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On the Cover

What is the diagnosis?

An 87-year-old male with a known history of hypertension and coronary artery disease presented with symptomatic anemia (Hgb 6.6 g/dL) and AKI with a serum creatinine of 8.4 mg/dL. He had microscopic hematuria and proteinuria of 2093 mg/g. Serum protein electrophoresis showed a faint M-spike, and immunofixation suggested IgG lambda monoclonal band. A serum free light chain assay showed a kappa/lambda ratio of 0.14, with a free kappa level of 57 mg/dL and lambda level of 394 mg/dL. Kidney biopsy showed lambda-restricted light chain crystalline cast nephropathy. Bone marrow workup subsequently revealed IgG lambda plasma cell myeloma comprising 60% of marrow cellularity. He required dialysis initiation and remains dialysis dependent to date, 3 months after his initial presentation.

Image Description:

Light microscopy showed acute tubular injury coupled with slightly pale eosinophilic (left image) and bright fuchsinophilic (middle image) intra-tubular geometric-shaped crystalline casts having sharp angulated edges. Some of these casts were associated with intense cellular reaction (as in left image) within the tubular lumina including occasional giant cells (not shown). Glomeruli were unremarkable. Electron microscopy showed tubular lumina filled with similar crystalline geometric casts (right image).

Teaching Points:

Light chain crystalline cast nephropathy shares etymological similarities with light chain cast nephropathy, but with important differences such as: (1) it is much rarer; (2) there is a predominance of lambda-restricted light chain casts; (3) crystalline casts can be seen in both proximal and distal tubules; and (4) morphologically, it presents as sharp multiple geometric-shaped crystalline casts within tubules. Treatment of light chain crystalline case nephropathy is not well defined and guided by case reports only, which encompass standard chemotherapy for multiple myeloma.

References:


(Images and text provided by Rajib K. Gupta, Ochsner-LSU Health, Department of Pathology and Translational Pathobiology, Shreveport, Louisiana; Rusella Mirza, Ochsner-LSU Health, Department of Pathology and Translational Pathobiology, Shreveport, Louisiana; and Narendra Goel, New Jersey Kidney Care, Jersey City, New Jersey)