Renal Involvement in Collagen Vascular Diseases and Dysproteinemias

**FIGURE 11-35 (Continued)**

B. Amyloid deposits in the renal arteries in a congo red-stained slide and viewed under polarized light. Amyloid has an apple-green color. C. Immunofluorescence. Amyloid deposits in the mesangium stained with anti-\(\alpha\) antibodies. (Panel A, methenamine silver. Original magnification \(\times550, \times350, \times400\), respectively.)

**FIGURE 11-36**

Light chain amyloidosis on electron microscopy. A. Characteristic fibrillar pattern of amyloid deposits. Long, randomly distributed, nonbranching fibrils with diameters of 8 to 12 nm. B. Amyloid fibrils in the capillary lumen and capillary wall with extension through the glomerular basement membrane (GBM) into the subepithelial space (arrow) fibrils arranged in parallel forming spicules. (Original magnification \(\times48,000, \times20,000\), respectively.)

**FIGURE 11-37** (see Color Plate)

Light chain deposition disease. In about 60% of patients with this renal lesion, nodular expansion of the mesangium is seen that resembles nodular diabetic nephropathy \(\{175,76\}\). The nodules stained purple with periodic acid-Schiff (PAS) stain have a homogeneous appearance, and those stained with methenamine silver are pink-brownish in color. In a few cases, a more mesangiocapillary pattern of injury is present. The tubular basement membranes (TBMs) are thickened, as seen in the PAS-stained sections. In the remaining cases, no renal lesions can be seen on light microscopy. On immunofluorescence, linear staining of basement membranes of glomeruli, tubuli, and vessels can be observed for one of the light chains (\(\kappa > \lambda\)). In most cases, the TBMs are more heavily stained than the glomerular basement membranes (GBMs). Congo red staining is negative for amyloid. On electron microscopy, fine granular electron-dense material can be found along the endothelial side of the GBM, in the mesangium, and along the interstitial side of the TBM. A few cases of heavy chain and of light and heavy chain deposition disease have been described, in most cases with identical morphologic characteristics as described in light chain deposition disease \(\{177,78\}\).

A. Nodular glomerulosclerosis with nodular increase of mesangial matrix. B. Linear staining of the GBM, mesangium, Bowman’s capsule, and TBM for the \(\kappa\) light chain.

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FIGURE 11-37 (Continued)

C and D. Electron-dense granular deposits in the GBM (C) and around the TBM (D). L—capillary lumen; Pod—podocyte. (Panel A, methenamine silver. Original magnification ×400, ×400, ×15,000, ×6500, respectively.)

FIGURE 11-38

Cast nephropathy. The casts have a homogeneous, fractured, or crystalline appearance with sharp angular or irregular edges and are present in the distal and collecting tubules [73]. These casts are composed of aggregated κ or λ light chains mixed with Tamm-Horsfall protein (THP). Sometimes the tubular cells show necrosis accompanied by disruptions of the tubular basement membrane (TBM). Proximal tubular cells show hyaline droplets or vacuoles with needlelike, tubular, or complex crystalline material. Casts are surrounded by macrophages and multinucleated giant cells. On electron microscopy, the casts have a granular, homogeneous, or fibrillary appearance with occasional needlelike crystals. The fibrils that surround the casts are probably THP. In most cases, a varying degree of interstitial fibrosis exists, accompanied by mononuclear cell infiltration and tubular atrophy. Congo red staining for amyloid is usually negative. The glomeruli are normal.

Fanconi's syndrome in a patient with λ light chain proteinuria. **A**, Vacuolization of proximal tubular epithelial cells. Vacuoles contain light-brown-colored material. **B**, Immunofluorescence. The granular material in tubular cells is stained for λ light chains. **C**, Low-power view of a proximal tubular epithelial cell with vacuoles containing organized or crystalline material. **D**, High-power view of the vacuoles containing tubular or ladderlike crystalline structures. BB—brush border. (Panel A, methenamine silver. Original magnification ×600, ×400, ×7000, ×19,000, respectively.)