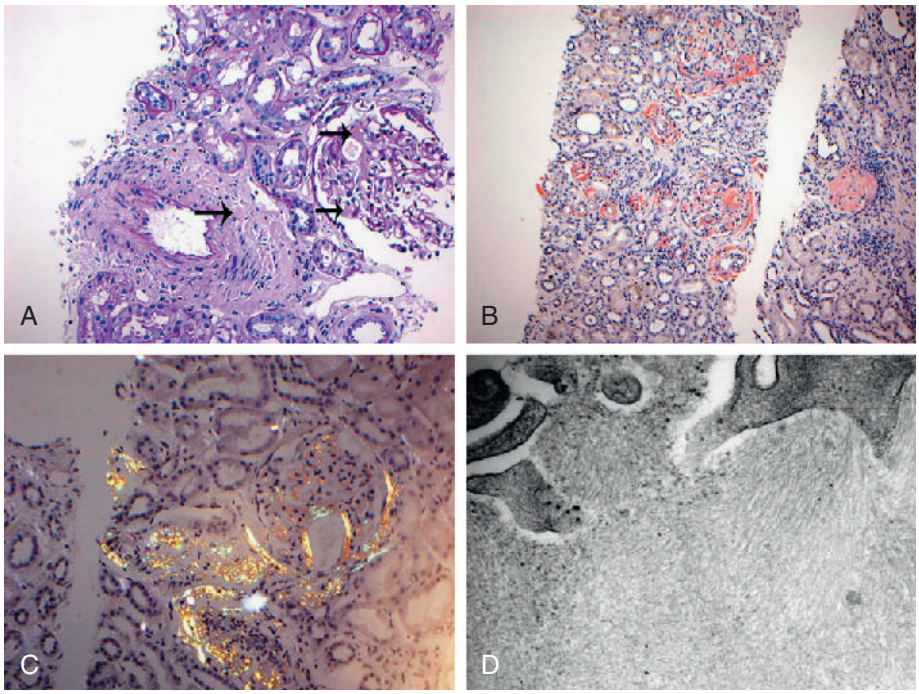

FIGURE 28-12 Amyloidosis. **A,** Light microscopy shows amyloid deposits characterized by mesangial expansion *(small arrows)* with material negative for staining. The material is also seen in vessel walls, where the *arrow* points to vascular deposits (periodic acid–Schiff stain, ×20). **B,** Congo red staining is positive for amyloid and shows reddish brown material in the glomeruli, interstitium, and vessel walls (×10). **C,** Amyloid deposits show apple green to orange-yellow birefringence under polarized light (×20). **D,** Electron microscopy shows randomly oriented amyloid fibrils. The fibrils measured 9 nm thick (×49,000).

common in patients with rheumatoid arthritis, inflammatory bowel disease, chronic infection, or familial Mediterranean fever. Treatment of AA amyloidosis is directed at the underlying inflammatory process with antimicrobials or anti-inflammatory medications.

Light Chain Deposition Disease

Light chain deposition disease is a paraprotein-associated disorder. The peak incidence is in the sixth decade of life, and men are affected more commonly than women. Approximately 30% to 50% of patients with light chain deposition disease have multiple myeloma. Most have a detectable monoclonal protein (usually κ light chain) in the serum or urine, but no hematologic abnormality is identified in about 10% of cases. Renal involvement manifests as proteinuria, and renal insufficiency is the most common initial presentation. Immunoglobulin deposits in other organs may result in myriad associated clinical symptoms.

Renal biopsy specimens show acellular, eosinophilic mesangial nodules that stain strongly positive with PAS, often mimicking diabetes mellitus. The deposited monoclonal proteins do not form fibrils and are Congo red negative. Immunofluorescence microscopic findings are diagnostic, with diffuse linear immunoglobulin light chain deposition (κ in 80% of cases) along the GBM and tubular basement membranes. On electron microscopy, amorphous, noncongophilic, monoclonal immunoglobulin proteins can be seen along the GBM (Fig. 28-13).

Encouraging results have emerged with the use of bortezomib and dexamethasone and with high-dose chemotherapy and autologous stem cell transplantation. Unless remission is achieved after chemotherapy, the disease will recur in the kidney allograft.

GLOMERULONEPHRITIS ASSOCIATED WITH HEPATITIS B VIRUS INFECTION

HBV-mediated glomerular disease usually manifests as membranous nephropathy, especially in children. The diagnosis of HBV-mediated glomerular disease requires detection of the virus in the blood and the exclusion of other causes of glomerular diseases.

HBV-mediated glomerular disease usually has a favorable prognosis, with a high spontaneous remission rate in children, but it is often progressive in adults. Patients with HBV infection and glomerulonephritis should receive treatment with interferon-α and/or with nucleoside analogues as recommended by standard clinical practice guidelines for management of HBV infection. Those with severe vasculitis or RPGN may be candidates for immunosuppressive therapy in combination with antiviral therapy. Rituximab treatment of patients who are positive for HBV has been associated with fatal acute hepatitis.

THROMBOTIC MICROANGIOPATHIES

Thrombotic microangiopathy is characterized by thrombocytopenia, microangiopathic hemolytic anemia, and microvascular occlusion, resulting in various degrees of organ dysfunction. Markers of hemolysis include low haptoglobin levels, increased levels of lactate dehydrogenase and unconjugated bilirubin, and a high reticulocyte count. Schistocytes are seen in peripheral blood smears.

The quintessential forms of thrombotic microangiopathy include hemolytic uremic syndrome (HUS) and thrombotic thrombocytopenic purpura (TTP). Although previously thought to represent different manifestations of the same disease, these disorders are distinct clinically and mechanistically. In adults,