anemia, thrombocytopenia, and

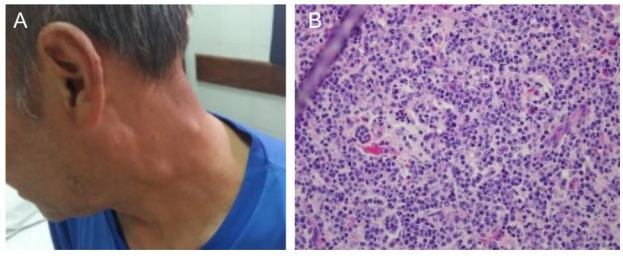


**Figure 1.** Clinical and histopathological findings in case 1. (A) Enlarged cervical lymph node. Lymph nodes were 0.8-1.2 inches in size, firm, and nontender. (B) Histological section stained with hematoxylin and eosin of cervical lymph node (40×) of the interfollicular area with the presence of lymphoplasmacytic infiltrates and histiocytes with abundant eosinophilic cytoplasm with intracytoplasmic lymphocytes (emperipolesis).

eosinophilia, increased erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP), low serum complement levels (C3 and C4), polyclonal hypergammaglobulinemia, high IgE levels, hypoalbuminemia, acute kidney injury with a serum creatinine of 4.14 mg/dL (normal range 0.50-0.90 mg/dL), and urinalysis with hematuria and 24-h proteinuria of 1518 mg. Serum IgG4 levels were normal. Antinuclear antibodies (ANA) were positive, with negative rheumatoid factor and anti-dsDNA, anti-SSA/Ro, anti-SSB/La, anti-Sm, and anti-RNP antibodies.

A kidney biopsy showed tubulointerstitial nephritis with infiltration of lymphocytes, plasma cells, and histiocytes and with mild mesangial proliferation, tubular atrophy (20%), and non-storiform interstitial fibrosis. Immunostaining showed 15 IgG4+ plasma cells/high-power field (HPF) with an IgG4/IgG ratio of 25%. Cervical lymph node biopsy revealed lymphoid follicles with germinal centers. The interfollicular zones were expanded at the expense of lymphoplasmacytic infiltrate with foci of emperipolesis (Figure 1B). Immunostaining showed CD68+, S100+, CD1a-histiocytes, and 100 IgG4+ plasma cells/HPF with an IgG4/IgG ratio of 45%. A bone marrow biopsy showed hypercellularity with 50% CD138+ plasma cells and emperipolesis. A minor salivary gland biopsy revealed periductal lymphoplasmacytic infiltrate with mild non-storiform fibrosis, without fulfilling criteria for focal lymphocytic sialadenitis. Immunostaining showed 12 IgG4+ plasma cells/HPF with an IgG4/IgG ratio of 25%. Even though ANA were positive, the patients did not fulfill criteria for systemic lupus erythematosus or Sjögren's syndrome.

A diagnosis of nodal and extranodal IgG4-associated RRD was made. Treatment with 1 gr boluses of methylprednisolone for 3 days and then prednisone 1 mg/kg/day for 4 weeks was implemented with subsequent prednisone taper. Hydroxychloroquine 200 mg/day was added. After a follow-up of 4.5 years, the patient did not present relapses and was under remission. The patient died of sepsis unrelated to RDD in June 2022.

### Main Points

- Rosai–Dorfman disease may display histopathological features of immunoglobulin G4 (IgG4)-related disease.
- Both conditions may mimic each other.
- Rheumatologists are often involved in the work-up of Rosai–Dorfman disease patients.
- The evidence does not support a link between IgG4-associated Rosai–Dorfman disease and IgG4-related disease.

### Patient 2

A 23-year-old man presented to the Rheumatology clinic with a 1-year history of an excised right mandibular tumor. The tumor appeared at the right mandibular angle and had grown to a diameter of 2 inches within a month (Figure 2A). A magnetic resonance imaging (MRI) of the face showed an increase in the size of the subcutaneous tissue overlying the superficial lobe of the right parotid gland (Figure 2B). An 18-fludeoxyglucose ($^{18}$FDG) PET/CT revealed $^{18}$FDG uptake in a lymph node at the right pectoralis muscle. A biopsy of the lymph node demonstrated follicular and sinus lymphoid hyperplasia, hemophagocytosis and emperipolesis, and 50 IgG4+ plasma cells/HPF. Excision of the tumor was performed, and a diagnosis of IgG4-related sialadenitis was made. He was referred to our Institution for further evaluation.

Physical exam and laboratory tests were unremarkable, including normal CRP, ESR, C3, C4, IgE, IgG, and IgG4. A revision of both biopsies (Figure 2C–F) revealed subcutaneous tissue and lymph nodes with a dense lymphoplasmacytic infiltrate and CD68+, S100+, CD163+, CD1a- histiocytes exhibiting emperipolesis; the mandibular lesion had 90 IgG4+ and 100 IgG+ plasma cells/HPF with an IgG4/IgG ratio of 90% and storiform fibrosis. The lymph node had 50 IgG4+ and 20 IgG+ plasma cells/HPF with an IgG4/IgG >100%.

A diagnosis of IgG4-associated nodal and extranodal RDD was made, and he was referred to the Hematology Department. No treatment was deemed necessary. After a follow-up of 3 years, the patient remained asymptomatic without treatment, and he has not relapsed.

### Patient 3

A 40-year-old woman had an 8-year history of bone tumors at the right tibia and talus that have required multiple orthopedic interventions and had been diagnosed as plasmacytoma. She also had of a left maxillary sinus tumor that was diagnosed as angiocentric eosinophilic fibrosis. During the evaluation, an asymptomatic left extra-axial temporal lesion was seen on CT images and was referred to our Institution in 2015.

The physical exam was unremarkable. CRP was elevated, and there was polyclonal hypergammaglobulinemia. A paranasal sinuses CT revealed complete obliteration of the left

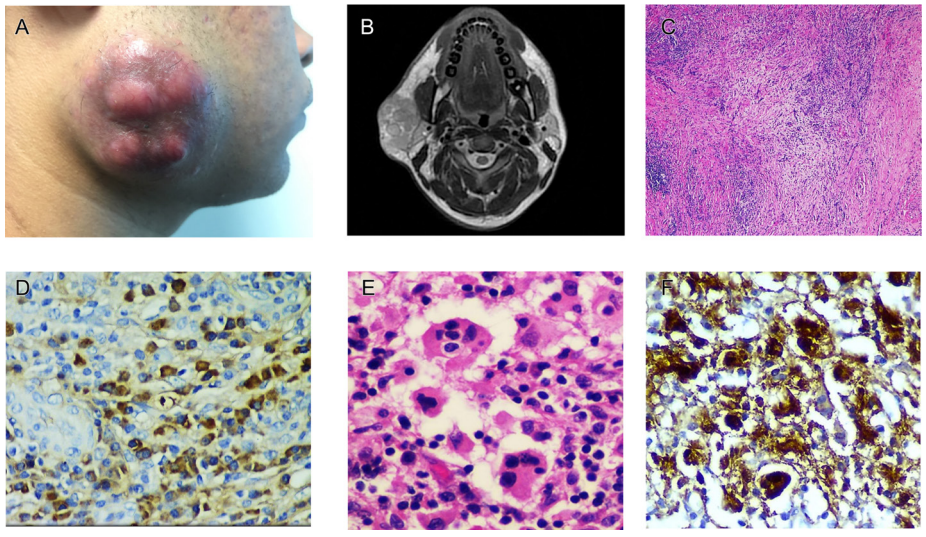


**Figure 2.** Clinical, radiological, and histopathological findings in case 2. (A) Tumor-like lesion at the right mandibular angle with erythema and telangiectasias. (B) T2-weighted head MRI showing an increase in the size of the subcutaneous tissue overlying the superficial lobe of the right parotid gland, due to the presence of multiple nodular hypointense lesions. (C) Biopsy of the mandibular lesion showing storiform fibrosis and abundant mature plasma cell infiltration (H&E 10×). (D) Immunohistochemistry for IgG4 (40×) of the mandibular tumor with the presence of plasma cells of mature morphology positive for IgG4 (90/HPF). (E) Hematoxylin and eosin section (100×) with histiocytes with extensive eosinophilic cytoplasm with intracytoplasmic lymphocytes (emperipolesis). These histiocytes stand out with positive immunohistochemistry for protein s100 (F).

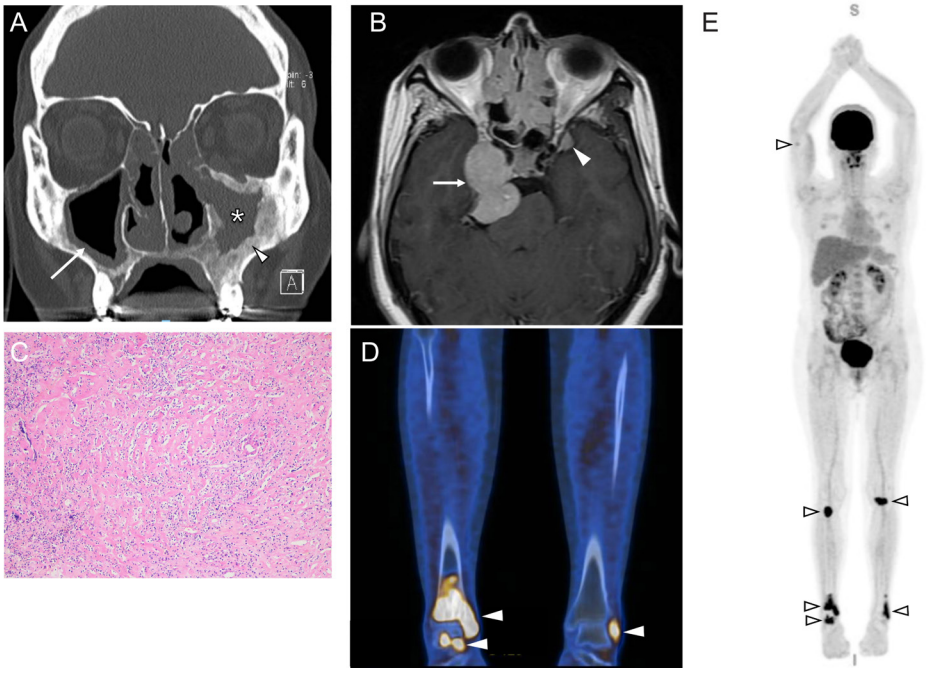


**Figure 3.** Radiological and histopathological findings in case 3. (A) Coronal paranasal sinuses CT showing thickening of the right maxillary sinus mucosa (arrow) and complete obliteration of the left maxillary sinus by a soft tissue mass (asterisk) with findings suggestive of severe osteitis (arrowhead). (B) Axial T1-weighted contrast-enhanced brain MRI showing extra-axial mass at the right cerebellopontine angle extending into the ipsilateral trigeminal tract and with infiltration and expansion of the Meckel's cavum and ipsilateral cavernous sinus (arrow); a similar lesion adjacent to the left sphenoid cleft molding the temporal lobe was seen (arrowhead). (C) Dural lesion biopsy showing storiform fibrosis and abundant mature plasma cell infiltration (H&E 10×). (D and E) $^{18}$FDG PET/CT revealed $^{18}$FDG uptake at the right proximal and distal tibial epiphyses, left proximal tibial epiphysis, left distal fibular epiphysis, right talus, and right humerus with a SUVmax of up to 11.8.

maxillary sinus by a soft tissue mass (Figure 3A). An MRI of the brain revealed an extra-axial mass in the right cerebellopontine angle and a similar lesion adjacent to the left sphenoid cleft (Figure 3B).

In May 2016, the right temporal mass was completely excised. The histopathological report showed dense lymphoplasmacytic infiltrates, storiform fibrosis (Figure 3C), CD68+ and S100+ histiocytes showing emperipolesis; CD138+ plasma cells showed no light chain restriction. Immunostaining showed 44 IgG4+ and 246+ plasma cells/HPF with an IgG4/IgG ratio of 18%. A diagnosis of extranodal IgG4-associated RDD was made.

At the Hematology clinic, the bone tumors were considered a manifestation of RDD, and she was managed only with local radiotherapy due to bone pain. In 2021, she presented with nasal congestion. A review of the previous maxillary sinus tumor biopsy revealed findings consistent with RDD. An $^{18}$FDG PET/CT revealed $^{18}$FDG uptake at multiple bones (Figure 3D and E). In September 2021, she complained of bone pain, and a left retroauricular lymph node was noticed at the physical exam. At that time, she was diagnosed with disseminated RDD, and systemic glucocorticoid treatment was indicated with prednisone at a dose of 60 mg/day. Response to bone pain was observed, and prednisone was tapered over the course of 6 months until discontinuation. At the last follow-up visit, 1 year after the start of prednisone, bone pain was under control and an $^{18}$FDG PET/CT showed diminished metabolic activity of the bone lesions.

Ethical committee approval was received from the Institutional Review Board of Instituto Nacional de Ciencias Médicas y Nutrición Salvador Zubirán (IRE-2549). Written informed consent was obtained from the patients. This research was conducted in compliance with the Helsinki Declaration.

## Literature Review

Rosai–Dorfman disease shares clinical and histopathological features with IgG4-RD. Both conditions may involve a similar array of organs, such as lymph nodes, major salivary gland, orbit, pachymeninges, and pancreas.[1,5] Furthermore, RDD shares morphological features with IgG4-RD such as the presence of dense lymphoplasmacytic infiltrates, storiform fibrosis, and IgG4+ plasma cells.[2,3,6,7] As such, both conditions may mimic each other, and rheumatologists are often involved in the work-up of RRD patients.[5] Similarities and differences between RDD and IgG4-RD are summarized in Table 1.

Several studies have investigated the relationship between RDD and IgG4-RD. Kuo et al[6] first proposed a possible association between both conditions. They evaluated 12 cases of cutaneous RDD and found >30 IgG4+ plasma cells/HPF in all cases except one and an increased IgG4/IgG ratio in 3 patients. Furthermore, some cases exhibited the characteristic storiform fibrosis of IgG4-RD. They concluded that cutaneous RDD might belong to the spectrum of IgG4-RD.

Later, Shrestha et al found that 6 out of 8 RDD cases had increased IgG4+ plasma cells and 3 had an elevated IgG4/IgG ratio.[16] In a similar vein, Zhang et al studied 26 RDD biopsies from 15 patients and found that 73% of RDD cases had more than 10 IgG4+ plasma cells/HPF, 46% of the cases had more than 30 IgG4+ plasma cells/HPF, and 30.8% an IgG4/IgG ratio over 40%.[13] In a similar study including 70 cases of nodal and extranodal RDD, 40% demonstrated elevated IgG4+ plasma cells and 17.4% an IgG4/IgG ratio over 40%.[14] Another study by Liu et al assessed the number of IgG4+ plasma cells and the IgG4/IgG ratio in 32 biopsy specimens from 29 patients with RDD and compared the findings from those of IgG4-RD of the pancreas and reactive lymph nodes. They also assessed the number of FoxP3+ regulatory T cells (Tregs) since they were reported to be increased in IgG4-RD. They found that RDD cases had much lower numbers of IgG4+ plasma cells and lower IgG4/IgG ratios compared with IgG4-RD but were similar to reactive lymph nodes. Three of the 29 cases had an IgG4/IgG ratio >40%. Furthermore, RDD had a lower number of FoxP3+ Tregs than IgG4-RD. There were no significant differences in the number of IgG4+ plasma cells and the IgG4/IgG ratio between the nodal and extranodal RDD cases.[15]

Recent reports have corroborated the aforementioned findings, including nodal and extranodal RRD patients with storiform fibrosis, obliterative phlebitis, and even high IgG4 serum levels, which complicates the differential diagnosis between both conditions.[3,12,17,18]

**Table 1.** Clinical, Serological, and Pathological Features in IgG4-Related Disease and Rosai–Dorfman Disease

| | IgG4-Related Disease | Rosai–Dorfman Disease | References |
|---|---|---|---|
| Age of onset | 48-60 years | 20.6 years | 2, 8, 9 |
| Male to female ratio | 1:1-4:1 | 1.4:1 | 2, 8, 9 |
| Lymphadenopathy | 44% | Almost always present | 8, 9, 10 |
| Massive cervical lymphadenopathy | Rare | Common | 8, 10 |
| Orbit | 6.5% | 11% | 2, 11 |
| Pachymeninges | 2.7% | 5% | 2, 11 |
| Lacrimal gland | 26% | Limited to case reports | 8, 10 |
| Major salivary gland | 37.7% | Limited to case reports | 8, 10 |
| Paranasal sinus | 6.5% | 11% | 9, 10 |
| Pancreato-hepatobiliary | 47.7% | Limited to case reports | 8, 10, 11 |
| Lung | 14.2% | 2% | 10 |
| Retroperitoneum | 15.8% | Uncommon | 8, 10 |
| Kidney | 15.6% | 4% | 8, 10 |
| Skin | Rare | 10% | 2, 9, 10 |
| Fever | Rare | Common | 10, 11 |
| Bone involvement | 0% | 5-10% | 10, 11 |
| High IgG4 serum levels | 78.7% | Rare | 9, 12 |
| Hypocomplementemia | 29.8% | Not present | 9 |
| Dense lymphoplasmacytic infiltrates | Almost always present | Common | 2, 11 |
| Storiform fibrosis | 39.6% | Uncommon | 8, 10 |
| Obliterative phlebitis | 18.5% | Not present | 11 |
| > 10 IgG4+ plasma cells/HPF | Almost always present | 27-73% | 11, 13-15 |
| IgG4/IgG ratio >40% | Almost always present | 10-30% | 11, 13-15 |
| Emperipolesis | Not present | Almost always present | 2, 11 |
| Histiocytic infiltration with positive S100, CD68, CD14, and CD163 immunostaining | Not present | Always present | 2, 11 |

HPF, IgG4, immunoglobulin G4; HPF, high-power field.

Our cases showed varying degrees of IgG4+ plasma cell infiltration, all with more than 10 IgG4+ plasma cell/HPF and 2 of them with an IgG4/IgG ratio >40%. Of note, the only patient with a ratio <40% displayed storiform fibrosis. These histopathological findings raised concern about the possibility of concomitant IgG4-RD. However, the diagnosis of IgG4-RD requires a strict correlation of clinical, serologic, and histologic features, and the presence of IgG4+ plasma cells is not sufficient to make a diagnosis as many inflammatory and neoplastic conditions may exhibit these findings.[7,19] As such, our cases could be regarded as IgG4-associated RDD rather than an overlapping condition.[4]

In cases where the histopathologic features suggest IgG4-RD, features that point toward RDD are the presence of large histiocytic cells with smooth contoured hypochromatic nuclei, small distinct centrally placed round nucleoli and ill-defined, pale cytoplasm, and emperipolesis.[2,5,19] With immunostaining, those histiocytes are positive for S100, CD68, CD14, and CD163.[2-10] These features may also help distinguished Sjögren's syndrome from RRD of the salivary glands, as the former lacks histiocytic infiltrates in tissue.[20]

Regarding management of RRD, there is no uniform approach, and treatment is tailored according to the clinical scenario of each patient. As such, the notions of first-line and second-line treatments are not readily applicable to RDD.[10] Observation may be an option in cases with uncomplicated lymphadenopathy or asymptomatic cutaneous RDD. Surgery is warranted for debulking in upper airway obstruction, meningeal involvement with compression or any other bulky lesion causing end-organ damage. Furthermore, surgery may be curative for unifocal disease.[1,10] Systemic therapy with corticosteroids and/or sirolimus is used for multifocal or unresectable disease. Other treatment options used for refractory or relapsing disease are cladribine, methotrexate, vinblastine, thalidomide, lenalidomide, rituximab, or imatinib mesylate.[10]

## Conclusion

In summary, a significant number of RDD shows histopathological features characteristic of IgG4-RD. Although the most recent classification of histiocytoses recommends evaluating IgG4+ plasma cell infiltration and IgG4/IgG ratio in all RDD cases, the usefulness of this approach is currently unanswered, and no further guidelines are provided on how these results should be interpreted.[2,4] At present, the evidence does not support a link between IgG4-associated RDD and IgG4-RD, and one condition should not be considered part of the spectrum of the other.[2,4,19] We believe it is of paramount importance to increase the awareness of IgG4-associated RDD for pathologists who interpret the biopsies and clinicians who integrate the diagnosis and treat such patients to not overdiagnose IgG4-RD. Further studies are needed to address if IgG4-associated RDD behaves differently in terms of treatment response and prognosis compared to RDD without associated IgG4.

**Data Availability:** The data that support the findings of this study are available from the corresponding author upon reasonable request. All data relevant to the study are included in the article.

**Ethics Committee Approval:** Ethical committee approval was received from the Institutional Review Board of Instituto Nacional de Ciencias Médicas y Nutrición Salvador Zubirán (IRE-2549).

**Informed Consent:** Written informed consent was obtained from the patients who participated in this study.

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