UNDETECTED ANCA-ASSOCIATED VASCULITIS: A CASE OF MIXED NEPHRITIC-NEPHROTIC SYNDROME CULMINATING IN DIALYSIS IN AN OLDER WOMAN

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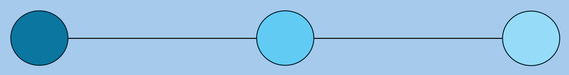
INTRODUCTION

- Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) is a rare autoimmune disorder causing inflammation of small blood vessels, often affecting the kidneys, lungs and heart.
- AAV may go undetected for years, before a sudden flare causes severe organ failure.
- We present a case of AAV masked for years by hypertensive cardiomyopathy, unveiled only by a severe renal crisis requiring permanent dialysis, highlighting the need for heightened autoimmune awareness in multimorbid patients.

TIMELINE

2022
- CHF symptoms
- LVH, low EF
- ANA+, IgM elevated

2025
- AKI (Cr 23.5)
- Nephritic-nephrotic
- Dialysis started



2022-2024
- Recurrent symptoms
- Pericardial effusion
- Pulmonary HTN

WHY THIS CASE MATTERS
- Demonstrates how multimorbidity can hide rare autoimmune disease, delaying diagnosis
- Risk of anchoring bias
- Need for re-evaluation when clinical course deviates from expected progression

CASE PRESENTATION

- 62-year-old African American woman with HTN, CAD, HFpEF, AF (s/p ablation), obesity, and CKD stage 3 (baseline Cr 1.0)
- Presented with 3 weeks of worsening dyspnea (NYHA 3-4), edema, confusion, and tremors
- On examination: confusion, dry mucous membranes, 2+ edema of lower limbs, clear lungs
- CT chest: concern for aspiration pneumonia and right lung nodules
- Labs showed severe AKI (Cr 23.5 mg/dL), initially attributed to hypovolemia
- UA: heavy proteinuria (>2000mg/dL), hematuria, leukocyturia revealing a mixed nephritic-nephrotic picture
- Hemodialysis was initiated due to renal failure, and workup for intrinsic renal disease began.

Prior clues:

- In 2022, patient had dyspnea/edema attributed to hypertensive CHF
- Echo revealed LVH, EF 35%, and pericardial effusion
- CT: ground-glass opacities, pleural effusions
- Cardiac MRI ruled out ischemic/infiltrative disease
- Right heart cath: precapillary pulmonary HTN
- Labs: ANA positivity + elevated IgM (these incidental findings were not worked up)

Definitive autoimmune workup done:

- Autoimmune panel:
  p-ANCA +
  dsDNA +
  low C3
- Renal biopsy: pauci-immune ANCA-associated vasculitis with necrotizing crescentic glomerulonephritis and C3 dominant deposits.
- Diagnosis: SLE-AAV overlap syndrome

- Treatment: IV steroids and rituximab, but renal function did not improve.
- Outcome: progression to ESRD requiring permanent dialysis

DISCUSSION

- ANCA-associated vasculitis (AAV) is a small-vessel autoimmune disease that can present subtly and mimic common conditions.
- This case highlights diagnostic delay due to attribution of symptoms to hypertension and heart failure.

Early warning signs:
- Pulmonary hypertension
- Recurrent pericardial effusions
- Positive ANA and abnormal immunologic markers

- Overlapping features of SLE and AAV complicated the clinical scenario.
- Mixed nephritic-nephrotic syndrome should have prompted early autoimmune workup.
- Delays in diagnosis contributed to the outcome of irreversible renal damage and ESRD.

CONCLUSION

- AAV can remain clinically silent for years before presenting with severe organ damage, as it has in this case.
- Early recognition of
  Unexplained renal dysfunction
  Pulmonary hypertension
  Autoimmune markers

  Is critical for timely diagnosis.
- Delayed diagnosis may lead to irreversible outcomes such as ESRD. Clinicians should maintain a low threshold for autoimmune screening in atypical or progressive cardiorenal disease.