

INTERSTITIAL NEPHRITIS AS THE ONLY RENAL LESION WITH C-ANCA POSITIVE WEGENER'S DISEASE- AN UNUSUAL RENAL PRESENTATION

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A 54 year old woman was admitted with one week history of bodyaches, right eye redness and joint pain. In the past one year patient reports having recurrent bilateral otitis media and mastoiditis requiring placement of bilateral myringotomy tubes. She also reported recurrent episodes of "pyelonephritis". She was found to have microscopic hematuria on urinalysis and proteinuria of 1 to 1.5 g/day. Her renal function was normal. Cytoplasmic Antineutrophilic Antibodies (C-ANCA) obtained as part of the work up of the urinalysis findings was positive in high titers (46 U/ml, Normal < 6 U/ml), ESR was 124. A previous biopsy of the nasal mucosa showed chronic nonspecific inflammation without evidence of necrotizing granulomas. A clinical diagnosis of Wegener's disease was made and a diagnostic renal biopsy was performed which showed acute and chronic interstitial nephritis, tubulitis and mild to moderate tubulointerstitial scarring. Glomeruli were unremarkable. She was started on Prednisone **1 mg/kg/day** and Cyclophosphamide **125 mg/day orally**. Her proteinuria improved to less than 300 mg/day **after 4 months of treatment**. C-ANCA titers became negative (<6 U/ml) and the ESR normalized. ANCA mediated neutrophil activation has been implicated to play a role in pauci immune vasculitis and interstitial nephritis (Weidner et al, Arth & Rheum, Nov 2004, 50: 11)

Interstitial changes typically occur along with a segmental necrotizing glomerulonephritis in patients with ANCA associated Vasculitis (AAV). However, isolated interstitial involvement without glomerular disease is rare. This case illustrates an atypical renal presentation of Wegener's disease with good response to therapy.