A RARE CAUSE OF RAPIDLY PROGRESSIVE GLOMERULONEPHRITIS

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INTRODUCTION

- Hydralazine induced ANCA vasculitis (HIAV) is a small vessel vasculitis that typically presents with multiorgan system involvement sometimes including severe renal and pulmonary impairment
- On literature review, there have only been 4 previously reported cases of HIAV that were limited to renal involvement[1]
- Here, we present a 5th case of HIAV that displayed renal limitation

INITIAL PRESENTATION

- A 65-year-old male presented to the hospital with fatigue, weight loss, and shortness of breath for the past three months

Past Medical History:
- HTN, CKD II, CAD, and Microscopic Hematuria

Medications:
- Aspirin 81 mg, Carvedilol 25 mg twice a day, and Hydralazine 100 mg three times a day

Social History:
- Jehovah's witness

Physical Exam:
- Vitals: 98.6 F, 126/71 mmHg, 65 beats per minute, 23 respirations per minute, 97% on room air
- Chronic psoriatic rash over lower extremities

Pertinent Laboratory Findings		
Measurement (units)	Result	Normal Range
WBC (K/mcL)	4.0	4.2 - 11.0
HGB (g/dL)	8.4	13.0 - 17.0
BUN (mg/dL)	65	6 - 20
Creatinine (mg/dL)	7.22	0.67 - 1.17
Urinalysis Erythrocytes (cells/hpf)	>100, few dysmorphic RBCs	None Seen, 1 to 2
Urinalysis Protein (mg/dL)	30	Negative
Urine Protein/Creatinine Ratio (mgPR/gCR) (done 2 days prior to arrival)	1060	<=199
Blood Culture x2	No growth for 6 days	No growth
ESR (mm/hr)	101	0-20
CRP (mg/dL)	1.6	<1.0
ANA Titer	Positive, 1:1280	Negative
ANA Pattern	Homogenous	N/a
MPO (AI)	Positive, >8.0	<1.0
PR3 (AI)	Positive, >8.0	<1.0
C3 (mg/dL)	64	79-152
C4 (mg/dL)	13.9	16.0-38.0

HOSPITAL COURSE

- Hydralazine was stopped and added to allergy list
- Patient completed pulse dose steroids then transitioned to PO
- Patient completed six sessions of plasmapheresis and four rituximab infusions
- Patient's kidney function improved and was discharged
- A kidney biopsy was deferred due to bleeding risk, low platelet count, platelet dysfunction from aspirin, low fibrinogen levels due to plasmapheresis, and patient's religious contraindication to transfusion

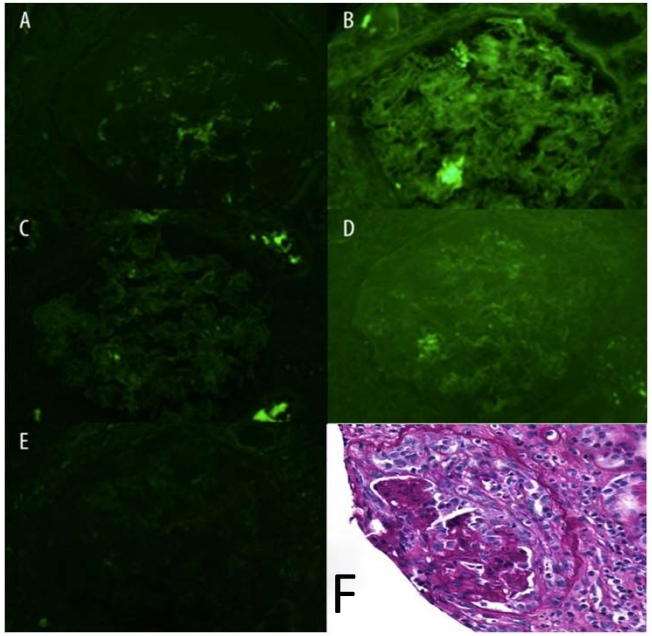

Figure: Images A-E depict immunofluorescence of a kidney biopsy stained with IgM statin, IgG statin, IgA statin, C3 statin, and C1q statin respectively. Notice the minimal signal throughout these photos as there is little immune complex deposition in HIAV. Image F shows a light microscopy slide from a separate patient displaying a cellular crescent. Of note, these pictures were not from our patient. They are just a representation of characteristic findings for a patient with HIAV.

Images A-E obtained from:
https://pubmed.ncbi.nlm.nih.gov/33993184/
Image F obtained from:
https://www.pathologyoutlines.com/topic/kidneyANCArelatedgngen.html

LABORATORY TRENDS

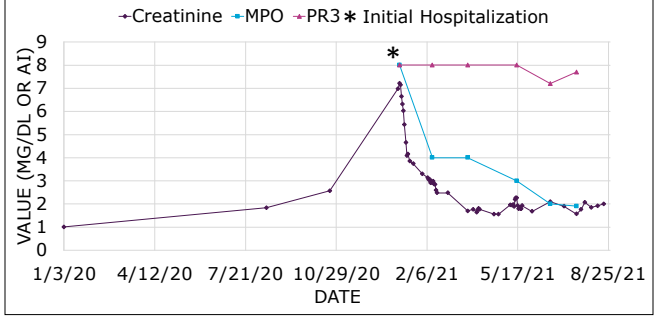

Figure 2: Kidney function baseline prior to hospitalization compared to labs during and after hospitalization. Response to treatment was seen as creatinine returned to near baseline and MPO and PR3 decreased. Of note, the first three PR3 values were >8.0 and the fourth was exactly 8.0 limiting representation of improvement in PR3 in this case.

DISCUSSION

- HIAV presents with constitutional symptoms but may impact many systems including musculoskeletal, cutaneous, pulmonary, and renal
- Diagnosis is made primarily by serology which will often show positive ANA, MPO, PR3, occasional anti dsDNA, low complement levels, and kidney biopsy showing crescentic glomerulonephritis with minimal immune complex deposition[2]
- Treatment includes cessation of hydralazine and initiation of immunosuppression in severe presentations
- Hydralazine induced lupus is often misdiagnosed but differs by usually not presenting with severe renal impairment, will be negative for MPO and PR3, and positive for anti-histone antibody
- Aggressive immunosuppressive therapy without kidney biopsy was initiated in our patient due to degree and rate of renal impairment in the setting of a likely diagnosis

CONCLUSION

- HIAV has a diverse presentation, including a renal limited form, for which greater awareness will result in proper treatment including cessation of hydralazine and initiation of immunosuppressive therapies

REFERENCES

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2. Grau, R.G. Drug-Induced Vasculitis: New Insights and a Changing Lineup of Suspects. *Curr Rheumatol Rep* 17, 71 (2015). https://doi.org/10.1007/s11926-015-0545-9