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On the Cover

What's the diagnosis? Plasma cell dyscrasias may result in production of an abnormal immunoglobulin (paraprotein), with affinity to deposit in the kidney (amyloidosis or monoclonal immunoglobulin deposition disease) or to filter into the tubule, with consequent reabsorption by the tubular epithelium (light chain tubulopathy, crystal storing histiocytosis) or with intraluminal cast formation (cast nephropathy, myeloma kidney). Therefore, patients with multiple myeloma may have a variety of presentations of the kidney disease, but "myeloma kidney" specifically refers to cast nephropathy. Light chain casts may obstruct many tubules and are characterized by PAS-negative reaction, waxy texture and fractured appearance, and the presence of brisk inflammatory reaction of the surrounding tissue with multinucleated giant cells on light microscopy. Immunofluorescence studies show restriction to one of the light chains. In typical cases of cast nephropathy, electron microscopy confirms the absence of dense deposits in the tissue and shows very dark (electron dense) intraluminal cast material, with homogeneous texture and no substructural details. Occasionally, light chains will form unusual crystals within the lumen of the tubules, as shown on this electron micrograph. The crystals are intraluminal (extracellular), they are very electron dense, mostly needle-shaped or rod-like, but generally of variable shape and size. This variant of cast nephropathy with extraordinary and breathtaking morphological features is of no clinical relevance, as these patients usually present with typical features of cast nephropathy. (Image and text provided by Dr. Vanesa Bijol, Brigham and Women's Hospital)