Nephritic Syndrome

The nephritic syndrome is defined by oliguria, edema, hypertension, proteinuria (usually <3.5 g/24h), and abnormal urinalysis with dysmorphic red blood cells or casts on microscopic examination.

Rapidly Progressive Glomerulonephritis

RPGN is a clinical syndrome characterized by progressive loss of kidney function with a time course of days to months in a patient with active urinary sediment such as red blood cell casts. Patients may have oliguria. Most of the pulmonary-renal syndromes manifest in this fashion, and the pathologic corollary is often a focal, necrotizing, crescentic glomerulonephritis. When RPGN is suspected, renal biopsy with immunofluorescence studies is extremely helpful.

Linear deposition of immunoglobulin G (IgG) points to Goodpasture disease or anti–glomerular basement membrane (anti-GBM)–mediated glomerulonephritis. Immunoglobulins and complement suggest systemic lupus erythematosus (SLE), cryoglobulinemia, immunoglobulin A nephropathy (IgAN), or postinfectious glomerulonephritis. Negative or weak immunofluorescence (pauci-immune) findings usually indicate an antineutrophil cytoplasmic autoantibody (ANCA) vasculitis (Fig. 28-1).

GLOMERULAR DISEASES MANIFESTING WITH NEPHROTIC SYNDROME

Minimal Change Disease

In a patient with nephrotic syndrome, minimal change disease (MCD) is defined by a renal biopsy with no significant glomerular abnormalities on light microscopy, negative immunoglobulin and complement deposition on immunofluorescence, and widespread foot process effacement on electron microscopy (Fig. 28-2). MCD is the most common cause of nephrotic syndrome in children and accounts for up to 20% of adults with primary nephrotic syndrome.

The pathogenesis of MCD is unknown. The association with Hodgkin's lymphoma suggests that MCD may be a consequence of T-lymphocyte abnormalities, with T cells producing a lymphokine that is toxic to glomerular epithelial cells. Most cases of MCD are idiopathic, although drugs (e.g., NSAIDs), hematologic malignancies (mainly Hodgkin's lymphoma), and thymoma are well-recognized causes of secondary MCD. Concomitant interstitial nephritis suggests drugs (e.g., NSAIDs) as the likely cause of MCD.

In children, MCD usually manifests with nephrotic syndrome of acute onset. Hematuria, hypertension, or impaired renal function is unusual and suggests another diagnosis. When nephrotic syndrome occurs in a child with normal urinalysis results, the

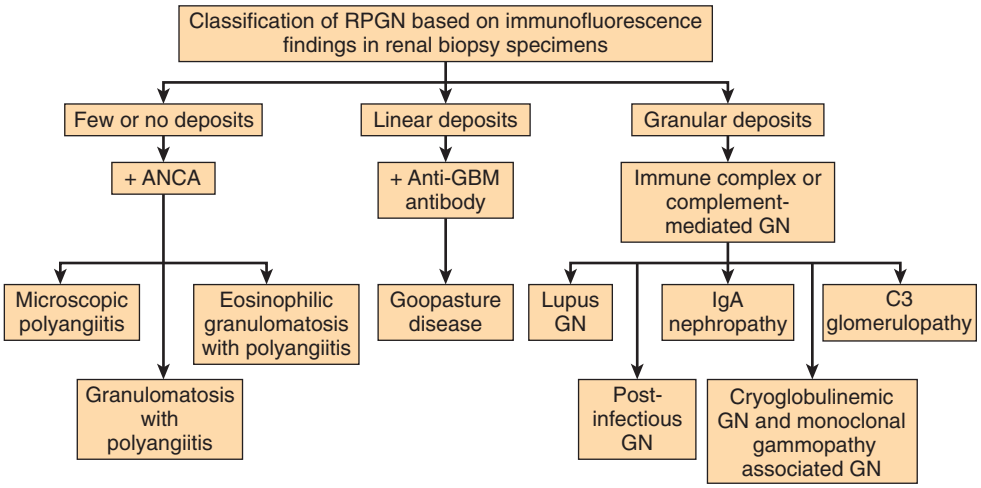

FIGURE 28-1 Rapidly progressive glomerulonephritis (RPGN) is classified according to immunofluorescence microscopy findings in renal biopsy specimens. ANCA, Antineutrophil cytoplasmic autoantibody; GBM, glomerular basement membrane; GN, glomerulonephritis; IgA, immunoglobulin A.

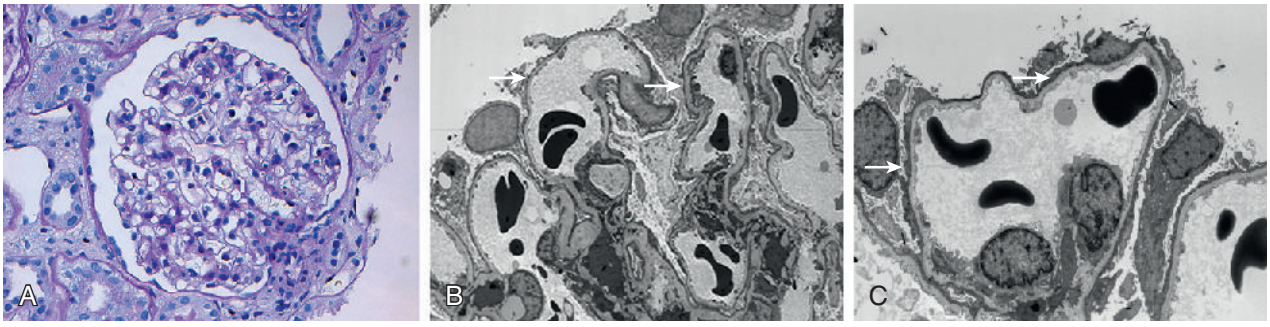

FIGURE 28-2 Minimal change disease. **A,** Light microscopy shows a normal-appearing glomerulus (periodic acid–Schiff, ×40). **B** and **C,** Electron microscopy shows diffuse foot process effacement *(arrows)* (**B,** ×2500; **C,** ×4200). Immunofluorescence studies were negative for immune deposits.