Table 21.10.2.2 Classification of rapidly progress

Table 21.10.2.2 Classification of rapidly progressiveglomerulonephrit is according to renal immunofluorescence andcirculating immune reactants

21.10.2 The kidney in systemic vasculitis 4991 are present in granulomata and at sites of vasculitic injury. They have specificity for ANCA autoantigens and demonstrate features of af- finity maturation. Autoantibodies to endothelial antigens are found in over 50% of patients with vasculitis, but their targets have not been defined and their contribution to pathogenesis is unclear. Role of infection The involvement of the respiratory tract in GPA has led to interest in the interaction between respiratory tract infection and a dysregulated immune response in the epithelium. Colonization with Staphylococcus aureus is associated with a higher relapse rate, and bacterial strains ex- pressing toxic shock staphylococcal toxin are implicated. Damage to the

respiratory tract resulting from vasculitic inflammation impairs its ability to eradicate microbial infection and a cycle of vasculitis and recurrent infection develops. Cytokine-induced up-regulation of endothelial adhesion molecules promotes leucocyte adhesion and in- jury, providing an additional mechanism by which inflammation sec- ondary to infection can stimulate vasculitis. Epidemiology The incidence of AAV is 15 to 20/million population per year, with prevalence rates of 200 to 250/million. The incidence is similar between Europe and Japan, but there is phenotypic variation with a predomin- ance of MPA with MPO-ANCA in Japan and China and an even distri- bution of GPA and MPA with PR3 and MPO-ANCA in Europe. There is a paucity of epidemiological data from Southern Asia or black people of African descent. A latitudinal gradient with GPA predominating in colder, temperate climates has been reported in both hemispheres. Both GPA and MPA are very rare in children and have an increasing incidence with age, with a mean age at diagnosis of GPA of 55 years, and MPA at 10 years older. Renal involvement is very common in MPA, occurring in 90%, and in 70% of GPA and 15% of EGPA cases. Renal function at diagnosis is worse in older patients, indicating that not only is renal involvement more frequent, but also more aggressive. Renal vasculitis is less frequent in Takayasu's arteritis at 0.2/million per year, with polyarteritis nodosa and anti-GBM disease at 1/million per year each. IgA vasculitis is much more common in children, but when it occurs in adults, renal disease is frequent and the outcomes more severe. Cryoglobulinaemia is typically associated with hepatitis C infection, but idiopathic forms with renal involvement are fewer than 1/million per year. Clinical features Patients with primary systemic vasculitis vary in their prodromal features, in the pattern and severity of organ involvement, in their response to therapy, and in their subsequent disease course and prognosis. A high index of suspicion is required to make the diag- nosis in any scenario where there is suspected nephritis or un- explained chronic inflammation (Box 21.10.2.1). Clinical and laboratory evaluation confirms the extent and severity of organ involvement, which is used to guide therapy (Table 21.10.2.2). Treatment aims to recover renal function and to obtain and sustain disease remission, but relapses are common and refractory disease or chronic, persisting low disease activity states are therapeutic challenges. Box 21.10.2.1 Clinical features that should raise suspicion of vasculitis Features of nephritis • Haematuria with proteinuria, with or without impaired renal function Features of chronic inflammation, otherwise unexplained • Constitutional disturbance: — Polymyalgia, polyarthralgia, flitting polyarthritis — Fatigue, malaise, weight loss, fevers, night sweats • Ear, nose, and throat: — Nasal obstruction and epistaxis — Recurrent sinusitis — Deafness — Facial pain • Eye: — Episcleritis/scleritis — Corneal ulcer — Retinal vein thrombosis — Visual loss • Lung: — Haemoptysis — 'Maturity-onset' asthma, chronic breathlessness — 'Antibiotic-resistant pneumonia' — Respiratory failure — Subglottic or endobronchial stenosis • Heart: — Pericarditis — Aortic valve disease, aortitis • Skin: — Nailfold infarction (splinter haemorrhages) — Purpura — Nonhealing ulcer • Nervous system: — Cranial or peripheral neuropathy (sensory or motor) — Cerebral mass lesion(s) — Myelitis Table 21.10.2.2 Classification of rapidly progressive glomerulonephritis according to renal immunofluorescence and circulating immune reactants Type Renal immunofluorescence Compatible serology Diagnosis I Linear IgG Anti-GBM antibodies Anti-GBM disease II Granular IgG/IgA/IgM ANA/anti-dsDNA/low C3/4 Systemic lupus erythematosus Granular IgG/IgA/IgM Low C3/4 Postinfectious glomerulonephritis Granular IgG/IgA/IgM Low C3/4 Mesangiocapillary glomerulonephritis Granular IgG/IgA/IgM Low C3/4, cryoglobulins Cryoglobulinaemia Granular IgA None IgA vasculitis (Henoch–Schönlein purpura) III Pauci-immune (absent or scanty deposits) ANCA ANCA-associated vasculitis (GPA, MPA, or EGPA) ANA, antinuclear antibodies; ANCA, antineutrophil cytoplasm autoantibodies; dsDNA, double-stranded DNA; GBM, glomerular basement membrane.

Revision #1
Created 2026-01-22 16:42:15 UTC by Omar Ayman
Updated 2026-01-22 16:42:15 UTC by Omar Ayman