

**Title:** *IgG1 Heavy Chain Deposition Disease with Fibrillary Glomerulonephritis as a Manifestation of MGRS*

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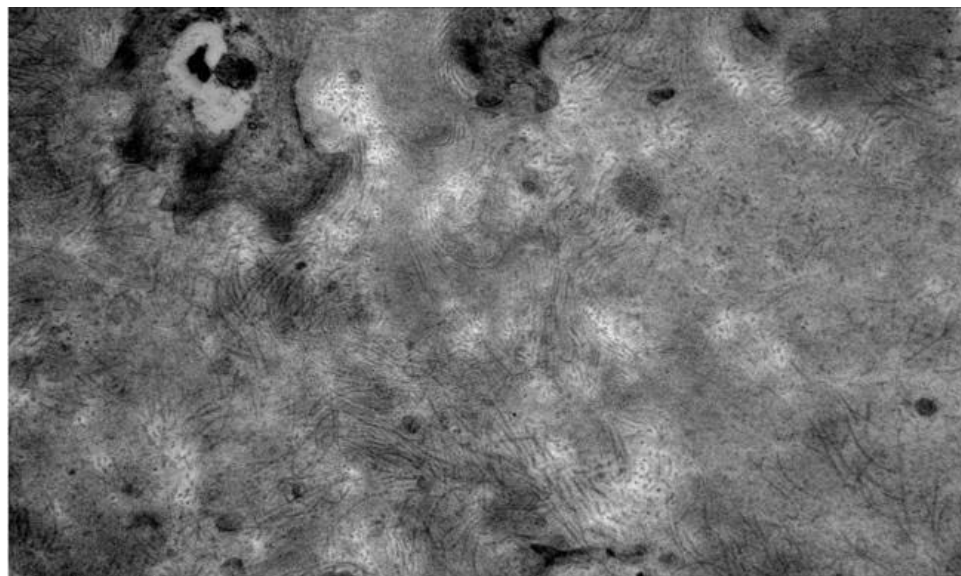
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**Introduction:** Monoclonal gammopathy of renal significance (MGRS) refers to kidney diseases caused by pathogenic monoclonal immunoglobulins. Heavy chain deposition disease (HCDD) with fibrillary glomerulonephritis (FGN) is an exceedingly rare MGRS variant.

**Case Description:** A 67yo F with HTN and HFpEF presented with AKI on CKD & nephrotic syndrome. Evaluation revealed an IgG lambda monoclonal gammopathy with markedly elevated serum free lambda light chains (2742 mg/L, K/L ratio 0.00) and a minimal M-spike (0.09g/dL). Kidney biopsy demonstrated mesangial expansion and segmental sclerosis on light microscopy. IF showed mesangial and capillary wall staining for IgG (2–3+), lambda light chain restriction, and IgG1 subclass positivity, with C3 and C1q 1+ staining. EM revealed powdery electron-dense deposits along glomerular and tubular basement membranes with associated mesangial fibrillary deposits, consistent with HCDD with FGN. Bone marrow biopsy revealed 15–25% plasma cells without evidence of amyloidosis & with intermediate-risk cytogenetics. There was paucity of heavy chain production, but she still had heavy chain renal disease. She was started on daratumumab-based CyBorD therapy with improvement in K/L ratio to 0.86, Cr down from 2.6 mg/dL to 1.6 mg/dL and proteinuria down from 3.5g/g to 2.2g/g within 6 weeks.

**Discussion:** This case highlights a rare overlap of HCDD and FGN and underscores the discordance between hematologic burden and severity of renal involvement in MGRS. This case emphasizes the importance of timely kidney biopsy and early clone-directed therapy. Early recognition of rare MGRS variants is critical to guide prompt treatment and potentially preserve kidney function.



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Direct Mag: 25000 x  
AMT Camera System