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Pauci-immune glomerulonephritis

INSERM

Source

INSERM. (1999). *Orphanet: an online rare disease and orphan drug data base. Pauci-immune glomerulonephritis. ORPHA:93126*

Pauci-immune glomerulonephritis (GN) is one of the most frequent causes of rapidly progressive GN (RPGN, see this term). It is characterized clinically by renal manifestations of RPGN (hematuria, hypertension) leading to renal failure within days or weeks, and may be associated with manifestations of systemic vasculitis (arthralgia, fever, seizures, mono neuritis and lung involvement). Pauci-immune GN is histologically characterized by focal necrotizing and crescentic GN, with mild or absent glomerular staining for immunoglobulin and complement by fluorescence microscopy, which may manifest either as part of a systemic small vessel vasculitis (including microscopic polyangiitis, granulomatosis with polyangiitis and eosinophilic granulomatosis with polyangiitis (see these terms)), or rarely as part of renal-limited vasculitis (RLV, idiopathic crescentic GN). Immunologic classification is based on the presence or absence of circulating anti-neutrophil cytoplasmic antibodies (ANCAs), namely pauci-immune-GN with ANCA and pauci-immune GN without ANCA (see these terms).