Microscopic polyangiitis presenting as predominantly pulmonary disease with mild renal symptoms and quiescent RPGN: a case report

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INTRODUCTION
• Vasculitis is defined by presence of inflammatory leukocytes in blood vessel walls
• Classification based on size of vessels involved (Figure 1)
  - Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) includes granulomatosis with polyangiitis (GPA), eosinophilic granulomatosis with polyangiitis, microscopic polyangiitis (MPA)
  - AAV incidence 3.3 per 100,000, occurs in older adults
  - MPA typical clinical manifestations include non-specific symptoms: fever, malaise, anorexia, weight loss, myalgias, and arthralgias
  - Prodromal symptoms last weeks to months without evidence of specific organ dysfunction
  - Organ specific dysfunction EXT, pulmonary, kidney, cutaneous, and neurological involvement
  - Evident glomerulonephritis present in only 18% of patients at presentation, but develops in 80% in first two years

PRESENTATION
A 69-year-old male with PMH of HTN, T2DM, s/p CABG in 2021 presented to outside hospital with increasing SOB. Symptoms began one month, described as a non-painful "out of breath" feeling that originally noted with activity, now effecting patient at rest. Patient has additionally experienced generalized fatigue, night sweats, and several months of weight loss (20+lbs in four-month period). Denies sputum production, travel history and recent hospitalizations. ROS was remarkable for for an episode of a retroorbital headache and temporary jaw claudication for four weeks, which he was prescribed a course of antibiotics for a presumed sinus infection, which did not improve symptoms, pt subsequently started on 60 mg prednisone, Notably, patient endorses a specific finding with obvious cause +/- specific organ involvement
• Early ANCA testing is critical to support the diagnosis of AAV
• This specific discrepancy between clinical phenotype and ANCA values only occurs in 10–20% of patients
• Case highlights how AAN represents a spectrum of diseases with overlapping presentations necessitating a thorough history-taking and timely rheumatologic workup
• Tissue biopsy vital to diagnosis and can be performed on kidney even with limited renal symptoms
• Creatine and other markers not a perfect surrogate for disease staging

DISCUSSION:
• AAV should be considered in patient with non-specific findings with obvious cause and +/- specific organ involvement

Figure 7: Kidney biopsy showing pauci-immune crescentic glomerulonephritis showing remarkable fibrinoid necrosis surrounded by crescent formation

REFERENCES: