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The glomerulus in bloom- ultrastructural tapestry of collagenofibrotic glomerulopathy

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Abstract

We highlight an ultrastructural image of collagenofibrotic glomerulopathy, a rare disorder marked by mesangial and subendothelial type III collagen deposition, and place it within the current consensus on diagnostic criteria and clinical implications. The fibrils show a woven, thread-like pattern of delicate interlacing bundles, reminiscent of fine flowering twigs in a tapestry, highlighting the beauty of high-quality electron microscopy visualization in nephropathology.

Keywords Collagenofibrotic glomerulopathy, Type III collagen, Electron microscopy, Non-immune glomerulopathy

Collagenofibrotic glomerulopathy (CFG), also known as collagen type III glomerulopathy, is an exceptionally rare glomerular disease defined by deposition of type III collagen within mesangial and subendothelial regions. Somewhat more than 100 cases of CFG have been reported worldwide [1]. CFG is considered a sporadic disease, however, an autosomal recessive type of inheritance affecting some families has been reported predominantly in the pediatric population [2, 3]. The etiology of this disease remains unknown.

Type III collagen is not normally present in glomeruli; its emergence within the capillary wall and mesangial matrix is pathological and reflects a distinct, non-immune mechanism of extracellular matrix dysregulation [1]. Although type III collagen may be detected in glomeruli as a secondary change in majority of advanced chronic glomerular diseases, its prominent and primary accumulation as a defining feature is characteristic of CFG.

Despite being reported worldwide, CFG remains under-recognized, in part due to its histopathological overlap with more common disorders such as membranoproliferative pattern of glomerular injury, diabetic nodular sclerosis, chronic thrombotic microangiopathy, and amyloidosis. Light microscopy may demonstrate lobular accentuation or amorphous PAS-positive mesangial widening, but neither pattern is specific [1, 4, 5]. Immunofluorescence is typically negative for immunoglobulins and complement, consistent with a non-immune pathogenesis. Because nodular and/or deposition-like patterns on light microscopy can be shared across multiple entities, ultrastructural evaluation is often essential to resolve the differential diagnosis.

Electron microscopy (EM) remains the decisive diagnostic modality in CFG. Recognition of this entity is important because visualization of the diagnostic collagen fibrils may be enhanced by phosphotungstic acid contrast; in some cases, without this additional contrast the characteristic deposits can be subtle or missed on routine EM preparation [2]. Its defining ultrastructural pattern consists of abundant fibrils with a periodicity of 40–65 nm, deposited within the mesangial matrix and

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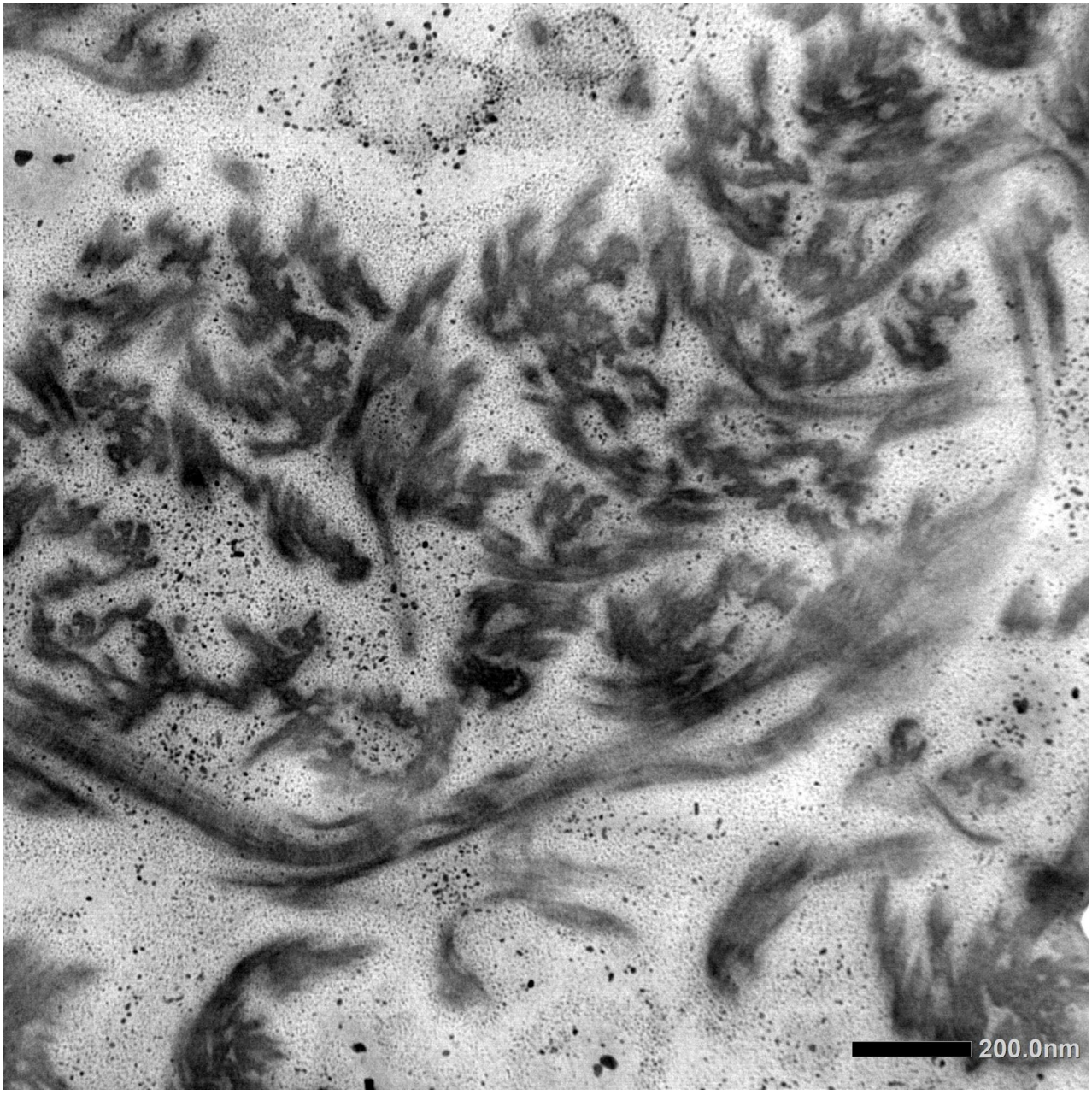

Fig. 1 Electron micrograph demonstrating a markedly expanded mesangial matrix filled with collagen fibrils showing characteristic D-periodicity (banded pattern) and measuring approximately 50 nm in diameter, arranged in irregular, slightly curved bundles extending into the subendothelial zone. This ultrastructural pattern is characteristic of type III collagen deposition in collagenofibrotic glomerulopathy. In this case, the fibrils display a "woven, thread-like pattern," forming delicate interlacing bundles. Scale bar: 0.2 µm

along the subendothelial aspect of the capillary wall [1, 6]. These fibrils show a distinctive morphology: they are thicker than normal interstitial collagen fibrils, form irregular and at times loosely bundled curved aggregates, and often display frayed or tapering ends. Immunohistochemistry or immunoelectron microscopy provides confirmatory evidence by demonstrating type III collagen within the deposits [3].

The presented images (Fig. 1, Supplementary Figures) demonstrate these hallmarks with striking clarity. The fibrils create irregular, interwoven bundles that expand the mesangial matrix and displace the adjacent glomerular basement membrane—an appearance that is readily distinguishable from amyloid, immunotactoid glomerulopathy, diabetic glomerulosclerosis, and fibrillary glomerulonephritis. Such high-quality EM visualization is particularly valuable in biopsies with extensive global

sclerosis, where only a few viable glomeruli remain to establish the diagnosis.

From a clinical perspective, CFG typically manifests with proteinuria, hypertension, and progressive renal dysfunction [1, 7, 8]. Serum procollagen type III N-terminal propeptide (PIIINP) has been reported as elevated in several published cases, but it is non-specific and not required for diagnosis. Both sporadic and familial cases have been documented. The latter demonstrate autosomal recessive inheritance in some reports [3], although genetic testing is not widely accessible.

Management remains supportive, with renin–angiotensin system blockade as the primary strategy to reduce proteinuria and slow progression. No disease-specific therapy exists, and the role of immunosuppression has not been validated [1, 8]. According to published literature, progression to kidney failure is common, typically within a decade, and recurrence after kidney transplantation has been documented [6].

The present case illustrates these principles. The patient was a 71-year-old man with a history of hypertension who presented with nephrotic-range proteinuria (3.7 g/24 h) and hypoalbuminemia (serum albumin 2.8 g/dL), accompanied by chronic kidney disease (eGFR 44 mL/min/1.73 m^2 and serum creatinine 137 µmol/L at the time of biopsy). He had no erythrocyturia, extrarenal manifestations, or syndromic features. There was no history of diabetes mellitus and no clinical or laboratory evidence of monoclonal gammopathy based on the available work-up. Family history for kidney disease was negative.

Light microscopy of the biopsy, containing 28 glomeruli, demonstrated marked lobular accentuation, a nodular sclerosis–like appearance, and global glomerulosclerosis in 5 glomeruli, with amorphous mesangial material as the dominant feature. Immunofluorescence revealed negative immunoglobulins and complement without evidence of light-chain restriction. These histomorphological pattern indicated deposition of non immune material in glomeruli, resembling of diabetic glomerulopathy or other deposition diseases. Given the non-specific nodular deposition-like pattern on light microscopy and the negative immunofluorescence, EM was performed to clarify the nature of the mesangial and capillary wall deposits and to resolve the differential diagnosis.

For EM, samples were contrasted with lead citrate and phosphotungstic acid following the method described by Gubler et al. [2]. Ultrathin sections were examined using a JEM-1400 Flash transmission electron microscope (JEOL, Japan). EM was therefore decisive, demonstrating the characteristic type III collagen fibrils; immunostaining confirmed their identity. Genetic testing for collagenofibrotic glomerulopathy was not available. Serum procollagen type III N-terminal propeptide (PIIINP) levels were not assessed. Despite optimal supportive therapy, kidney function progressively declined, and the patient reached kidney failure within five years; however, he remains clinically stable on maintenance dialysis.

Conclusions

This descriptive, image-based case exemplifies the importance of EM in resolving the differential diagnosis of mesangial expansion and deposition-like lesions. As precision nephropathology increasingly integrates molecular tools, high-quality ultrastructural visualization remains indispensable for recognizing disorders such as CFG that lack specific clinical or serological signatures. Keeping in mind the full spectrum of glomerular diseases, including rare entities such as CFG, is essential for guiding the appropriate use of EM, including the application of phosphotungstic acid contrast when indicated.

Abbreviations

CFG	Collagenofibrotic glomerulopathy
CKD	Chronic kidney disease
eGFR	Estimated glomerular filtration rate
EM	Electron microscopy
PAS	Periodic acid–Schiff
PIIINP	Procollagen type III N–terminal propeptide

Supplementary Information

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Supplementary Material 1

Supplementary Material 2

Supplementary Material 3

Supplementary Material 4

Supplementary Material 5

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Author contributions

Ž. Večerić-Haler – clinical nephrology, case analysis, manuscript preparation; N. Kojc – histopathology and electron microscopy interpretation; manuscript editing; J. Pleško – electron microscopy interpretation and figure preparation.

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Data availability

Detailed data are available at corresponding author upon request.

Declarations

Ethics approval and consent to participate

Not applicable (single case report).

Consent for publication

Written informed consent for publication of this image and clinical data was obtained from the patient.

Reporting guidelines

This case report adheres to the CARE guideline; the completed CARE checklist is provided as a supplementary file.

Artificial intelligence (AI) assistance
During the preparation of this manuscript, the authors used a generative AI tool to assist with grammar and style editing. The authors reviewed and revised all content and take full responsibility for the final manuscript.

Competing interests
The authors declare no competing interests.

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