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Case Report

Rash, Lymphadenopathy, and Nephritis: Is It Always Lupus?

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Abstract:

Background: IgG4-related disease (IgG4-RD) is a multisystem fibroinflammatory condition capable of affecting nearly any organ. Renal involvement, although recognized, remains uncommon and frequently leads to diagnostic uncertainty because of overlap with autoimmune, infectious, and lymphoproliferative disorders.

Case Presentation: A 34-year-old woman presented with a three-year course of intermittent painful purpuric plaques, chronic cough, Raynaud-like digital pallor, and episodic arthralgia. Examination revealed generalized non-tender lymphadenopathy. Laboratory studies showed anemia, eosinophilia, low complement levels, elevated IgG, and renal dysfunction. Autoimmune markers were negative. Imaging identified right-sided bronchiectatic changes and axillary lymphadenopathy. Skin biopsy showed leukocytoclastic vasculitis, while lymph node histology suggested reactive plasma cell-rich changes. A markedly raised serum IgG4 level (>53 g/L) prompted kidney biopsy, which demonstrated storiform fibrosis and abundant IgG4-positive plasma cells, confirming IgG4-related tubulointerstitial nephritis.

Discussion & Conclusion: The combination of renal, cutaneous, pulmonary, and lymph node abnormalities, together with serological and histopathological findings, established the diagnosis of systemic IgG4-RD and illustrated its ability to mimic a broad differential diagnosis.

Prednisolone therapy resulted in clinical and biochemical improvement, including normalization of renal function and improvement in complement levels. The patient remains in remission at 1 year.

Keywords: IgG4-related disease, tubulointerstitial nephritis, hypocomplementemia, lymphadenopathy, vasculitis

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Introduction

IgG4-related disease (IgG4-RD) is a chronic fibroinflammatory disorder characterized by tumefactive lesions, a lymphoplasmacytic infiltrate rich in IgG4-positive plasma cells, and a tendency to involve multiple organs simultaneously [1]. Although the pancreas and salivary glands are more frequently affected, renal manifestations, particularly IgG4-related kidney disease (IgG4-RKD), are increasingly recognized and may lead to progressive renal dysfunction if not promptly treated [4]. The protean nature of IgG4-RD often results in diagnostic confusion, as its clinical picture may mimic rheumatic diseases, chronic infections, or hematologic malignancies [2,10].

This report describes a young woman with multisystem involvement in whom renal biopsy ultimately confirmed IgG4-RKD. The case highlights the diagnostic challenges of IgG4-RD and reinforces the importance of integrating histopathology, serology, and clinical judgment for early recognition [3].

Case Presentation

Patient History

A 34-year-old woman with no prior comorbidities reported recurring purpuric patches that initially appeared on her legs and gradually extended to the thighs and forearms over three years. She also described a persistent cough that worsened in cold seasons, episodic joint pains, and transient pallor of the fingers when exposed to cold water. There were no complaints of fever, chest discomfort, abdominal symptoms, hematuria, or urinary irritation. Her mother had rheumatoid arthritis.

Physical Examination

Vital parameters were stable. Multiple non-tender lymph nodes were palpable in the cervical and axillary chains, the largest being a 4-cm node in the right axilla. The remainder of the systemic examination was unremarkable.

Investigations

Table 1 Laboratory Findings

Parameter	Result
Hemoglobin	9.1 g/dL
Total leukocyte count	10,000/μL
Differential count	P 59%, L 28%, E 11%
Absolute eosinophil count	**990/μL**
Platelet count	3.4×10^5/μL
Urea	41 mg/dL
Creatinine	1.9 mg/dL
Uric acid	7.9 mg/dL
Sodium	132 mEq/L
Potassium	4.6 mEq/L
Calcium	9.76 mg/dL
Phosphorus	3.59 mg/dL
Total protein	**10.34 g/dL**
Albumin	**2.78 g/dL**
CRP	13.39 mg/L
Urine routine	No protein, no RBCs, no pus cells
Urine ACR	<30 mg/g

Table 2 Autoimmune Serology and Immunoglobulin Profile

Category	Test	Result
ANA Profile	ANA (IFA)	Negative
	ANA Extended Profile	Negative
ANCA Profile	c-ANCA	Negative
	p-ANCA	Negative
	MPO-ANCA	Negative
	PR3-ANCA	Negative
Immunoglobulins	**IgG**	**5702 mg/dL**
	IgA	62 mg/dL
	IgM	267 mg/dL
	IgE	75 IU/mL
	IgG4	**>53 g/L**

Table 3 Radiology

Imaging Modality	Key Findings
Chest X-ray	Hyperinflated lung fields; no infiltrates
X-ray PNS	Bilateral maxillary sinusitis
Ultrasound KUB	Kidneys 9.5 × 4.2 cm; increased medullary echogenicity
CECT chest & abdomen	Right middle lobe collapse/consolidation; bronchiectasis in RUL & RLL; interlobular septal thickening; axillary lymphadenopathy largest 4 × 3 cm
MRI abdomen	No significant abnormalities detected

Nerve Conduction Studies

Electrophysiology showed reduced amplitudes in lower limb nerves and absent peroneal F-waves, suggestive of mild neuropathic involvement.

Lymph Node

Excisional biopsy showed expanded interfollicular areas enriched with plasma cells of mixed light-chain expression and prominent high endothelial venules, compatible with reactive lymphadenopathy.

Skin

Biopsy of the purpuric lesion demonstrated leukocytoclastic vasculitis with neutrophils, eosinophils, fibrinoid necrosis, and nuclear debris.

Kidney

Seventeen glomeruli were sampled. The glomeruli appeared minimally altered, but the interstitium contained dense aggregates of plasma cells, eosinophils, and lymphocytes. Storiform fibrosis was evident. More than 10 IgG4-positive plasma cells per high-power field were identified. Immunofluorescence did not show significant immune deposition. These findings confirmed IgG4-related tubulointerstitial nephritis.

Diagnosis

Based on the clinical features, markedly elevated serum IgG4, multi-organ involvement, and kidney biopsy findings, the patient fulfilled the 2019 ACR/EULAR classification criteria for IgG4-RD with a total score of 26, supporting a diagnosis of systemic IgG4-RD with renal involvement.

Treatment

Prednisolone was initiated at 1 mg/kg/day and slowly tapered over six weeks to a maintenance dose of 5 mg daily. Hydroxychloroquine (200 mg/day) was added as adjunctive therapy.

Outcome And Follow-Up

During a year of follow-up:

- The purpuric plaques resolved fully.
- Lymph nodes regressed appreciably.
- Respiratory symptoms improved.
- Complement levels improved (C3 76.3 mg/dL; C4 10.3 mg/dL).
- Serum creatinine decreased from 1.9 mg/dL to 0.9 mg/dL.
- No relapse has been documented to date.

Discussion

The heterogeneous nature of IgG4-RD and its resemblance to several autoimmune and inflammatory diseases make it a recognized diagnostic challenge [1,10]. In this patient, the presence of cutaneous vasculitis together with pulmonary, lymphatic, hematologic, and renal abnormalities warranted consideration of a broad differential diagnosis, including autoimmune conditions, infectious causes, and lymphoproliferative disorders [2,10].

Hypocomplementemia, elevated serum IgG levels, and eosinophilia are recognized laboratory clues in IgG4-RKD and may support clinical suspicion [6]. However, serum IgG4 levels alone are insufficient for diagnosis and must be interpreted alongside tissue findings and overall clinicopathological context [3,10]. Lymph node pathology in IgG4-RD can be heterogeneous and may resemble reactive hyperplasia, making nodal biopsy non-definitive in some cases [10].

In contrast, renal biopsy provides highly specific diagnostic information when IgG4-RKD is suspected. Storiform fibrosis, a plasma cell-rich infiltrate with increased IgG4-positive cells, and the absence of significant immune complex deposition are characteristic histopathological findings, as seen in this case [4-6]. Cutaneous vasculitic lesions have also been described in IgG4-RD, although they remain uncommon [10].

IgG4-RD may affect the respiratory system through airway disease, parenchymal involvement, or bronchiectatic changes that are increasingly recognized on thoracic imaging [7,10]. The patient's imaging profile was compatible with these reported pulmonary manifestations.

Glucocorticoids remain the cornerstone of initial therapy for IgG4-RD, and early treatment is associated with favorable clinical and biochemical responses in many patients [9]. The dramatic improvement observed in our patient, together with sustained remission over one year, underscores the value of early intervention in preventing chronic fibrosis and long-term organ damage.

Conclusion

IgG4-related disease can present with diverse organ involvement and pose significant diagnostic challenges. In this patient, integration of clinical, serological, radiological, and histopathological findings led to the diagnosis of IgG4-related kidney disease. Appropriate corticosteroid therapy achieved sustained remission, highlighting the importance of early diagnosis for optimal outcomes.

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Appendix

Fig 1 Raynaud phenomenon

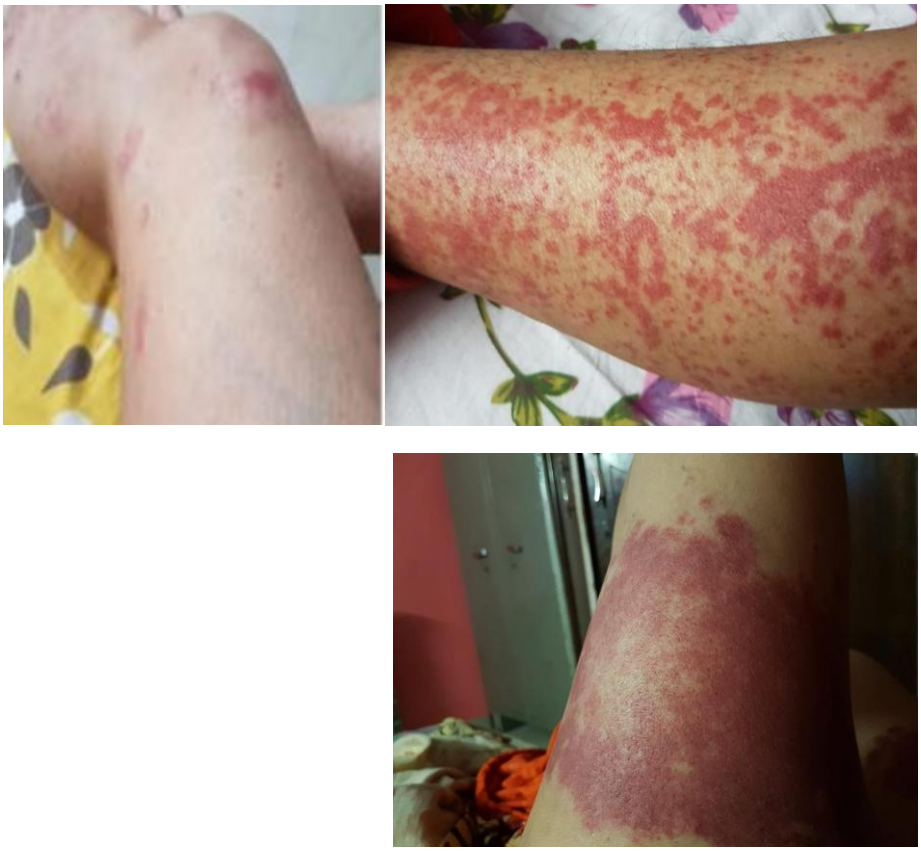

Fig 2, 3, 4 Evolution of skin lesions

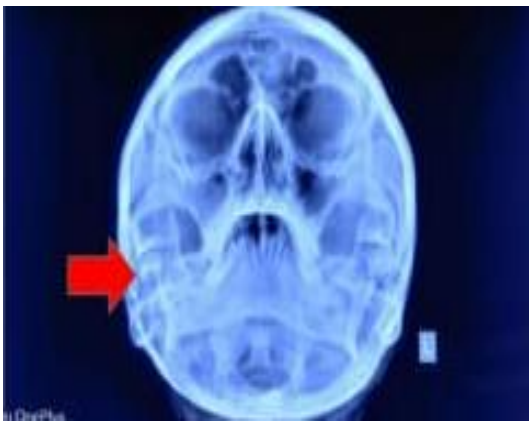

Fig 5 X-Ray PNS showing B/L maxillary sinusitis

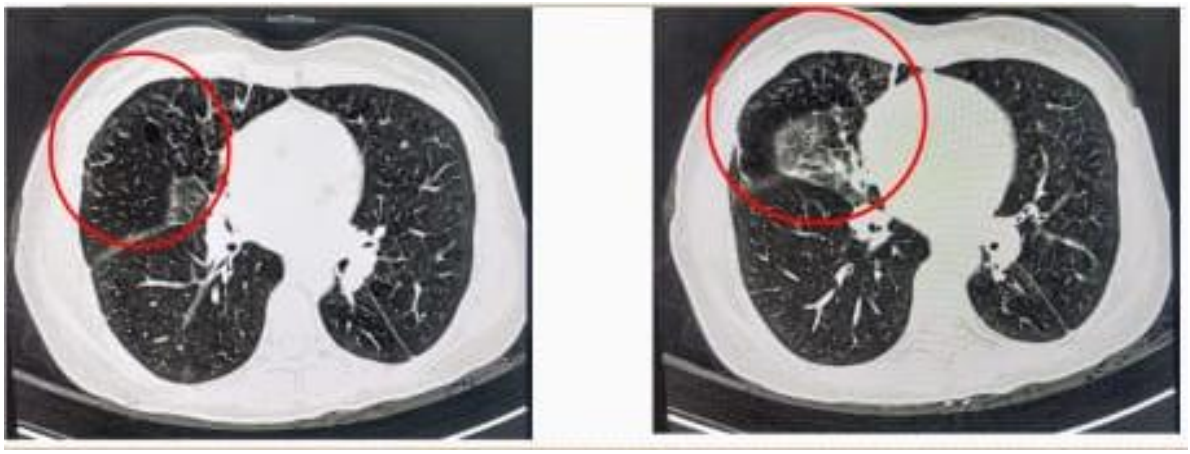

Fig 6 CECT Chest

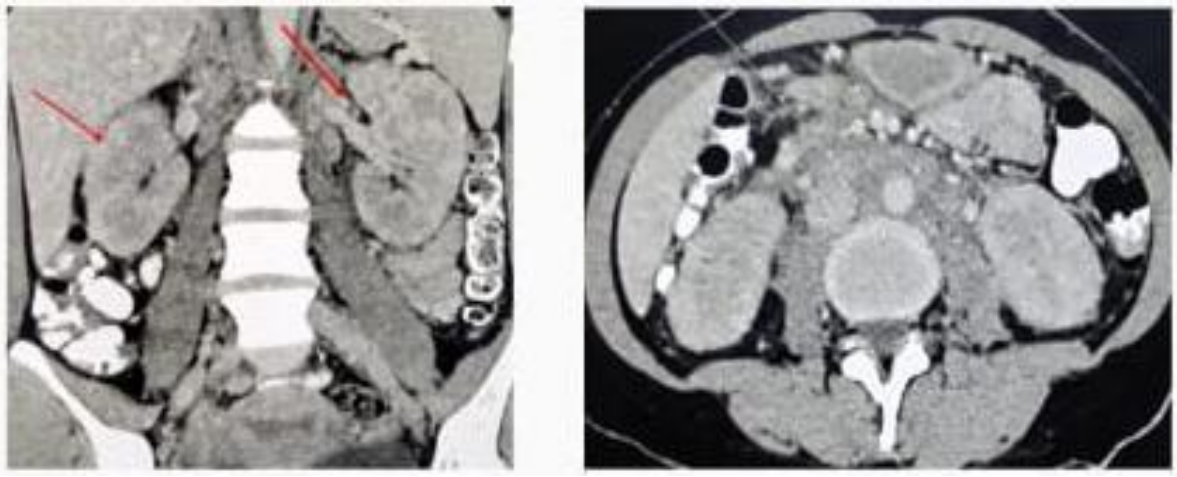

Fig 7 CECT Abdomen

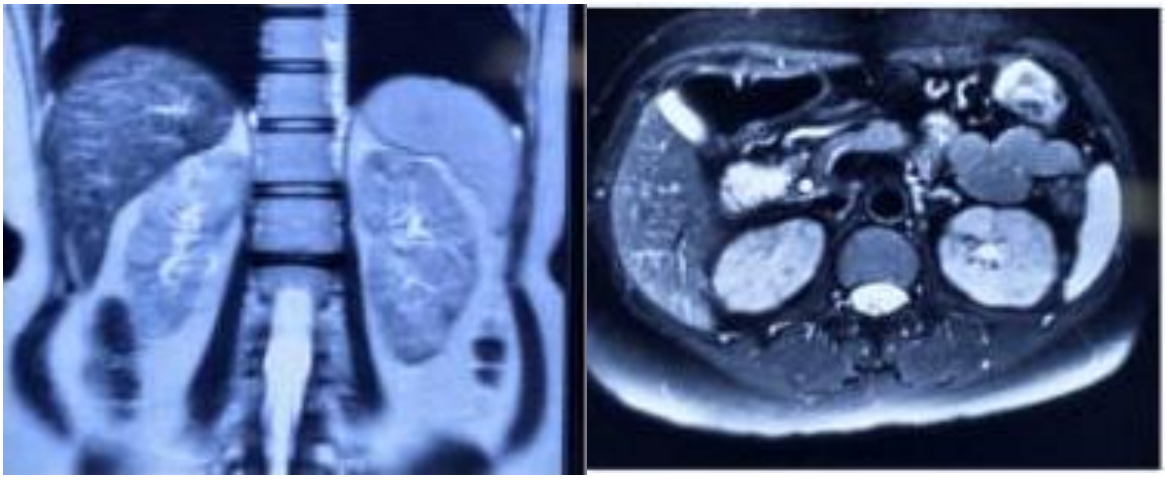

Fig 8 MRI Abdomen

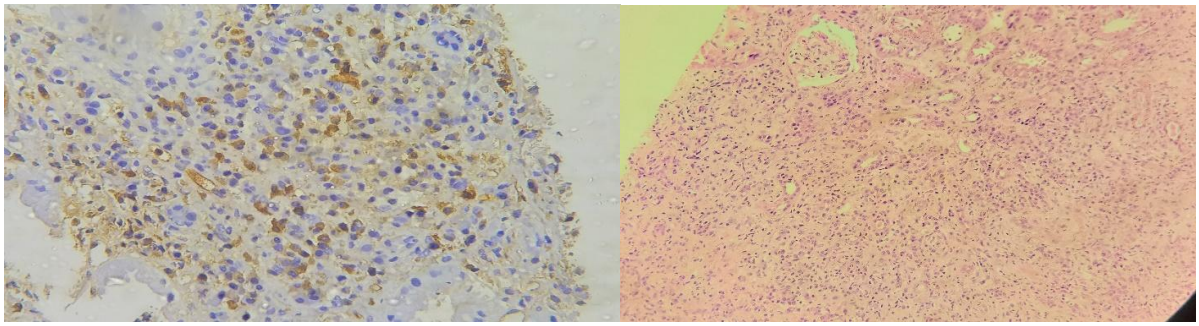

Fig 9 HPE showing plasma cell infiltrates in interstitium with IgG4 positivity