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Nephrotic syndrome as an atypical presentation of polymyositis: A case report.

José Lucas Daza [1]®, Salma Yulieth Yasno López [2], Nicole Stephany Cifuentes Peralta [2], Stefanía Macías Villermo [2], Elba Vanessa Villavicencio Cerón [3].

1. Colombian Association of Nephrology and Arterial Hypertension, Colombia
2. Faculty of Medicine, University of Tolima, Colombia.
3. Nephrology Service, IESS Hospital, Portoviejo, Manabí, Ecuador.

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*** Autor de correspondencia**

Email: José Lucas Daza <drlucasdaza@gmail.com>/Dirección: Cra 12 sur nro 93-21, Clinica Medicaz-Sede Samaria Consultorio 804., Ibagué, Tolima, Colombia. CP 730001.
Teléfono: 318 (278) 1476.

Abstract

Introduction: Polymyositis is an idiopathic autoimmune myopathy characterized by collagen disorders that primarily affect proximal muscles and the lungs. It has a subacute onset and progresses over several weeks. It is more prevalent in women than in men. Renal manifestations are uncommon, but when present, the main manifestations include acute tubular necrosis related to hemoglobinuria and myoglobinuria associated with acute rhabdomyolysis. In addition, symptoms such as dysphagia and dyspnea may occur.

Case report: We report the case of a 40-year-old female patient with a history of dermatitis herpetiformis and hyperprolactinemia who presented with a one-month history of fever, anasarca, muscle weakness, significant proteinuria, and pericardial effusion. Electromyography revealed findings consistent with inflammatory myopathy, and muscle biopsy confirmed the diagnosis of polymyositis. Renal biopsy revealed an increased mesangial matrix with IgM deposits on immunofluorescence, which was consistent with mesangial glomerulopathy. The patient was treated with corticosteroid pulses and had a favorable clinical course.

Discussion: Renal involvement in patients with polymyositis (PM) is uncommon and is usually limited to tubular damage secondary to rhabdomyolysis or glomerulopathies, such as focal segmental glomerulosclerosis or tubulointerstitial nephritis. The development of nephrotic syndrome in this context requires a detailed etiological evaluation. In the present case, renal biopsy revealed IgM mesangial nephropathy, a rare and still controversial entity that has been discussed as a variant of minimal change disease or FSGS. The literature reports few cases of PM-associated glomerulopathies, with membranous or immune complex glomerulonephritis predominating. The pathophysiology may involve common immune mechanisms, including the activation of CD8+ T cells and the release of cytokines capable of inducing glomerular damage.

Conclusion: Complete remission of nephrotic syndrome following immunosuppressive treatment directed at the PM suggests a possible shared immune-mediated origin.

Keywords: Polymyositis, glomerulonephritis, dermatomyositis, renal biopsy, mesangial glomerulopathy.

Introduction

Polymyositis is an idiopathic autoimmune myopathy characterized by collagen disorders [1, 2] that primarily affects the proximal muscles and lungs in 36% of cases [3]. It typically has a subacute onset, developing over several weeks. This condition occurs predominantly in women and has an approximate incidence of 1 case per 100,000 inhabitants [1]. Renal manifestations are rare, but two specific types of lesions have been described: acute tubular necrosis related to myoglobinuria and acute rhabdomyolysis [2, 4].

It typically presents with progressive muscle weakness that interferes with daily activities, frequently affecting the posterior pharyngeal wall and the proximal third of the esophagus, causing dysphagia and dyspnea, and is associated with impaired renal function [2]. Furthermore, this condition can be associated with various diseases, including systemic lupus erythematosus, rheumatoid arthritis, Crohn's disease, human immunodeficiency virus (HIV), hepatitis C virus, and human T-cell lymphotropic virus (HTLV) [2,5].

Proliferative mesangial glomerulopathy is the most common histological type of polymyositis [1]. In this context, we present the case of a patient with polymyositis diagnosed by electromyography and the presence of antibodies. Subsequently, a renal biopsy was performed, which revealed increased mesangial matrix and diffuse granular IgM deposition in the mesangium, leading to a diagnosis of IgM mesangial nephropathy.

Case report

Patient information

A 40-year-old woman with a history of dermatitis herpetiformis was diagnosed in 2016 and has been on a gluten-free diet. G3P1C1Ab1, with spontaneous hyperprolactinemia treated with cabergoline, presented with a one-month history of edema, foamy urine due to anasarca, fever, arthralgia, and muscle weakness. She also reported a dry cough, which led her to seek medical care and be hospitalized.

Clinical findings

The physical examination revealed elevated vital signs, including tachycardia, and bibasilar crackles on auscultation. The patient reported generalized pain from her knees to her shoulder girdle, proximal weakness, and edema of the lower extremities.

Diagnostic tests

A 24-hour proteinuria test was performed, yielding a result of 3.5 g/24 hours. The transaminase (AST and ALT) levels were elevated to three times the baseline values (Table 1). Blood cultures (BCs) and urine cultures (UCs) were negative. A chest CT scan revealed bibasilar interstitial infiltrates with air bronchograms, and the echocardiogram was unremarkable. Thyroid,

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abdominal, and pleural ultrasounds were performed, all of which were normal. Five days later, a two-dimensional echocardiogram was performed, revealing tachycardia, a left ventricular end diameter (LVEDD) of 42 mm, a right ventricular end diameter (RVEDD) of 106 mm, a PPD of 0.87, a normal left ventricular ejection fraction (LVEF), and right heart chambers of normal shape and size. Additionally, moderate to severe pericardial effusion with right atrial collapse was observed, along with a nondilated vena cava with inspiratory collapse. Based on echocardiographic findings, the patient was admitted to the intensive care unit (ICU). Owing to a suspected diagnosis of primary or secondary polymyositis, electromyography (EMG) was ordered, revealing findings consistent with acute or subacute myopathic involvement. A noncontrast chest CT scan was subsequently performed, which revealed a small pericardial effusion, bilateral laminar pleural effusion with passive atelectasis of the adjacent parenchyma, a fissural effusion, ground-glass opacities in both inferior posterior lobes (likely related to vascular ectasia), mild hepatic steatosis, and a homogeneous and enlarged right adrenal gland (28 HU). Spirometry revealed that although the FEV1 and FEF 25–75% were reduced, the FEV1/FVC ratio was elevated. The diffusion capacity was severely reduced. Reduced lung volume and an elevated FEV1/FVC ratio, in the absence of parenchymal disease, are suggestive of muscle weakness. Owing to a history of dermatitis herpetiformis and poor dietary adherence, antibodies for Crohn's disease and endoscopic retrograde cholangiopancreatitis (ERCP) were ordered, along with a biopsy, which revealed a sliding hiatal hernia and erosive antritis. Anti-gliadin, anti-transglutaminase, anti-endomysium, and anti-Jo antibodies were positive. Two days later, positive results were also observed for anti-Ro, anti-La/Sm, and anti-RNP antibodies. Antinuclear antibodies (ANAs) and anti-DNA were negative, complement levels were normal, hepatitis B virus (HBV) and hepatitis C virus (HCV) serology were negative, HIV was negative, and muscle biopsy confirmed inflammatory myopathy. Renal biopsy (Figures 1, 2, 3, and 4) revealed the following: corticomedullary casts with up to 22 glomeruli and mild segmental matrix thickening. PAS staining revealed normal cellularity and capillary walls. Immunofluorescence (IF) (Figures 5, 6) revealed IgM with a mesangial and segmental paramesangial parietal pattern. Therefore, glomeruli with mild mesangial matrix increase and IgM deposits are diagnosed (mesangial glomerulopathy with IgM deposits).

Table 1. Renal and hepatic function tests

	05/08	20/08	23/09	25/09	27/09	30/09	08/10	15/10
AST/ASTG (UI/L)	178/118	179/286	613/786	457/632			141/433	
Alkaline phosphatase (IU/L)		87	791	579			500	
LDH (UI/L)			3165	11828			148	
Urea (mg/dL)	22	21	52	64	83	32	33	22
Creatinine (mg/dL)	0.7	0.57	0.43	0.48	0.68	0.4	0.4	0.4
PCR (mg/L)				0.21				
Proteinuria (g/24 h)	3.5							
Urinary sediment								
White blood cells			23		80-90			
Red blood cells			20-25		20-30			
Granular cylinders			Thin and wide		Dark			

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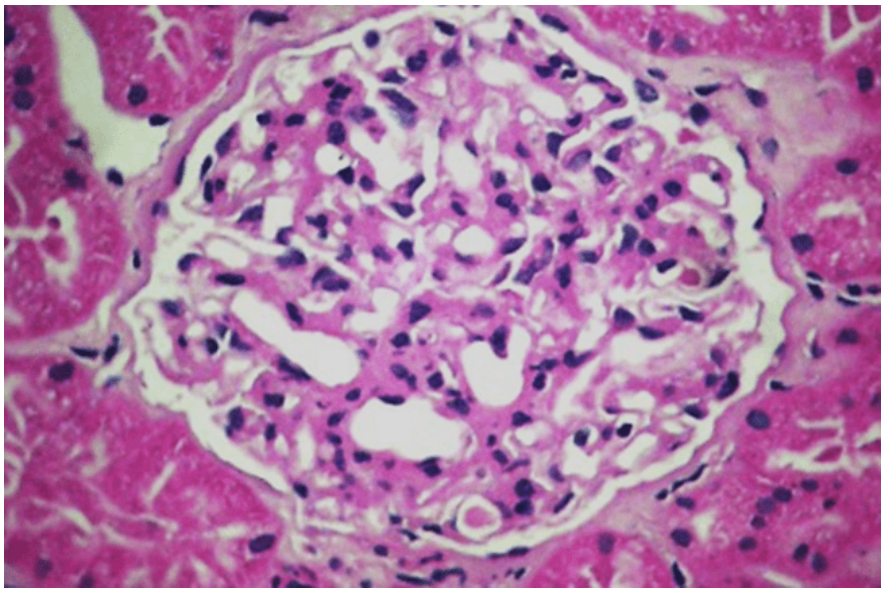
Antibodies

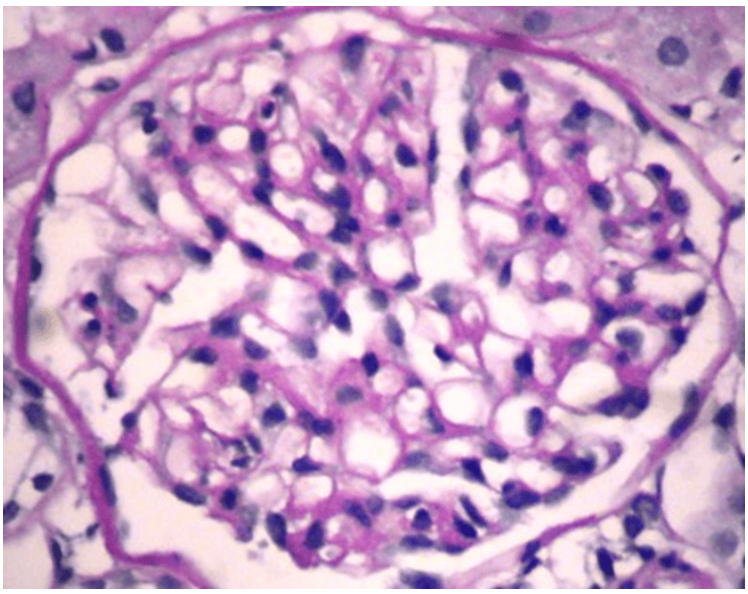
Anti-gliadin positive. Anti-transglutaminase: positive. Anti-endomysium: positive. Anti-Jo: positive. Ac anti-Ro: positive.

Figure 1. Renal biopsy, H&E.



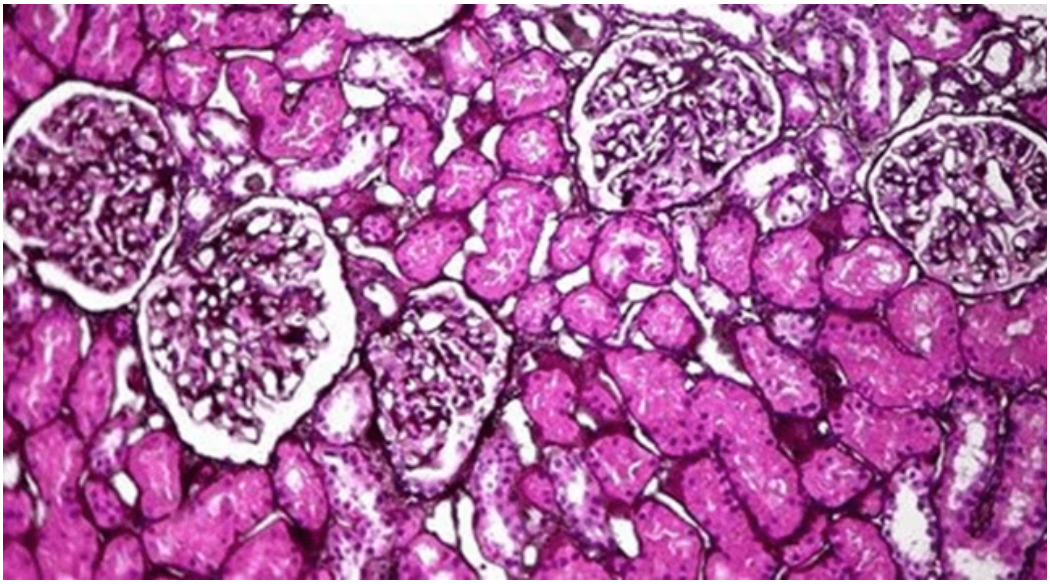
A glomerulus is observed with H&E staining and a slight increase in the mesangial matrix is evident.

Figure 2. Renal biopsy, PAS technique



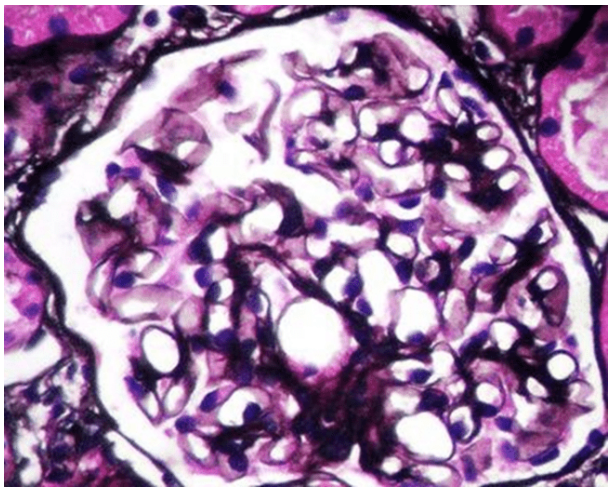
It is evident that the cellularity is normal, as are the capillary walls.

Figure 3. Corticomedullary cylinder.



Renal corticomedullary cylinder containing up to 22 glomeruli. This low-magnification image is representative of the analyzed sample, and as seen, the 5 glomeruli that appear in this cortical sector, the tubules, and the interstitium appear normal.

Figure 4. Renal biopsy, PAS technique



Slight segmental matrix increase.

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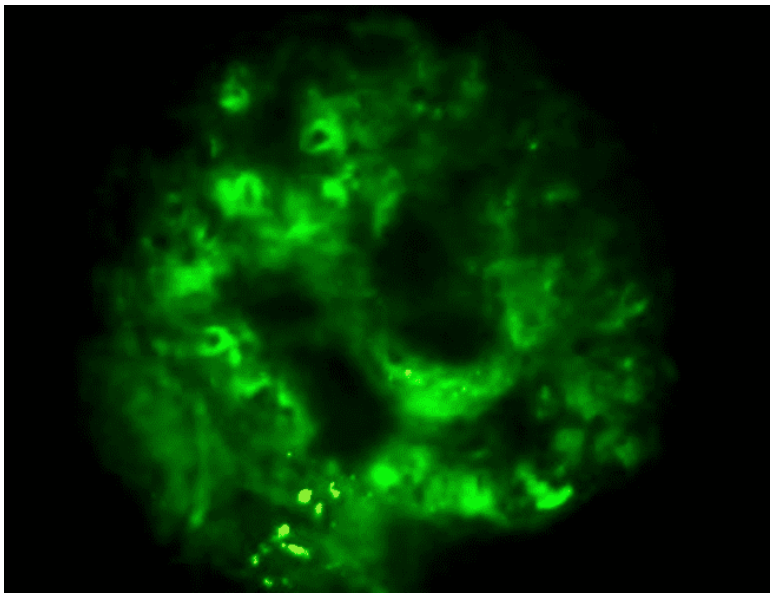
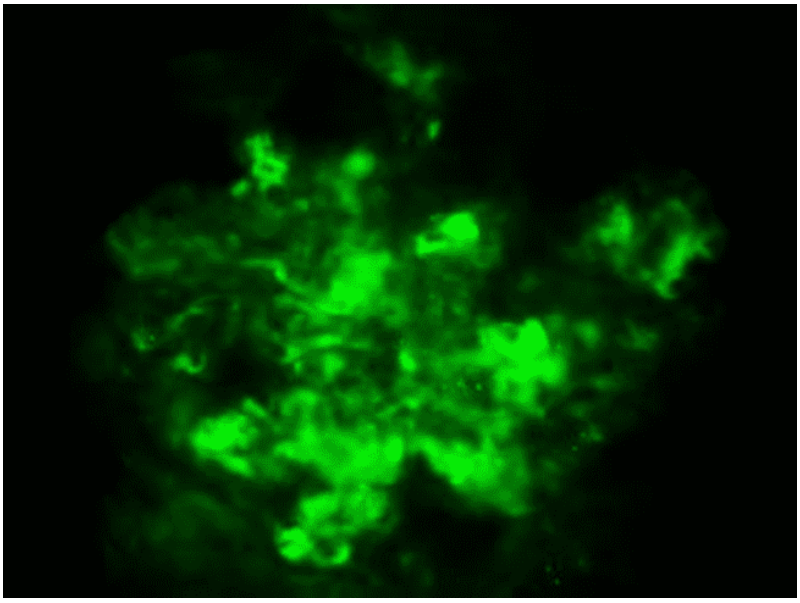
Figure 5. IgM immunofluorescence.



Immunofluorescence. IgM with mesangial and paramesangial parietal pattern.

Figure 6. IgM immunofluorescence.



Immunofluorescence. IgM with mesangial and paramesangial parietal pattern.

Therapeutic intervention

Initially, the patient was treated with ampicillin-sulbactam and clarithromycin. This was complemented by a negative fluid balance; subsequently, 40 mg furosemide, 5 mg enalapril, and aspirin

were administered. On the basis of echocardiogram findings, the patient was transferred to the intensive care unit (ICU) 5 days after admission; however, subsequent follow-up revealed no progression of the condition. Methylprednisolone (1 g pulses) was administered for 3 days, followed by the administration of 80 mg/day Deltisone, with partial improvement of the pericardial effusion. Owing to suspected polymyositis (primary vs. secondary), electromyography and collagenography were subsequently ordered; additionally, celiac disease antibodies and a biopsy were requested.

Evolution and monitoring

Partial improvement of the pericardial effusion was observed after steroid treatment. No progression of the effusion was reported in subsequent echocardiographic follow-up. The diagnosis of inflammatory myopathy consistent with polymyositis, with mesangial glomerulopathy and associated IgM deposits, was confirmed, and the pericardial effusion resolved completely after 4 weeks. The patient is being monitored for systemic involvement, hematuria, and proteinuria.

Discussion

The association between polymyositis (PM) and kidney disease is rare and is generally limited to manifestations such as membranous glomerulonephritis, focal segmental glomerulosclerosis (FSGS), tubulointerstitial nephritis, or kidney damage secondary to rhabdomyolysis [6 , 7]. The development of nephrotic syndrome in a patient with PM should prompt a thorough investigation to rule out secondary causes. In the present case, the histology revealed IgM mesangial nephropathy, a rare and controversial entity that has been debated as a variant or subtype of minimal change disease or FSGS [8 , 9].

In 2004, Yen et al., in a retrospective study, evaluated the incidence of acute or chronic kidney damage in a cohort of 65 patients with inflammatory myopathy (IM). Acute kidney injury (AKI) was reported in 14 patients (21%), chronic kidney disease (CKD) was reported in 1 (1.5%), and 4 (6%) additional patients presented with isolated proteinuria. For these patients, a renal biopsy was performed in two patients with dermatomyositis (DM) and overt proteinuria, revealing membranous nephropathy in one patient and IgA nephropathy in the other; however, some patients included in the study had systemic lupus erythematosus (SLE), which represents a clear bias in the analysis of renal involvement in that cohort [4]. In 2005, Takizawa et al. described twenty-one patients who developed biopsy-proven glomerulonephritis associated with PM/DM. The underlying disease was PM in 15 patients and DM in 6 patients. Both glomerulonephritis and PM/DM were diagnosed concurrently in most of these patients. The pathological diagnoses were as follows: membranoproliferative glomerulonephritis (MPG) in 12 patients, membranous glomerulonephritis (NM) in 6 patients, minimal change disease in 1 patient, and crescentic glomerulonephritis in 2 patients [1]. In 2014, Couvrat et al. conducted a retrospective study that included 150 patients with myositis and carefully excluded those with other systemic autoimmune diseases. Renal involvement was found in 35 (23.3%) patients in the cohort, including 21 (22%) with DM and 6 (23%) with PM. Renal involvement consisted of AKI in 16 patients (10.7%) and CKD in 31

(20.7%). Proteinuria was 90.3 g/d in 26 of 86 patients (30%), with ATN being the main cause of AKI in patients with myositis. Within the cohort, 14 patients (7 men and 7 women) with myositis (6 PM, 5 DM) who underwent renal biopsy at 10 French nephrology centers were identified; immune complex-related glomerulonephritis (n=4) and IgA nephropathy (IgAN) were identified in 2 patients, and NM was identified in 2 patients, one of whom presented with crescentic NM [3].

IgM nephropathy is characterized by mild to moderate mesangial proliferation on light microscopy and diffuse mesangial IgM deposits on immunofluorescence. Although its existence as a separate entity has been questioned, multiple studies have shown that some patients with this histology present a distinct clinical course characterized by increased proteinuria, a reduced response to steroids, and a greater risk of progression to chronic kidney disease [10–12].

In systemic autoimmune diseases, infrequent associations between IgM nephropathy and systemic lupus erythematosus (SLE) or Sjögren's syndrome have been described; however, the relationship with polymyositis is extremely rare, with very few documented cases [13]. These findings suggest that polyclonal B-cell activation in autoimmune diseases could facilitate mesangial deposition of immunoglobulins, including IgM, as part of a dysregulated immune response.

From a pathophysiological point of view, the relationship between polymyositis and renal alterations, on the one hand, is a well-identified cause of toxic ATNs, which are frequently found in patients with inflammatory myopathies, especially myoglobinuria, and the use of nephrotoxic drugs such as intravenous immunoglobulins (IVIGs), which, through renal medullary vasoconstriction and ischemia, cause tubular damage [3]. Furthermore, PM is considered to be caused by a cell-mediated immune process in which clonally expanded, self-aggressive CD8+ T cells contact and invade muscle fibers. These findings suggest that various cytokines released by these self-aggressive CD8+ T cells may stimulate mesangial cells [1], thus contributing to the observed glomerular injury through underlying immune mechanisms in the PM—such as complement activation, aberrant cytokine production, and lymphocyte dysfunction [14]. The presence of predominant IgM deposits suggests an immune pattern distinct from that of classic PM-associated glomerulonephritis, raising the question of whether this nephropathy represents a coincidence or a true secondary manifestation.

The optimal treatment for IgM-induced mesangial nephropathy is not standardized. Most patients receive steroids, but the response is variable, especially in adults [14]. In this case, management focused on systemic immunosuppression with corticosteroids targeting the PM, which led to complete remission of nephrotic syndrome, supporting the hypothesis of a common immune-mediated origin.

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Conclusions

Nephrotic syndrome secondary to polymyositis represents an infrequent but clinically significant manifestation of the systemic spectrum of idiopathic inflammatory myopathies. Its occurrence poses diagnostic and therapeutic challenges, as renal involvement may precede, coincide with, or follow muscular symptoms and reflect a range of glomerular lesions, from membranous nephropathy to immune complex glomerulonephritis. This case underscores the importance of a systematic renal evaluation in patients with polymyositis, even in the absence of initial urinary symptoms, as well as the need for a multidisciplinary approach that integrates clinical, serological, histopathological, and therapeutic findings. Given the scarcity of reports, prospective studies and systematic analyses are needed to characterize the underlying pathogenic mechanisms, establish specific therapeutic guidelines, and optimize the renal and systemic prognoses of these patients.

Abbreviations

DM: dermatomyositis.
GNRs: glomerulonephritis.
GEFS: focal segmental glomerulosclerosis.
ATN: acute tubular necrosis.
IgM: immunoglobulin M.
PM: polymyositis.

Supplementary information

The supplementary materials have not been included.

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Not declared.

Authors' contributions

José Lucas Daza: Conceptualization, data curation, research, visualization, original draft writing.
Nicole Stephany Cifuentes Peralta: Conceptualization, data curation, research, visualization and writing of the original draft.
Salma Yasno López: Conceptualization, formal analysis, methodology, project management, resources, software, supervision, validation, writing–review and editing.
Stefanía Macías Villermo: Conceptualization, data curation, research, visualization and writing of the original draft.
Elba Vanessa Villavicencio Cerón: Conceptualization, visualization and writing of the original draft

All the authors read and approved the final version of the manuscript.

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Clinical cases are not needed.

Consent for publication
The authors have the patient's authorization to publish the pathology images and the clinical case.

Conflicts of interest
The authors declare that they have no conflicts of interest.

Use of generative AI
The authors declare that they did not use generative AI in this document.

Author information
José Lucas Daza, internist and nephrologist. Member of the Colombian Association of Nephrology and Hypertension, Bogotá, Colombia. Master's degree in renal pathology. Ibagué, Tolima, Colombia.
Email: drlucasdaza@gmail.com
https://orcid.org/0000-0002-6430-5415

Salma Yulieth Yasno López, medical student at the University of Tolima.
Email: syyasnol@ut.edu.co
https://orcid.org/0009-0005-1174-8924

Nicole Stephany Cifuentes Peralta, Medical Student at the University of Tolima.
Email: nscifuentesp@ut.edu.co
https://orcid.org/0009-0008-7848-155X

Stefanía Macías Villermo, Medical Intern at the University of Tolima.
Email: smaciasv@ut.edu.co
https://orcid.org/0009-0008-2044-2128

Elba Vanessa Villavicencio Cerón, MD, graduated from the Technical University of Manabí (Portoviejo, 2007). Specialization in Nephrology from the Pontifical Catholic University of Argentina (Buenos Aires, 2016). Attending Physician in the Nephrology Department, IESS Hospital, Portoviejo, Manabí, Ecuador.
Email: elvavice@hotmail.com
https://orcid.org/0000-0002-5184-0427

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