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CASE REPORT

C3 glomerulopathy in a pediatric patient: case report of a rare and complex nephropathy

Glomerulopatia C3 em paciente pediátrico: relato de caso de nefropatia rara e complexa

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ABSTRACT

Introduction: C3 glomerulopathy presents as a group of rare renal diseases caused by the dysregulation of complement system pathways and deposition of C3 components in the glomeruli, leading to inflammatory reactions in the glomeruli at varying levels. Therapeutic approaches based on immunosuppressants and inhibitors of the terminal complement pathway show variable efficacy and do not guarantee universal cure. **Objective:** We present the case of a 6-year-old male patient diagnosed with glomerulopathy C3. **Methods:** Descriptive observational study aimed at reporting a case of C3 glomerulopathy. **Case Description:** We describe a case of a patient diagnosed with C3 glomerulopathy, confirmed by renal biopsy with immunofluorescence microscopy analysis, which revealed a granular mesangial pattern and starry-sky type capillary loops. Therapeutic approaches used for clinical management of the condition are also discussed. **Discussion**: C3G is rare, it has significant clinical implications due to its association with progressive renal insufficiency, high rates of recurrence following kidney transplantation, and graft failure. Currently, there are no specific or curative therapies available for managing C3G.3 However, treatment is directed at reducing proteinuria and controlling renal inflammation. **Conclusion:** C3 glomerulopathy is a rare, severe renal disease with a challenging approach, as there is no specific therapy. Ongoing clinical trials are testing the efficacy of first-generation drugs targeting the alternative complement pathway, which may change this scenario.

Keywords: Glomerulonephritis; Proteinuria; C3 complement; Biopsy; Microscopy, electron.

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RESUMO

Introdução: A glomerulopatia C3 apresenta-se como um conjunto de patologias renais raras, acarretadas pela desregulação de vias do sistema complemento e deposição do componente C3 nos glomérulos, o que provoca reações inflamatórias glomerulares em níveis variáveis. As terapêuticas são baseadas em imunossupressores e inibidores da via terminal do complemento apresentam eficácia variável e não garantem cura universal. **Objetivo:** Apresentamos o caso de um paciente masculino de 6 anos de idade com diagnóstico de glomerulopatia C3. **Métodos:** Estudo observacional descritivo, que objetiva descrever relato de caso de glomerulopatia C3. **Descrição do Caso:** Descrevemos um caso de paciente com diagnóstico de glomerulopatia C3 constatado por biópsia renal com análise por microscopia com imunofluorescência, que evidenciou padrão granular mesangial e em alças capilares tipo céu-estrelado (*starry-sky*) e terapêuticas utilizadas para o manejo clínico do quadro. **Discussão:** Apesar da raridade, a C3G apresenta impacto clínico significativo devido à associação com insuficiência renal progressiva, altas taxas de recidiva pós-transplante renal e de falhas no aloenxerto. Atualmente, não há terapêuticas específicas e curativas disponíveis para o manejo de glomerulopatia. Todavia, a abordagem é direcionada ao tratamento para atenuar a proteinúria e o quadro inflamatório renal. **Conclusão:** A glomerulopatia C3 é uma doença renal rara, grave e de abordagem desafiadora, uma vez que não há terapêutica específica. Ensaios clínicos estão em andamento para testar a eficácia de drogas com alvo na via alternativa do complemento, que podem alterar esse cenário.

Palavras-chave: Glomerulonefrite; Proteinúria; Complemento C3; Biópsia; Microscopia eletrônica.

Introduction

C3 glomerulopathy (C3G) encompasses a rare and recently defined group of kidney disorders characterized by dysregulation of the complement system, primarily the alternative pathway, resulting in predominant C3 deposition within the glomeruli and a spectrum of inflammatory injury[1]. C3G, a condition that has been recently defined, is subclassified into two main entities: dense deposit disease (DDD) and C3 glomerulonephritis (C3GN), both associated with impaired regulation of complement activation[2].

Current treatment strategies, including immunosuppressive agents and terminal complement inhibitors, demonstrate variable efficacy and do not achieve sustained remission in most cases. Although several clinical trials are evaluating complement-targeted therapies, particularly agents that inhibit the alternative pathway, no definitive cure is currently available. Renal transplantation carries a high risk of disease recurrence, further complicating management[3].

In this article, we present the case of a patient with biopsy-confirmed C3G, in which immunofluorescence microscopy revealed a granular mesangial and capillary loop deposition pattern, consistent with a "starry-sky" appearance. Additionally, we provide a comprehensive review of the current and emerging therapeutic approaches under investigation for C3G.

Objective

In this report, we present the case of an 8-year-old male patient diagnosed with C3 glomerulopathy. The aim of this article is to describe the clinical findings and correlate them with current knowledge available in the literature regarding this rare renal disorder.

Methods

This is a descriptive observational study aimed at reporting a clinical case of C3 glomerulopathy. Case information was obtained through medical chart review, a clinical interview with the patient, and photographic documentation, all of which were carried out with the patient's assent. These

data were subsequently correlated with findings from the scientific literature. The patient, a minor, provided assent through the signing of the informed assent form, and the legal guardian provided written informed consent. The study was approved by the Research Ethics Committee (CEP) under protocol number 86868225.9.0000.5342, and conducted in accordance with the principles outlined in the Declaration of Helsinki.

Case Report

A 6-year-old male patient was referred to the Pediatric Nephrology outpatient clinic with clinical and laboratory findings (Table 1) compatible with nephritic syndrome, presenting with hypertension, hematuria, and nephrotic-range proteinuria, associated with persistent laboratory evidence of C3 consumption. A percutaneous renal biopsy, guided by ultrasonography, was performed to further investigate the condition. Histopathological analysis of the biopsy specimen under light microscopy revealed mild and diffuse expansion of the mesangial matrix and cellularity in the glomerular compartment (Figure 1.1). In the tubulointerstitial compartment, red blood cells and red blood cell casts were observed filling the tubules, while the vascular compartment showed no significant abnormalities (Figure 1.2). Immunofluorescence microscopy demonstrated a coarse, moderately intense granular pattern with positivity for C3 fragment immunostaining (Figure 2), showing a characteristic mesangial and capillary loop "starry-sky" pattern. Trace amounts of IgM were also detected, while other immunologic markers were negative. Electron microscopy revealed numerous small, electron-dense glomerular deposits, primarily in subendothelial and transmembrane regions, some of which resembled "dense bands" (Figure 3). These findings are characteristic of C3-dominant glomerulonephritis and may be indicative of dense deposit disease. No subepithelial humps, mesangial or organized (fibrillar) deposits were observed. Complementary

Table 1. Laboratory findings at presentation.

Test	Result	Reference Range
Hemoglobin	10,5 g/dL	12-14,5 g/dL
Hematocrit	31,9%	36-43%
White blood cell count	7.410 mm3	3.400-10.800mm3
Platelet count	352.000/mm3	140-450.000/mm3
Total cholesterol	308 mg/dL	<170 mg/dL
LDL cholesterol	221 mg/dL	Inferior a 130 mg/dL
HDL cholesterol	51 mg/dL	>45 mg/dL
Triglycerides	181 mg/dL	<85 mg/dL
Serum creatinine	0,53 mg/dL	0,2 a 0,7 mg/dL
Protein-to-creatinine ratio	3,18	Até 0,20
Microalbuminuria	4.425,8 µg/mg	Inferior a 30 µg/mg
Cystatin C	1,04 mg/L	0,62-1,11 mg/L
Serum albumin	2,3 g/dL	3,5-5,5 g/dL
Potassium	4,6 mEq/L	3,5-5,1 mEq/L
Serum calcium	9,5 mg/dL	8,6-11,3 mg/dL
Serum phosphorus	5,0 mg/dL	4,0-7,0 mg/dL
PTH (intact)	44 pg/mL	18-94 pg/mL
Ferritin	90,2 ng/mL	7-140 ng/mL
Transferrin saturation	31%	20-50%
Complement C3	10 mg/dL	90-170 mg/dL
Complement C4	26 mg/dL	12-36 mg/dL
CH50 complement activity	12,98 U/mL	42-95 U/mL
Protein electrophoresis	No monoclonal proteins	No monoclonal proteins
Urinalysis	Proteins: 3+; Hemoglobin: 3+; Casts; Dysmorphic RBCs	Normal
Urine culture	Negative	Negative

Legend: Laboratory findings at investigation.

investigations included a genetic panel for kidney diseases through genomic analysis using next-generation sequencing (NGS) and nanopore sequencing to investigate variants in genes associated with glomerulopathies. No pathogenic variants were identified in the analyzed genes, including in the CFHR5 gene, that could, by themselves, explain the patient's clinical presentation. Active and chronic infectious diseases were also excluded, such as cytomegalovirus infection, toxoplasmosis, hepatitis B, hepatitis C, HIV, as well as autoimmune conditions and lymphoproliferative disorders.

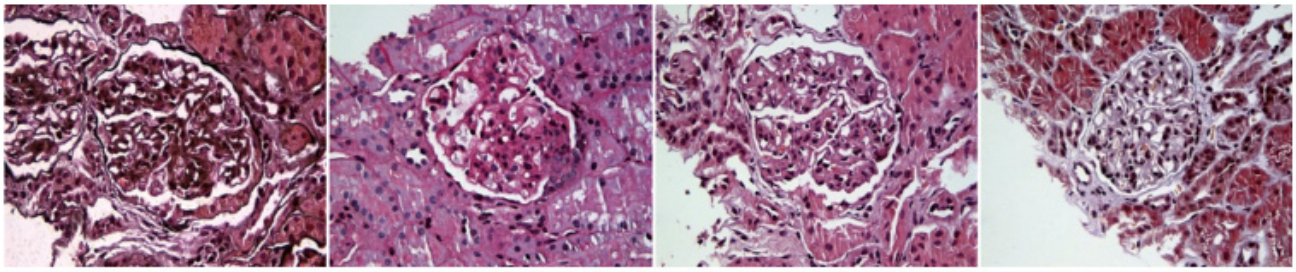

Figure 1. Light microscopy of renal cortex and medulla stained with H&E, PAS, Gomori's Trichrome, and Jones' Methenamine Silver.

Figure 1.1. Light microscopy of glomerular compartment.

Legend: The mesangial matrix and cellularity are mildly and diffusely expanded. The glomerular basement membrane appears unremarkable, with no evidence of holes, saw-tooth irregularities, splitting, or corrugation. Fibrous capsular adhesions are observed in Bowman's space. There are no signs of crescent formation, fibrin deposits, or necrosis. No unequivocal evidence of endocapillary hypercellularity, hyalinosis, segmental sclerosis, or fibrin microthrombi is observed. Occasional neutrophils are seen within glomerular capillary loops.

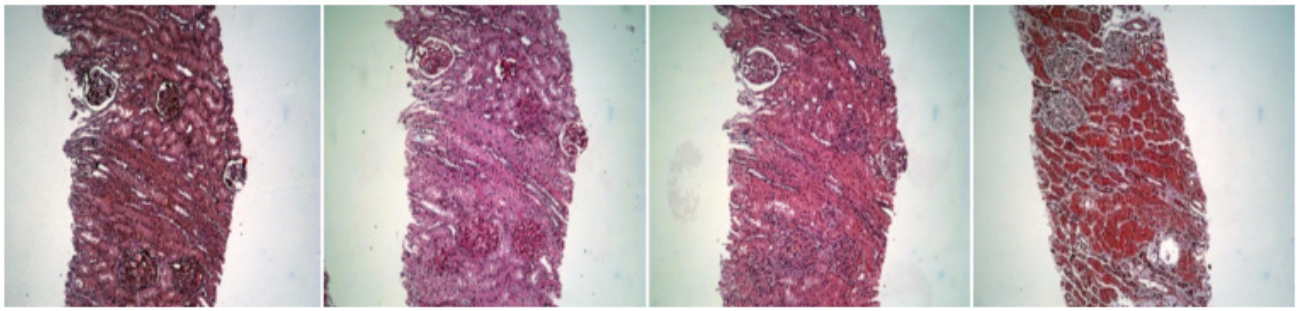

Figure 1.2. Light microscopy of tubulointerstitial compartment.

Legend: The tubulointerstitial compartment is well preserved, with no evidence of fibrosis or inflammatory cell infiltration. Red blood cells and hematic casts are observed filling some tubular lumina. Frequent vacuolization of epithelial cells is noted, consistent with protein reabsorption.

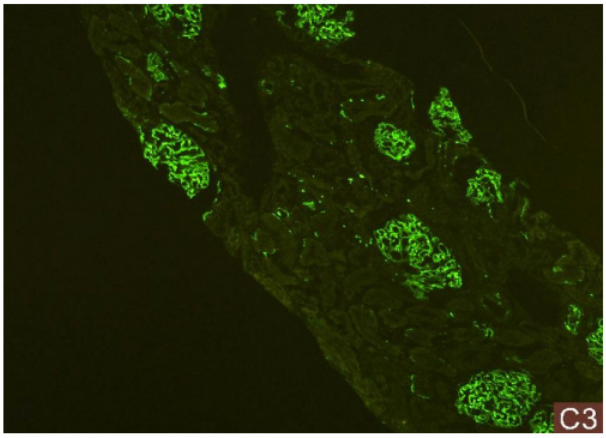

Figure 2. Immunofluorescence microscopy.

Legend: Analysis of renal fragment with C3 fragment antibodies at a 1:40 dilution, showing a positive result of 2+/3+, with granular mesangial and "starry-sky" capillary loop pattern.

The therapeutic approach focused on antiproteinuric management and non-specific immunomodulatory therapy. The patient was started on Enalapril 1 mg/mL (8 mL twice daily) to control proteinuria, along with corticosteroid therapy using Prednisolone 3 mg/mL (7 mL daily) and Mycophenolate Sodium 360 mg (twice daily). The patient achieved clinical remission and remains under quarterly follow-up at the Pediatric Nephrology Service.

Discussion

C3 glomerulopathy (C3G) is a rare, chronic renal disorder characterized by the presence of isolated or dominant C3 deposits in the glomeruli, identified through immunofluorescence microscopy of renal biopsy specimens. These deposits result from hyperactivity and dysregulation of the alternative complement pathway, which is the primary mechanism underlying glomerular injury in this disease. Based on electron microscopy findings and genetic investigations, the disease can be subclassified into two main presentations: C3 glomerulonephritis (C3GN) and dense deposit disease (DDD)[4].

Both C3GN and DDD are believed to share a pathogenesis based on acquired and genetic alterations in the alternative complement pathway. Advancements in understanding this association have contributed to the development of new therapeutic perspectives[5,6]. In some cases, mixed features of both C3GN and DDD can be observed on electron microscopy. Some researchers emphasize that electron microscopy is not specific for distinguishing these entities, suggesting that they may represent the same disease at different stages, referred to as Complement Alternative Pathway-Mediated Glomerulonephritis[3,6]. Nevertheless, it is anticipated that with ongoing genetic and molecular research, the differentiation between these two forms will become more feasible.

Although C3G is rare, it has significant clinical implications due to its association with progressive renal insufficiency, high rates of recurrence following kidney transplantation, and graft failure[2,7]. C3G predominantly affects children and young adults, although it can occur at any age. The clinical presentation of C3G is highly variable, ranging from asymptomatic cases with only laboratory alterations, such as hematuria and proteinuria detected on urinalysis, to more severe presentations involving nephritic syndrome, renal impairment, hypertension, as seen in the current case, and progressive renal failure[8]. Approximately half of patients with C3G progress to end-stage renal disease (ESRD) within 10 years of diagnosis[7]. Severe cases may progress rapidly to ESRD, and the disease typically follows a relapsing-remitting course, often triggered by infectious episodes[8].

The suspicion of C3G is raised by clinical presentations of nephrotic or nephritic syndrome, or urinary analysis changes, combined with persistent C3 consumption. Diagnosing C3G can be challenging, as confirmation requires renal biopsy and immunofluorescence microscopy to detect glomerular C3 positivity, along with electron microscopy for subclassification[4,8]. However, findings on electron microscopy can be variable, making it difficult to differentiate between C3GN and DDD[2,7].

In patients with a suspected C3 glomerulopathy (C3G), measurement of serum C3 and C4 levels, genetic testing, and screening for autoantibodies against CFH and nephritic factors, components that stabilize the alternative pathway C3 convertase, leading to excessive and prolonged complement activation, a key mechanism in disease pathophysiology and progression, are encouraged[9]. Recommended tests, such as assessment of C3 nephritic factor (C3NeF), an autoantibody that stabilizes C3 convertase, and evaluation of the CFHR5 gene, are important in the investigation of C3 glomerulopathies. Genetic analysis, including CFHR5 testing, is critical to identifying hereditary factors with variants that impair complement regulation and

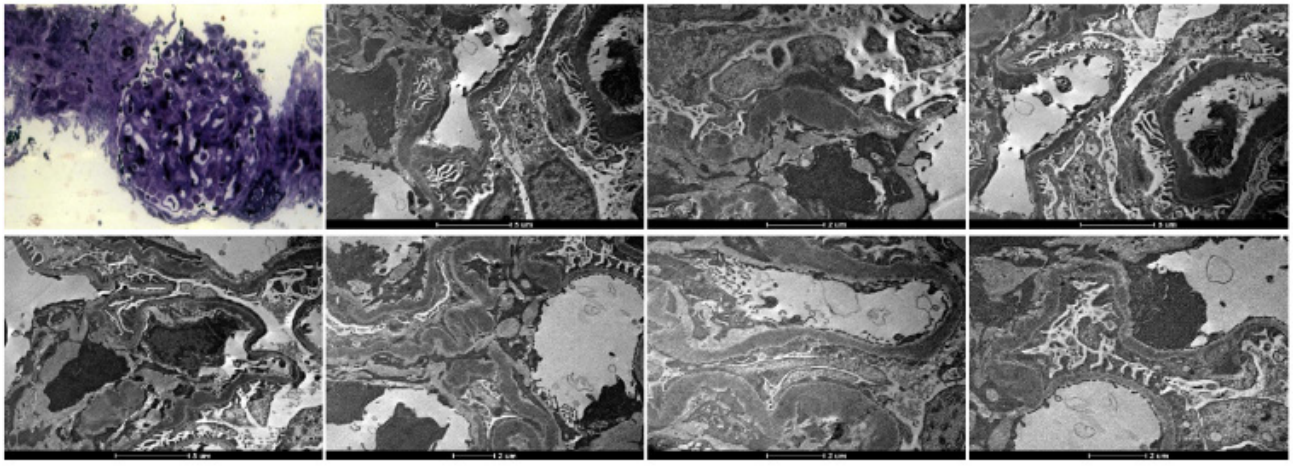

Figure 3. Electron microscopy.

Legend: Electron microscopy image showing numerous small electron-dense glomerular deposits, predominantly located in the subendothelial and transmembrane regions, resembling a "dense ribbon" pattern, characteristic of C3-dominant glomerulonephritis, and possibly corresponding to Dense Deposit Disease.

lead to dense subendothelial deposits similar to those observed in Dense Deposit Disease (DDD)[10-12]. Evaluation of these factors, with distinction between sporadic and familial forms, may aid in the diagnostic interpretation and prognosis of C3 glomerulopathy[9].

Although the genetic and laboratory investigations were detailed, tests for anti-Factor H antibodies and C3 nephritic factor (C3NeF) were not performed, limiting the assessment of potential autoimmune mechanisms in C3 glomerulopathy. Furthermore, this is a case report, with inherent limitations in generalizing the findings and evaluating long-term outcomes. These restrictions should be considered when interpreting the clinical relevance of the case and extrapolating the results to other patient populations.

It is also crucial to exclude chronic or active infectious diseases, genetic disorders, autoimmune conditions, and lymphoproliferative neoplasms[6]. Although no active or chronic infection was documented in this case, the exclusion of infection-associated C3 glomerulopathy (C3G) remains essential, as the rapid clinical recovery is consistent with this presentation. In this case, a comprehensive serological and infectious workup was performed and yielded negative results. Evidence indicates that dysregulated activation of the alternative complement pathway, frequently mediated by factors such as the nephritic factor (C3NeF), can be potentiated by infections, which may act as triggers for renal injury and disease progression, underscoring the necessity of systematically ruling out an infectious component[13,14].

It is important to note that, currently, there are no specific or curative therapies available for managing C3G[3]. However, treatment is directed at reducing proteinuria and controlling renal inflammation[7]. Among the therapeutic strategies, non-immunomodulatory therapy is used for patients with mild disease, while non-specific immunomodulatory therapy, such as Mycophenolate Mofetil combined with glucocorticoids, is reserved for moderate to severe cases[8]. It is important to emphasize that the use of Mycophenolate Mofetil in patients with C3G is off-label, especially in children. Caravaca-Fontán et al. (2020)[15] showed that corticosteroids combined with Mycophenolate Mofetil may induce remission and improve renal outcomes, although treatment response varies among patients.

Complement-targeted therapies are currently undergoing clinical trials and hold promise for better management of this disease[2,3,8]. Recent data on complement inhibitors highlight their therapeutic potential in C3G. The APPEAR-C3G trial showed that Iptacopan significantly reduced proteinuria and had an acceptable safety profile in adults, although children were not included in the study[16]. Clinical trials with Pegcetacoplan, which included only children older than 12 years, also demonstrated significant reductions in proteinuria, improvements in complement biomarkers, and stability of renal function[17-19]. However, it is important to emphasize that robust clinical data are not available for children under 12 years of age.

In terms of prognosis, registry data suggest that pediatric patients with C3 glomerulopathy (C3G), even in the presence of glomerular crescents, may exhibit more favorable outcomes compared to adults. Renal function tends to be better preserved in children, possibly due to differences in immune response and renal plasticity, although the risk of recurrence and disease progression still requires long-term follow-up[20,21].

Although there are no established guidelines for specific therapies for C3G, recent studies highlight the development of complement-targeted therapies, such as Iptacopan and Pegcetacoplan, which aim to prove efficacy in reducing proteinuria and controlling disease progression, with promising results[16,22,23]. These interventions may represent a paradigm shift in treatment, which has previously been reliant on corticosteroids and immunosuppressants, often with limited efficacy. The development of complement-specific therapies could offer a more effective and beneficial approach for managing C3G in the future.

Conclusion

C3 glomerulopathy is a rare and severe condition resulting from dysregulation of the complement system and the deposition of C3 in the glomeruli. Treatment remains complex, as current therapies exhibit variable efficacy and do not provide a definitive cure. Early diagnosis and continuous clinical monitoring are essential for effective management. Ongoing clinical trials hold promise for the development of more effective treatments. The advancement of research aimed at therapeutic strategies for C3 glomerulopathy should remain a priority for researchers in this field.

Author's contributions

We describe contributions to the papers using the taxonomy (CRediT) provide above: *Conceptualization, Investigation, Methodology, Visualization & Writing – review & editing*: A Duarte; L Renner; LA de Lima; LGR Raupp; I de A Brkanitch; LN da Rosa. *Project administration, Supervision & Writing – original draft*: A Duarte; L Renner; LA de Lima; LGR Raupp; I de A Brkanitch; LN da Rosa. *Validation & Software*: A Duarte, L Renner; LA de Lima; LGR Raupp; I de A Brkanitch; LN da Rosa. *Resources & Funding acquisition*: A Duarte; L Renner. *Data curation & Formal Analysis*: A Duarte; L Renner.

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