ANCA Negative Pauci-Immune Crescentic Glomerulonephritis Associated with Rheumatoid Arthritis: A Rare Case

Jose Aliling MD, Ronald Miick MD, Ruchika Patel MD
Division of Rheumatology
Einstein Medical Center
Philadelphia, PA
Case Presentation

A 53-year-old African-American Female came in with swelling of hands, wrists, knees, feet, and ankles.

- She has history of hypertension who came in with new onset of synovitis of bilateral hands, wrists, knees, ankles, and feet worsening over the last 2 months associated with bilateral lower extremity edema.
Physical Exam

• VS: normal

• HEENT:
  – (-) nasal discharge (-) sinus tenderness (-)
    tonsillopharyngitis (-) cervical lymphadenopathy

• CHEST
  – clear breath sound

• CVS
  – Normal rate and rhythm, (-) murmurs (-) gallops

• Abdomen:
  – (-) tenderness, (-) organomegaly
Physical Exam

• Tenderness and swelling of both wrists and MCP and PIP joints of the 2nd to 4th digit of both hands with no gross deformity
• Tenderness and synovitis of both knees with minimal effusion and with decreased knee flexion
• Tenderness and synovitis of both ankles and MCP joints of all digits of both feet
• Bilateral pitting edema of the lower extremities (grade 2-3)
Laboratory Data

- CBC: normocytic, normochromic anemia (hemoglobin of 9.9 gm/dl)
- Creatinine of 1.85 mg/dl (baseline 1.26)
- High titer RF and anti-CCP
- ANA of 1:80 with a negative specific serology
- C3 and C4 were normal
Laboratory Data

- ESR was 124 and the CRP was 3.66 mg/dl
- Urinalysis proteinuria with active urinary sediments with RBC casts 0-1/hpf.
- Urine protein/creatinine ratio was 2 grams of protein
- Urine toxicology was negative
- ANCA testing, anti-MPO, anti-PR3, and anti-GBM were all negative
Diagnostic Data

• Kidney biopsy: crescentic GN affecting 8 of 12 glomeruli with two additional glomeruli which were globally sclerosed.
• A moderate lymphoplasmacytic chronic interstitial nephritis was also identified; vasculitis was not present.
• Immunofluorescence was negative for IgG, IgA, IgM, kappa, lambda, C3 and C1q.
• Electron microscopic findings demonstrated crescent formation with no dense deposits identified.
Kidney Biopsy

Glomerulus with cellular crescent formation consisting of parietal epithelial cells and macrophages surrounding the glomerulus; the glomerular basement membrane is highlighted by the silver stain. PAMS stain, 400x.
Diagnosis

• The biopsy findings are consistent with a pauci-immune crescentic glomerulonephritis despite the negative ANCA serologies.
Treatment

• The patient was subsequently treated with high dose prednisone and monthly IV Cyclophosphamide with improvement of serum creatinine to baseline and reduction of proteinuria.
Discussion

• Renal involvement in RA is highly unusual with most cases being related to complications of therapy and not to the disease itself.

• Most common forms of renal disorders in RA patients are usually glomerular disease (membranous glomerulopathy and mesangial proliferative glomerulonephritis), amyloidosis, and tubulointerstitial lesions.\(^3,4\)

• Few cases of crescentic GN that have been associated with RA have all been positive for the p-ANCA antibody in the setting of systemic vasculitis\(^5\).

•
Discussion

- We report a patient with RA who presented with high disease activity with concomitant acute kidney injury due to ANCA negative crescentic GN.
- Renal involvement in RA is rare and the need for a kidney biopsy should not be delayed to aid in the diagnosis and prompt initiation of appropriate therapy to prevent further deterioration of renal function which may lead to irreversible damage.\(^6\)


