

Abstract

Wegener's Granulomatosis (WG), also known as Granulomatosis with Polyangiitis (GPA), is one of the Antineutrophil Cytoplasmic Antibodies (ANCA) associated small vessel vasculitides, strongly associated with cANCA. We report a rare presentation of WG in a 72 year old female with cANCA negativity. She was admitted with recurrent nasal congestion, symptoms of sinusitis, and saddle nose deformity. Her laboratory studies revealed high inflammatory markers, elevated serum creatinine, and microscopic haematuria. Her serum tested negative for cANCA and positive for pANCA. A computerized Tomography (CT) scan of her sinuses showed pansinusitis. Her renal biopsy revealed crescentic pauci-immune glomerulonephritis. A diagnosis of WG was made based on the clinical and laboratory findings. She was treated with steroids and cyclophosphamide.⁽¹⁾ cANCA is present in 80-90% of cases.⁽²⁾ We present a case of WG in a patient who tested positive for pANCA but negative for cANCA.