EXTREMELY RARE CASE OF ATYPICAL HUS INDUCED DIFFUSE RENAL CORTICAL NECROSIS

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Introduction

• HUS: Intravascular hemolysis, thrombocytopenia, and AKI
• Atypical HUS causing renal cortical necrosis is extremely rare.
• Diffuse cortical necrosis is very rare in developed countries, only 1-2% of AKI cases
• Usually irreversible.
Case Presentation

- 69-year-old female with a PMHx of breast cancer s/p left mastectomy
- Cough, watery diarrhea, fatigue and rash of 3 days
- Productive cough and chest congestion for 2 weeks
- Recently treated with prednisone
- Vital signs: Stable
- Diffuse maculopapular rash
Investigations:

• WBC 35K, creatinine 0.77 (GFR >90)
• U/A: hematuria, proteinuria, and pyuria.
• Lactic acid 2.6.
• CXR and CT chest: Concerning for Pneumonia
• Ceft + Azithromycin >> Moxifloxacin
Hospital Course

• Day 2: AKI and anuria
• Creatinine 4.55 (GFR 11) on day 3 and 7.72 (GFR 5) on day 6

• Negative ANA with Reflex and ANCA (MPO/PR3).
• Elevated LDH (2173), low C3 (20) and C4 (2.5)
• Haptoglobin, platelet, and HG normal and no schistocytes
• Direct Coombs, reticulocyte count, ADAMTS 13 activity normal.
• Renal US: Normal
• Atypical HUS genetic test positive
• Cobalamin C deficiency associated mutation
Renal Biopsy

• Extensive cortical coagulative necrosis with vascular fibrin deposit consistent with diffuse cortical necrosis.
Management

• Steroids
• Hemodialysis
• Hem Onc, Neph, Rheum, Geriatrics, ID
• Patient was discharged with outpatient HD(MWF).
Discussion

• This case demonstrates aHUS without hemolytic anemia or thrombocytopenia causing irreversible renal cortical necrosis.
• She has genetic predisposition to aHUS precipitated by severe sepsis/infection causing diffuse renal cortical necrosis.
• aHUS is not caused by infections or coexisting disease.
• Infections are considered as triggers not as a cause of disease
Treatment

• Steroids, Plasma exchange, Immunosuppressive therapy, Transplant
• Eculizumab: mAb against C5
• Hemodialysis
THANK YOU!
References

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