Antineutrophil cytoplasmic antibody (ANCA) associated lung-renal vasculitides: A single centre perspective

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Body: Introduction Small to medium vessel vasculitides are a rare cause of multi-organ failure. Methods: This is a single centre retrospective review of systemic vasculitides, with respiratory involvement. Results: 23 patients (14 Male; 9 Female, mean age 45 (Range 14 to 70) years), presented with vasculitis and significant respiratory involvement, including 17 with ANCA+ vasculitis, 3 ANCA-Neg, and 3 with Churg Strauss Syndrome (CSS). 2 patients had isolated airways disease, 4 systemic non-organ threatening disease, and 14 generalised disease, 7 requiring ventilation and renal replacement. 15 of 20 patients had a biopsy procedure including nasal, skin, bronchial and renal biopsy. 2 patients with ANCA+ vasculitis and 3 with CSS were diagnosed clinically. 16 of 20 patients recieved corticosteroids (CS) and cyclophosphamide induction. Maintenance therapy included CS and Azathioprine or Mycophenolate Mofetil. Patients presenting with diffuse alveolar haemorrhage or renal failure received plasma exchange. Rituximab was reserved for patients intolerant or not responding to Cyclophosphamide, or serious relapses, including cerebral vasculitis. Tracheal stenosis (n=1) required recurrent balloon dilatation. Cavitative pulmonary disease, pneumothorax and aspergillus disease, responded to CS, iv Immunoglobulin and Rituximab. 16 patients relapsed requiring further induction treatment. There were 3 deaths, 1 renal transplant, 1 lung transplant, and 1 end stage renal failure on haemodialysis. The majority of patients are asymptomatic on low dose immunosuppression. Conclusions: Pulmonary vasculitis requires a high index of suspicion to ensure prompt diagnosis and treatment to avoid end organ damage.