anemia and leukocytosis, mild hypergammaglobulinemia (particularly of the IgA class), and mildly elevated rheumatoid factor. Thrombocytosis may be seen as an acute-phase reactant. Approximately 90% of patients with active granulomatosis with polyangiitis have a positive antiproteinase-3 ANCA. However, in the absence of active disease, the sensitivity drops to ~60–70%. A small percentage of patients with granulomatosis with polyangiitis may have antimyeloperoxidase rather than antiproteinase-3 antibodies, and up to 20% may lack ANCA.

Patients with granulomatosis with polyangiitis have been found to have an increased incidence of venous thrombotic events. Although routine anticoagulation for all patients is not recommended, a heightened awareness for any clinical features suggestive of deep-vein thrombosis or pulmonary emboli is warranted.

DIAGNOSIS

The diagnosis of granulomatosis with polyangiitis can be established by the demonstration of necrotizing granulomatous vasculitis on tissue biopsy in a patient with compatible clinical features. Pulmonary tissue offers the highest diagnostic yield, almost invariably revealing granulomatous vasculitis. Biopsy of upper airway tissue usually reveals granulomatous inflammation with necrosis but may not show vasculitis. Renal biopsy can confirm the presence of pauci-immune glomerulonephritis.

The specificity of a positive antiproteinase-3 ANCA for granulomatosis with polyangiitis is very high, especially if active glomerulonephritis is present. However, the presence of ANCA should be viewed as adjunctive with tissue diagnosis being pursued when clinically inconsistent features are present or when ANCA is absent. False-positive ANCA has been reported in certain infectious and neoplastic diseases.

In its typical presentation, the clinicopathologic complex of granulomatosis with polyangiitis usually provides ready differentiation from other disorders. However, if all the typical features are not present at once, it needs to be differentiated from the other vasculitides, antiglomerular basement membrane disease (Goodpasture's syndrome) **(Chap. 314)**, relapsing polychondritis