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

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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 the 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

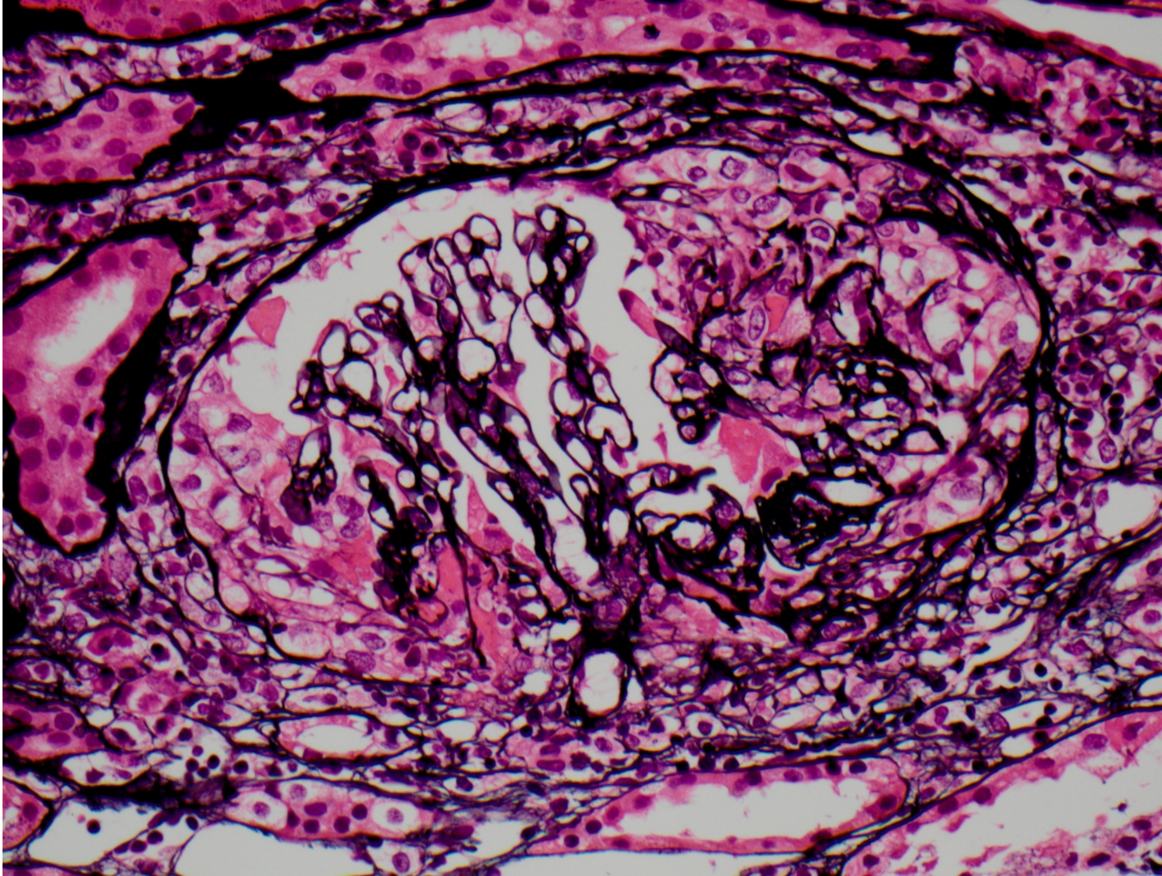
Laboratory Data

- 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].
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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]

References

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