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CASE REPORT

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Symmetrical polyarthritis in IgG4-related sialadenitis: a diagnostic challenge with seronegative rheumatoid arthritis

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Abstract

Background IgG4-related disease (IgG4-RD) is a systemic fibro-inflammatory condition characterized by elevated IgG4 serum levels and tissue infiltration by IgG4-positive plasma cells. While often presenting with organ-specific involvement, such as sialadenitis or pancreatitis, its rheumatologic manifestations are rare and poorly understood. IgG4-RD often overlaps with autoimmune diseases such as rheumatoid arthritis (RA), posing a diagnostic challenge, particularly in seronegative presentations.

Case presentation We report a 48-year-old male presenting with progressive symmetrical polyarthritis mimicking rheumatoid arthritis. Laboratory findings showed elevated serum IgG4 levels and inflammatory markers, while autoantibodies (RF, ACPA, ANA, ANCA) were negative. A biopsy of a submandibular gland revealed dense lymphoplasmacytic infiltrates + fibrosis and IgG4-positive plasma cells, confirming the diagnosis of IgG4-RD. The patient responded well to a combination of glucocorticoids and methotrexate, with complete symptom resolution within one month and normalization of inflammatory markers.The therapeutic response observed in this case demonstrates the effectiveness of immunosuppression therapy in IgG4-RD management, while emphasizing the need for long-term follow-up.

Discussion This case underscores the diagnostic challenges in recognizing arthritis in a patient with biopsy-confirmed IgG4-related sialadenitis particularly when seronegative rheumatoid arthritis (RA) remains a plausible differential diagnosis.The overlapping clinical features and shared treatment responses make it challenging to attribute the arthritis to a single etiology.This report emphasizes the importance of considering IgG4-RD in the differential diagnosis of atypical arthritis presentations, particularly in patients with systemic manifestations. Histopathological confirmation, supported by clinical and serological evaluation, remains pivotal in guiding diagnosis and management. Long-term follow-up is essential to monitor for evolving features, including the potential development of overlapping conditions, and to ensure optimal treatment outcomes. Early recognition and tailored interventions are critical to preventing complications and improving patient quality of life.

Conclusion IgG4-related disease (IgG4-RD) must be considered in the differential diagnosis of seronegative arthritis, especially when systemic symptoms or organ involvement are present. This case underscores the growing recognition of IgG4-RD in rheumatologic practice and the importance of a multidisciplinary approach to

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diagnosis and management. Clinicians should maintain heightened awareness of the overlap between IgG4-RD and rheumatoid arthritis (RA), advocating for the integration of histopathology, imaging, and serological testing to ensure accurate diagnosis. Additionally, long-term follow-up is essential to monitor disease progression, recurrence, development of new symptoms, and treatment response, ultimately enhancing patient care and outcomes.

Keywords Seronegative arthritis, Polyarthritis, IgG4-related diseases, IgG4-RD, Submandibular gland, RA

Introduction

IgG4-related disease (IgG4-RD) is a systemic fibroinflammatory condition characterized by elevated serum IgG4 levels and infiltration of IgG4-positive plasma cells in affected tissues. This condition involves dysregulated immune responses, driven by Th2 cells and regulatory T cells, leading to chronic inflammation and fibrosis. While the condition often presents with classic organ involvement such as autoimmune pancreatitis or sialadenitis, rheumatologic manifestations, including polyarthritis or myopathy, are exceedingly rare and less well-documented. Although IgG4-RD can overlap with immune conditions like seronegative RA, its rheumatologic manifestations, such as polyarthritis, remain rare and underrecognized. Both conditions may present with inflammatory arthritis, systematic symptoms, and respond to immunosuppressive therapies, making differentiation challenging without biopsy or specific serologic markers. This case emphasizes the rarity of IgG4-RD presenting in a patient with predominant rheumatologic symptoms and diagnostic challenges it poses in clinical practice.

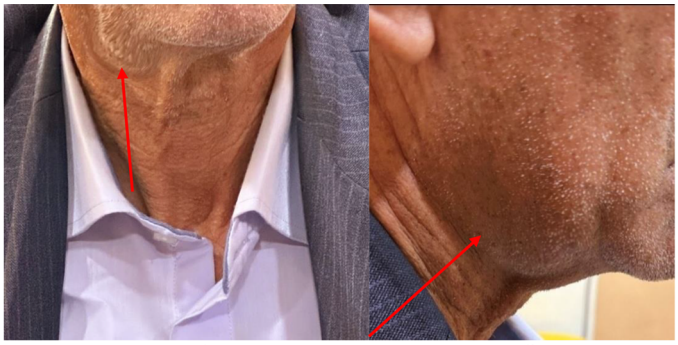
Case presentation

A 48-year-old male presented to the outpatient rheumatology clinic with a six-month history of progressive joint pain, stiffness, and fatigue. These symptoms were symmetrical and primarily involved the small joints of the hands, wrists, and knees, closely mimicking rheumatoid arthritis (RA). The patient denied systemic symptoms such as fever, chronic muscle pain, loss of appetite, weight loss, photosensitivity, or Raynaud's phenomenon, and his medical history was unremarkable, with no prior autoimmune disease or family history of rheumatologic conditions.

Upon physical examination, the patient exhibited swollen and tender metacarpophalangeal (MCP) and proximal interphalangeal (PIP) joints bilaterally, accompanied by mild synovitis in the wrists and knees. Despite the inflammation, no joint deformities, rashes, or nail changes were observed, and tests for Schirmer's sign and oral ulcers yielded negative results. The initial laboratory findings revealed negative rheumatoid factor (RF), anti-citrullinated peptide antibody (ACPA), antineutrophil cytoplasmic antibodies (ANCA), anti-Jo-1, and antinuclear antibodies (ANA). However, the patient's erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) were elevated at 58 mm/hr and 18 mg/L, respectively, indicating an inflammatory process.

While the illness closely resembled RA, the absence of joint erosions and seronegative status made a definitive RA diagnosis less likely. This conclusion was drawn from negative results for rheumatoid factor (RF) and anti-citrullinated protein antibodies (ACPA). Additionally, X-rays of the hands and feet showed no erosions or joint damage typical of rheumatoid arthritis (RA), and musculoskeletal ultrasound revealed an absence of erosions or power Doppler activity. There were also no typical joint deformities associated with RA. Despite the fact that the absence of joint erosions reduced the likelihood of RA, early or seronegative RA could not be ruled out, given the overlapping clinical features and early presentation.

Given the seronegative profile, further evaluation was performed. Serum IgG4 levels were elevated at 3.8 g/L (normal range: 0.08–1.4 g/L), while total IgG levels were 19.4 g/L (normal range: 7.0–16.0 g/L). IgM levels were measured at 1.2 g/L (normal range: 0.4–2.3 g/L), and IgE levels were 426 IU/ml (normal range: <150 IU/ml). Complement levels (C3 and C4) and HLA-B27 testing were normal. Furthermore, the patient also presented with a painless swelling in the right submandibular gland. This swelling was notable for being non-tender to the touch and was easily palpable (Fig. 1a & b). A contrast-enhanced CT scan of the neck and thorax demonstrated significant right-sided submandibular gland swelling without lymphadenopathy or contralateral gland involvement. A fine needle aspiration biopsy of a palpable submandibular gland revealed dense lymphoplasmacytic infiltrates & fibrosis, demonstrating more than 30 IgG4-positive plasma cells per high-power field (hpf), confirming the diagnosis of IgG4-related disease (IgG4-RD). This diagnosis was substantiated using the 2020 Revised Comprehensive Diagnostic Criteria (RCD) for IgG4-RD. Histopathological examination demonstrated dense lymphoplasmacytic infiltration + fibrosis; however, the observed fibrosis did not conform to typical obliterative or storiform patterns, and obliterative phlebitis was absent. The tissue exhibited an IgG4+/IgG + ratio of 42.9%. No evidence of necrotizing vasculitis, eosinophilic infiltration, or neutrophilic infiltration was observed. The 2020 Revised Comprehensive Diagnostic (RCD) criteria for IgG4-related disease (IgG4-RD) is outlined below in Fig. 1(c). Additionally, a thorough evaluation of the chest and abdominal organs on CT confirmed the absence of



(a) *(b)*

[Item 1] Clinical and radiological features ✓
One or more organs show diffuse or localized swelling or a mass or nodule characteristic of IgG4-RD. In single organ involvement, lymph node swelling is omitted.
[Item 2] Serological diagnosis ✓
Serum IgG4 levels greater than 135 mg/dl.
[Item 3] Pathological diagnosis ✓
Positivity for two of the following three criteria:
① Dense lymphocyte and plasma cell infiltration with fibrosis.
② Ratio of IgG4-positive plasma cells /IgG-positive cells greater than 40% and the number of IgG4-positive plasma cells greater than 10 per high powered field
③Typical tissue fibrosis, particularly storiform fibrosis, or obliterative phlebitis
Diagnosis (items):
Definite: 1) +2) +3) ✓
Probable: 1) +3)
Possible: 1) +2)

(c)

Fig. 1 (**a**, **b**) Two images of the patient are displayed, highlighting the palpable swelling of the right submandibular gland, as indicated by the arrows. (**c**) 2020 Revised Comprehensive Diagnostic (RCD) Criteria for IgG4-RD

systemic involvement, differentiating localized IgG4-RD from a systemic disease process.

The criteria/items met in this case are highlighted with blue ticks, indicating their fulfillment according to the diagnostic criteria framework. For this patient, the diagnosis is definitive, as items 1,2 and 3 (with subcomponents 1 & 2) are fully met.

Alternative diagnoses, including infections (e.g., tuberculosis, hepatitis B or C), malignancies, ANCA associated vasculitis and other autoimmune conditions such as Sjögren's syndrome, were systematically ruled out. The exclusion of Sjögren's syndrome was confirmed through negative results for anti-Ro/SSA and anti-La/SSB antibodies, a normal Schirmer's test, and the absence of xerostomia or keratoconjunctivitis sicca. Furthermore, ANCA-associated vasculitis was excluded based on negative ANCA testing and the absence of nonspecific symptoms, such as fever, weight loss, hypertension, kidney-related symptoms, or skin rashes. Additionally, there were no systemic vasculitis symptoms or evidence of necrotizing vasculitis in the biopsy findings.

The patient was started on a tapering dose of oral prednisone (initial dose: 40 mg daily), which led to rapid symptom resolution. Methotrexate (15 mg weekly) was added as a steroid-sparing agent. At six months follow-up, the patient remained asymptomatic with normalized inflammatory markers and stabilized IgG4 levels.

Clinical timeline

The clinical course and timeline are illustrated visually in Fig. 2 below.

Discussion

This case underscores the diagnostic challenges of IgG4-RD when it manifests with predominant rheumatologic features such as symmetrical polyarthritis. The seronegative status, absence of joint erosions, and biopsy-confirmed IgG4-related sialadenitis complicate differentiation from other autoimmune conditions, particularly seronegative rheumatoid arthritis. Given the overlapping clinical and therapeutic features, recognizing IgG4-RD as a potential mimic of autoimmune arthritis is essential for accurate diagnosis and appropriate management.

IgG4-RD is an under-recognized condition in rheumatology that overlaps with several autoimmune diseases, including rheumatoid arthritis (RA), Sjögren's syndrome, and systemic lupus erythematosus (SLE) [1, 2]. Rheumatologic manifestations of IgG4-RD, including polyarthritis, are rare, with arthritis reported in approximately 10% of the cases, which underscores the significance of this report [3].

It should be noted that reports describing inflammatory arthritis as a primary manifestation of IgG4-related disease (IgG4-RD) are limited. However, IgG4-related disease (IgG4-RD) can be a rare but significant cause of inflammatory arthritis and should be also considered in cases of aggressive inflammatory arthritis. While it is primarily known for causing fibroinflammatory lesions affecting multiple organ systems, its potential to manifest as inflammatory arthritis is much less recognized. This condition can present with various clinical features, including joint involvement, and may often be misdiagnosed due to its overlap with other rheumatic diseases. Therefore, a high index of suspicion is essential for accurate diagnosis and appropriate management [4].

Seronegative arthritis encompasses a variety of conditions that can present with similar clinical features, making diagnosis challenging. A comprehensive understanding of these conditions is crucial for appropriate management. Below, we provide a detailed tabular summary comparing key features of several relevant conditions, including rheumatoid arthritis, Sjögren's syndrome, sarcoidosis, infectious arthritis, and IgG4-related disease. Additionally, the rationale for excluding each alternative diagnosis based on clinical presentation and laboratory findings is also mentioned in the table. The differential diagnoses and rationale for exclusion for this patient are illustrated in Fig. 3.

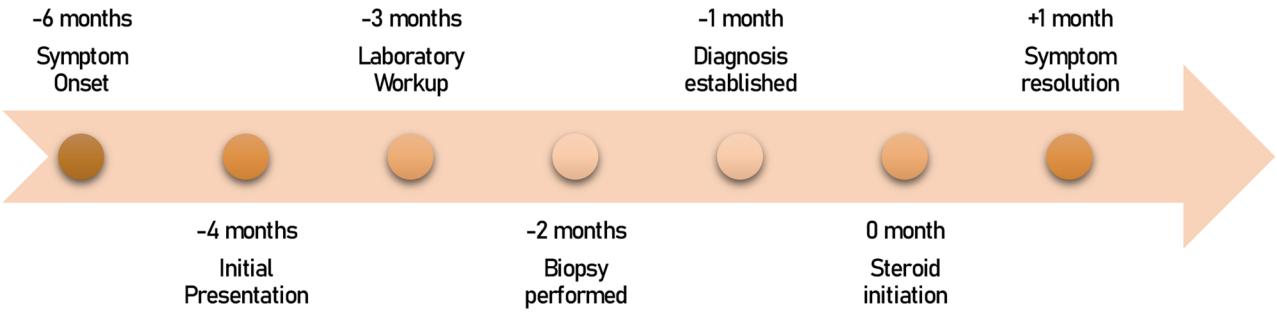


Fig. 2 A visual representation of the patient's clinical course, from the onset of symptoms to symptom resolution after treatment. The timeline highlights key milestones, including symptom onset, diagnostic evaluations (laboratory workup and biopsy), diagnosis confirmation, treatment initiation, and follow-up outcomes

Condition	Key Features	Rationale for Exclusion
Rheumatoid Arthritis (RA)	➢ Symmetrical polyarthritis ➢ Morning stiffness lasting >30 minutes ➢ Joint deformities (e.g., ulnar deviation) ➢ Extra-articular manifestations (e.g., nodules, vasculitis)	RA is typically associated with positive rheumatoid factor (RF) and anti-citrullinated protein antibodies (ACPA). In cases of seronegative RA, the absence of these markers may suggest a different underlying condition or a variant of RA that lacks classic features.
Sjögren's Syndrome	➢ Dry eyes (keratoconjunctivitis sicca) ➢ Dry mouth (xerostomia) ➢ Fatigue and malaise ➢ Positive antinuclear antibodies (ANA) or RF ➢ Lymphocytic infiltration in salivary glands	While joint pain can occur in Sjögren's syndrome, it is usually accompanied by significant dryness symptoms. Furthermore, the presence of specific autoantibodies (such as anti-SSA/Ro and anti-SSB/La) often helps differentiate it from other forms of arthritis. Seronegative cases may not exhibit the systemic involvement characteristic of this syndrome.
Sarcoidosis	➢ Granulomatous inflammation affecting multiple organs ➢ Respiratory symptoms (e.g., cough, dyspnea) ➢ Skin lesions (e.g., erythema nodosum) ➢ Elevated serum angiotensinconverting enzyme (ACE) levels ➢ Lymphadenopathy	Sarcoidosis typically presents with extra-articular manifestations that are not seen in isolated seronegative arthritis. The presence of granulomas on biopsy and elevated ACE levels can help confirm the diagnosis. Additionally, respiratory symptoms are often prominent in sarcoidosis patients.
Infectious Arthritis	➢ Acute onset of joint pain and swelling ➢ Fever and systemic signs of infection ➢ Localized tenderness over affected joints ➢ Joint aspiration revealing pathogens or inflammatory markers	Infectious arthritis usually presents acutely with systemic signs such as fever and chills. Joint aspiration typically reveals bacteria or other pathogens, which would not be present in non-infectious forms of arthritis. The rapid onset and systemic involvement are key differentiators from seronegative arthritis.
IgG4-Related Disease	➢ Organ involvement beyond joints (e.g., pancreas, salivary glands) ➢ Elevated serum IgG4 levels ➢ Fibrosis and lymphoplasmacytic infiltrate on biopsy ➢ Often presents with mass-like lesions in affected organs	IgG4-related disease is characterized by specific organ involvement and elevated serum IgG4 concentrations. The lack of typical inflammatory arthritis features—such as symmetrical joint involvement and significant morning stiffness—helps to exclude this condition from the differential diagnosis when assessing seronegative arthritis.

Fig. 3 Differential diagnosis chart & rationale for excluding other diagnosis. A tabular summary of the differential diagnosis for seronegative arthritis in this patient, comparing key features of conditions such as rheumatoid arthritis, Sjögren's syndrome, sarcoidosis, infectious arthritis, and IgG4-related disease. The rationale for excluding each alternative diagnosis is provided

Diagnosing IgG4-related disease (IgG4-RD) can be particularly challenging due to its nonspecific symptoms and the overlap it shares with other systemic diseases. Patients often present with vague complaints that can easily be misattributed to other conditions, complicating early recognition. This is concerning because untreated IgG4-RD can lead to irreversible fibrosis or significant organ damage, which may severely affect a patient's quality of life. Therefore, timely diagnosis and intervention are crucial to prevent these complications and ensure effective management of the disease [5]. IgG4-RD is primarily driven by Th2 and regulatory T-cell (Treg) cytokines, which promote fibro-inflammatory damage and plasma cell infiltration in affected tissues [6, 7]. Histopathological examination remains the gold standard for diagnosing IgG4-RD, with key findings including dense lymphoplasmacytic infiltrates, fibrosis, and IgG4-positive plasma cells. In this case, biopsy findings confirmed IgG4-RD but did not entirely exclude seronegative RA, highlighting the importance of a multidisciplinary diagnostic approach. Serum IgG4 levels, although elevated, lack specificity and must be interpreted alongside histopathology [8]. Nevertheless, In a study conducted by *Culver et al.*, a serum IgG4 level exceeding 2.8 g/L demonstrated a positive predictive value of 44.5% and a negative predictive value of 97.7% for distinguishing between IgG4-related disease (IgG4-RD) and non-Ig4-RD conditions [9].

In this case, the patient's biopsy findings and elevated serum IgG4 levels confirmed the diagnosis. Previous studies have highlighted its overlap with conditions such as RA, Sjögren's syndrome, and SLE [10, 11]. However, in most cases, the disease's glandular or organ involvement often overshadows its joint manifestations, complicating its recognition in rheumatologic practice [12].

The treatment of IgG4-RD involves glucocorticoids as the first-line therapy, which effectively suppress inflammation and resolve symptoms [13]. Although the response to glucocorticoids is generally rapid, tapering these medications is frequently associated with disease relapses. To address this issue, steroid-sparing agents are often considered. Medications such as methotrexate, mycophenolate, azathioprine, and cyclophosphamide have been utilized in these cases [14, 15]. This patient responded rapidly to a tapering regimen of prednisone, and methotrexate was introduced as a steroid-sparing agent. In refractory cases, biologics such as rituximab have shown promise in achieving disease remission [6].

While early or seronegative rheumatoid arthritis (RA) remains a plausible differential diagnosis, the histopathological confirmation of IgG4-related sialadenitis complicates this assessment. Bone erosions are known to develop early in RA, often within weeks of diagnosis, with over 10% of patients experiencing these changes within 8 weeks and up to 60% after one year [16]. Therefore, it is crucial to recognize that the presence of polyarthritis may not be solely attributed to IgG4-RD, as RA could also be a contributing factor.

Furthermore, monitoring patients with IgG4-related disease (IgG4-RD) is essential, regardless of the diagnostic certainty—whether definitive or uncertain. This long-term follow-up is vital for observing potential erosive joint changes or recurrence of IgG4-RD and enhances our understanding of the disease dynamics. Vigilance in recognizing evolving patterns is necessary to navigate the complexities associated with managing these overlapping conditions, ultimately leading to improved patient outcomes [17].

In this case, although the biopsy findings confirmed the diagnosis of IgG4-related disease (IgG4-RD), they did not rule out seronegative rheumatoid arthritis (RA) as a potential contributor to the patient's polyarthritis. The coexistence or overlap of IgG4-RD and seronegative RA is clinically relevant, particularly in cases where systemic inflammation is prominent. This highlights the necessity of considering all potential contributors when interpreting biopsy results. Given that both conditions can present with similar clinical features and may respond to comparable treatments, careful evaluation with long-term follow up is essential to ensure accurate diagnosis and appropriate management.

A tabular summary of the differential diagnosis for seronegative arthritis in this patient, comparing key features of conditions such as rheumatoid arthritis, Sjögren's syndrome, sarcoidosis, infectious arthritis, and IgG4-related disease. The rationale for excluding each alternative diagnosis is provided.

Conclusion

This case underscores the intricate nature of diagnosing symmetrical polyarthritis within the context of IgG4-related disease (IgG4-RD), as the overlapping characteristics with seronegative rheumatoid arthritis (RA) present significant diagnostic challenges. Both conditions share similar inflammatory pathways and responses, necessitating a cautious approach and diligent long-term follow-up. Long-term follow-up, including imaging, serological testing, and clinical evaluations, is essential for monitoring RA-specific features (e.g., erosions or persistent synovitis) and assessing IgG4-RD recurrence. Our findings highlight the critical importance of integrating histopathology, serology, imaging, and clinical expertise in effectively managing atypical presentations of inflammatory arthritis. Awareness of this complex condition among rheumatologists can facilitate timely and targeted interventions, ultimately improving patient outcomes. The treatment outcomes are illustrated in Fig. 4 below.

A graphical illustration showing the patient's response to treatment. The chart tracks the normalization of

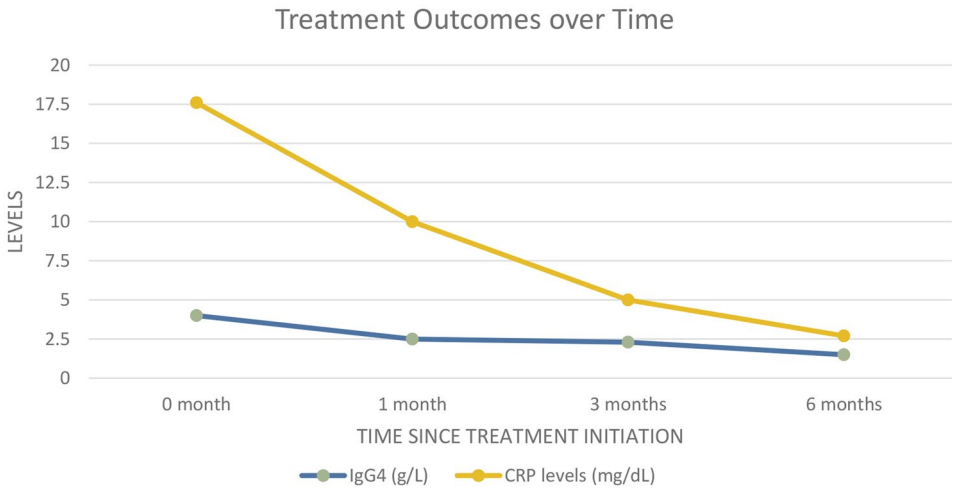


Fig. 4 Treatment outcomes over time. A graphical illustration showing the patient's response to treatment. The chart tracks the normalization of inflammatory markers (C-reactive protein) and serum IgG4 levels over six months following the initiation of glucocorticoids and methotrexate therapy

inflammatory markers (C-reactive protein) and serum IgG4 levels over six months following the initiation of glucocorticoids and methotrexate therapy.

Patient perspective

My journey began with mild joint pain that I initially brushed off as fatigue. However, it quickly escalated into severe stiffness that made even simple tasks like typing or cooking feel nearly impossible. When my joints became swollen, I started to worry that I might have rheumatoid arthritis. After months of testing and uncertainty, I was discovered to have elevated IgG4 levels and confirmed the diagnosis through a biopsy. Starting treatment was nothing short of life-changing. Within just a few weeks, the pain and stiffness that had dominated my daily life began to fade significantly. I felt a renewed sense of hope as I regained control over my life and could finally engage in activities I had once enjoyed.

Abbreviations

IgG4-RD	IgG4-related disease
Anti-Ro/SSA	Anti–Sjögren's-syndrome-related antigen A
Anti-La/SSB	Anti–Sjögren's-syndrome-related antigen B
Anti Jo 1	Anti-histidyl tRNA synthetase
RF	Rheumatoid factor
RA	Rheumatoid arthritis
ACPA	Anti-citrullinated protein/peptide antibodies
ANA	Antinuclear Antibodies
ESR	Erythrocyte sedimentation rate
CRP	C reactive protein
SLE	Systemic lupus erythematosus
Th2	Type 2 helper (T) cells
HLA	Human leukocyte antigens
mm/hr	Millimeters per hour
mg/L	Milligrams per litre

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None to declare.

Author contributions
Faizan Bashir & Moniza Rafiq, conceptualized and drafted the case report. Saide Honarmand conducted the literature review, and finalized the manuscript. Moiza Bashir, Ali Jafer collected data, literature search, edited and proofread the manuscript. All authors read and approved the finalized version of the manuscript.

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Data availability
No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate
This study was conducted in accordance with the principles of the Declaration of Helsinki. NA for consent to participate.

Consent to publish
Written informed consent to publish this case report and any accompanying images or information was obtained from the patient.

Clinical trial number
Not applicable.

Competing interests
The authors declare no competing interests.

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