

PAUCIIMUNE CRESCENTIC GLOMERULONEPHRITIS IN
ASSOCIATION WITH MYELOMA CAST NEPHROPATHY -A
PARANEOPLASTIC VASCULITIS.

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Abstract

The incidence of paraneoplastic vasculitis in patients with multiple myeloma is exceptional (0.8%) and the association with pauciimmune cresenteric glomerulonephritis has never been reported. Malignancy in general and paraprotein in myeloma specifically act as an antigenic factor playing a key role in mediating vascular damage. The triggering event of an initial ANCA response could be a synergistic infection or the inflammatory process priming neutrophils and monocytes and setting the stage for development of small vessel vasculitis. Proteinase 3 (PR-3) a target antigen for ANCA is identical to myeloblastin, a growth promoting protein from myeloid cells and myeloblastin mRNA is expressed in cells from patients with acute myeloid leukemia involved in differentiation of human leukemic cells which could be involved in Wegener's Granulomatosis and leukemogenesis. We report the first case of advanced renal failure with c-ANCA positivity, IgG kappa monoclonal gammopathy, hypocomplementemia, renal biopsy proven pauciimmune cresenteric glomerulonephritis with features suggestive of myeloma cast nephropathy. Bone marrow biopsy showing 10% plasma cells. We further describe favorable clinical response of renal failure to cyclophosphamide and prednisone. Vasculitis associated myeloma indicates additional poor prognostic sign and cyclophosphamide induced remission comes at a heavy price – infection, sepsis. Plasmapheresis best compliments to immunosuppressants in advanced renal disease. Ability to prospectively distinguish patients likely to relapse opens the possibility to target more effective and safe immunosuppressive therapy to those at higher risk.

