Crystal-Storing Histiocytosis in Monoclonal Gammopathy: A Case with Glomerular Involvement

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A 51-year-old woman was referred to the nephrology department for evaluation of proteinuria (358 mg/24h) and microscopic hematuria, with preserved kidney function. Kidney ultrasound was unremarkable. Laboratory evaluation revealed increased serum and urinary free kappa light chains, and serum protein electrophoresis with immunofixation identified an IgG kappa monoclonal component, consistent with IgG kappa monoclonal gammopathy of undetermined significance. She was placed under hematology surveillance.

Over two years, proteinuria increased to 624 mg/24 h, with albuminuria of 196 mg/24 h, suggesting a glomerular component of the proteinuria and prompting a kidney biopsy.

Light microscopy showed focal and segmental glomerulosclerosis with minimal chronic changes. Immunofluorescence (IF) on paraffin-embedded tissue showed granular deposits of kappa light chain, markedly exceeding lambda light chain, numerous in proximal tubular epithelial cells and rare in glomerular mesangium and capillary walls (Fig. 1). Electron microscopy (EM) revealed intralysosomal electron-dense material, some with crystalline appearance, within mesangial histiocytes (Fig. 2) and in proximal tubular epithelial cells.

These findings favored the diagnosis of renal crystal-storing histiocytosis (CSH) with glomerular involvement, associated with light chain–related tubulopathy. Bone marrow examination showed no CSH. Clone-directed therapy was initiated, and the patient achieved renal response after the second cycle.

CSH is rare, usually associated with plasma cell neoplasms or lymphoproliferative disorders.[1] Renal involvement is uncommon and typically interstitial[1,2]; glomerular involvement alone is exceedingly rare. Recognition is essential, as protease-digested IF on paraffin-embedded tissue is crucial to unmask monoclonal light chain deposits,[3] and EM is critical for diagnosis. This finding should prompt appropriate hematologic evaluation and has direct therapeutic implications.[4]

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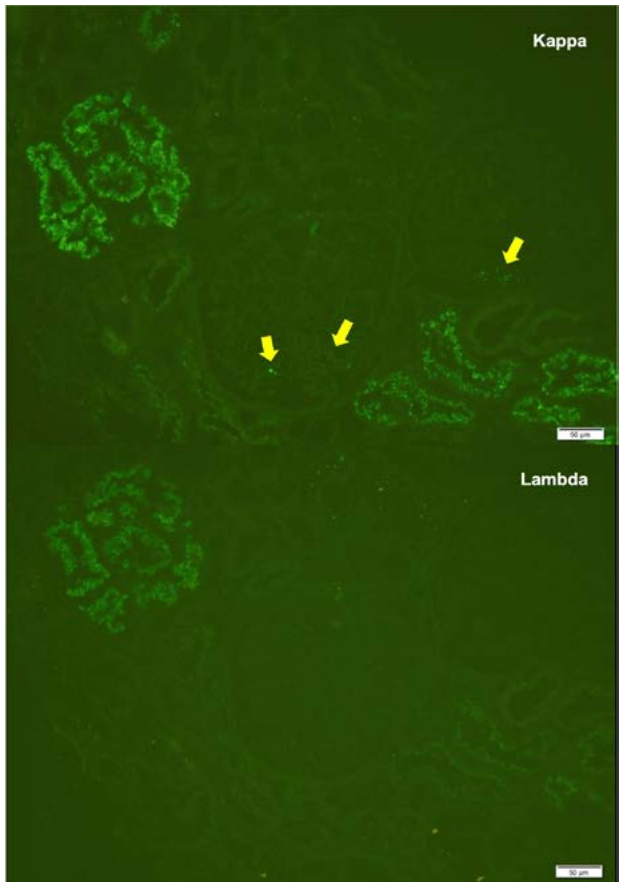

Figure 1. Immunofluorescence on paraffin-embedded tissue demonstrating kappa light chain staining in mesangium and glomerular capillary walls (arrow) and in proximal tubular epithelial cells, markedly exceeding lambda light chain staining.

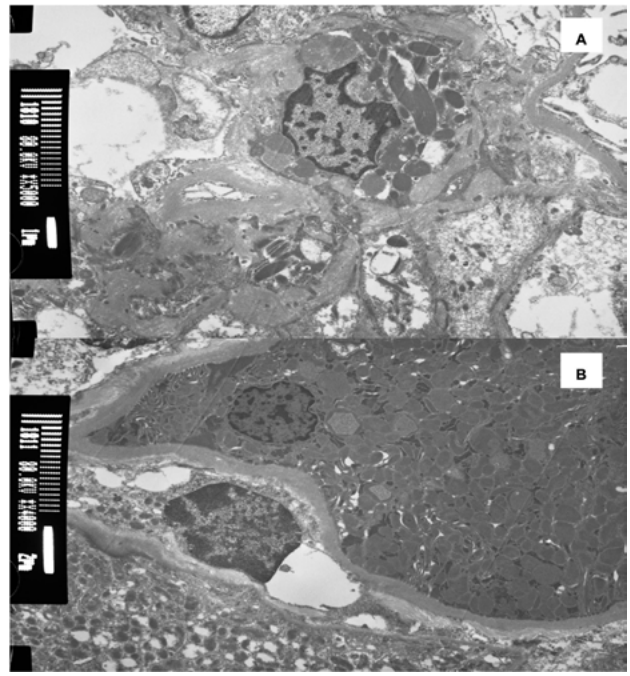

Figure 2. Electron microscopy revealed numerous intralysosomal electron-dense material, some of them with elongated, geometric crystalline appearance, within histiocytes in the glomerular mesangium (A) and in tubular epithelial cells of proximal tubules (B).

Ethical Disclosures

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REFERENCES

1. Gupta RK, Rosenberg AZ, Bagnasco SM, Arend LJ. Renal crystal-storing histiocytosis involving glomeruli: a comprehensive clinicopathologic analysis. Ann Diagn Pathol. 2019;43:151403. doi:10.1016/j.anndiagpath.2019.151403.
2. Shah S, Sethi S, Arend L, Geetha D. Crystal-storing histiocytosis. Kidney Int. 2016;89:507–11. doi:10.1016/j.kint.2015.12.012.
3. Larsen CP, Messias NC, Walker PD, Fidler ME, Cornell LD, Hernandez LH, et al. Membranoproliferative glomerulonephritis with masked monotypic immunoglobulin deposits. Kidney Int. 2015;88:867–73. doi:10.1038/ki.2015.195.
4. Randone P, Burdese M, Barreca A, Oliva S, Sanna E, Abbasciano I, et al. The matter of kidney biopsy in monoclonal gammopathy of renal significance: a case report of a new pattern of immunoglobulin-storing histiocytosis. Case Rep Nephrol Dial. 2023;13:191–6. doi:10.1159/000533913.