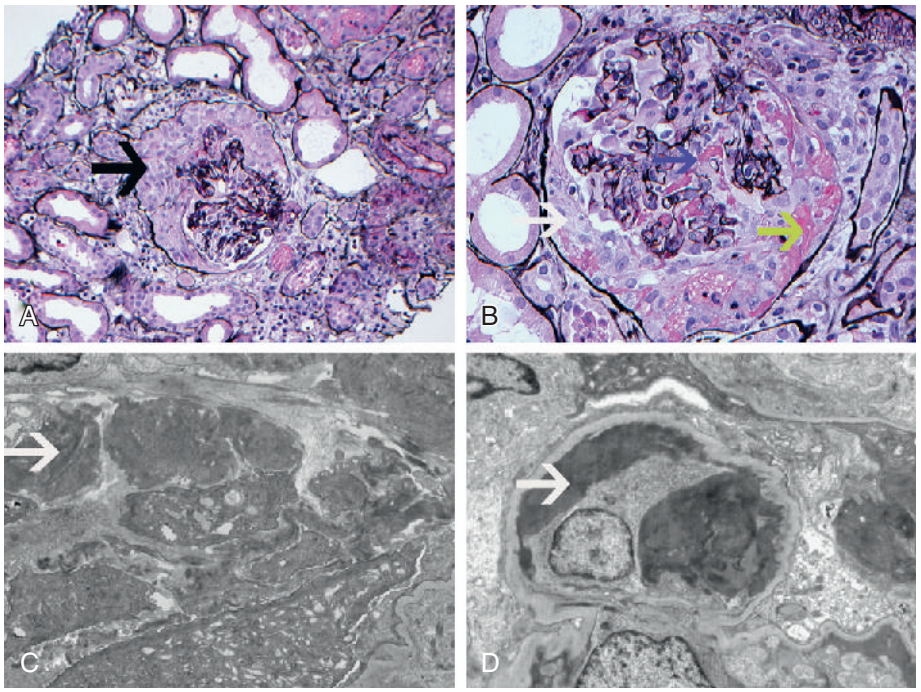

FIGURE 28-9 Crescentic glomerulonephritis. **A** and **B,** Light microscopy and silver methenamine staining show a large cellular crescent *(black arrow)* with fibrinoid necrosis *(blue arrow)*, hemorrhage into Bowman's capsule *(yellow arrow)*, and collapse of capillary tufts (**A,** ×20; **B,** ×40). **C** and **D,** Electron microscopy shows fibrinoid necrosis (i.e., necrotizing lesion) in Bowman's space *(white arrow)* and capillary loops *(short white arrow)* (both, ×11100).

TABLE 28-3 SIGNS AND SYMPTOMS OF ANTINEUTROPHIL CYTOPLASMIC AUTOANTIBODY VASCULITIS

Abdominal pain and gastrointestinal bleeding
Cutaneous purpura, petechiae, nodules, ulcerations, and necrosis
Facial pain, necrotizing (hemorrhagic) sinusitis, and septal perforation
Hematuria, proteinuria, and renal failure
Hemoptysis and pulmonary infiltrates or nodules
Muscle and pancreatic enzymes in blood
Myalgias and arthralgias
Peripheral neuropathy (mononeuritis multiplex)

Patients with newly diagnosed severe AAV vasculitis can be treated with a combination of high-dose corticosteroids and cyclophosphamide or high-dose corticosteroids and rituximab. Those with pulmonary hemorrhage, respiratory compromise, or severe renal failure (i.e., serum creatinine >5.5 mg/dL) also should undergo plasma exchange. The prognosis for AAV varies. Those with severe renal failure have the worst prognosis; and after successful therapy, AAVs have a relapse rate of 30% to 50% in the first 5 years. In some patients, rising ANCA titers are predictors of relapse. Patients with GPA or who are PR3-ANCA positive or presenting with relapsing disease are at higher risk for future relapses.

Goodpasture Disease: Anti–Glomerular Basement Membrane Antibody–Mediated Glomerulonephritis

Goodpasture disease is a pulmonary-renal syndrome (i.e., Goodpasture syndrome) caused by circulating anti-GBM antibodies. On immunofluorescence staining of biopsy specimens, a linear pattern is seen along the GBM and alveolar basement membrane (Fig. 28-10) using antibodies directed against the α3 chain of type IV collagen (COL4A3 protein). Patients usually have RPGN and various degrees of pulmonary hemorrhage.

The treatment of Goodpasture disease is based on high-dose pulse methylprednisolone (1 g/day for 1 to 3 days) followed by corticosteroids (prednisone, 1 mg/kg/day up to 80 mg daily) in combination with oral cyclophosphamide (2 to 3 mg/kg/day up to 200 mg daily, adjusted for age and creatinine level) and plasma exchange. The prognosis is predicted in part by the percentage of circumferential crescents on the renal biopsy specimen, oliguria, and the need for dialysis. Those with an initial serum creatinine level less than 5.0 mg/dL have a 90% probability of renal survival at 5 years; but those with 100% circumferential crescents and on dialysis do not recover renal function, and immunosuppressive regimens should be avoided except in the case of pulmonary hemorrhage.

Goodpasture disease rarely recurs. Patients with ESRD are candidates for renal transplantation after the antibody has disappeared (6 to 12 months).

Lupus Nephritis

Lupus nephritis occurs in up to 50% to 70% of patients with SLE and is associated with a poor prognosis. Proteinuria is the most common initial manifestation, and it is often in the nephrotic range and accompanied by a decline in renal function. Urinalysis does not always reflect the severity of the glomerular lesion, and kidney biopsy is indicated in those with proteinuria or active urinary sediment, or both, because the type of renal lesion