Acute Kidney Injury and Hemophagocytic Lymphohistiocytosis (HLH) caused by Human Granulocytic Anaplasmosis (HGA)

Grace Chong DO
Lehigh Valley Health Network, Grace.Chong@lvhn.org

Wael Hanna MD
Lehigh Valley Health Network, wael_a.hanna@lvhn.org

Joseph Scurozo
Lehigh Valley Health Network, joseph.scuorzo@lvhn.org

Tayyab Ali MD
Lehigh Valley Health Network, Tayyab.Ali@lvhn.org

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Acute Kidney Injury and Hemophagocytic Lymphohistiocytosis (HLH) caused by Human Granulocytic Anaplasmosis (HGA)

Grace Chong, DO[1], Wael Hanna, MD[2], Joseph Scuorzo, DO[1], Tayyab Ali, MD[2]

[1.] Department of Medicine, Lehigh Valley Health Network, Allentown, PA, United States
[2.] Department of Nephrology, Lehigh Valley Health Network, Allentown, PA, United States

INTRODUCTION

HLH is a hyperinflammatory syndrome characterized by unregulated macrophage and T-lymphocyte activation resulting in cytokine overproduction, hemophagocytosis, and tissue destruction. It can be either primary, due to a genetic etiology, or secondary, caused by infection & hematologic malignancy, and autoimmune disease. AKI has been shown to occur in up to 62% of cases of HLH. HGA is a tick-borne illness characterized by flu-like symptoms, cytopenias, and transaminitis.

CASE PRESENTATION

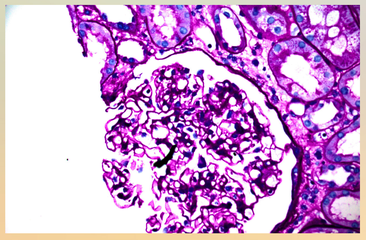
- A 62-year-old male with a history of DM2 presented with fever and vomiting. Labs showed acute renal failure, thrombocytopenia (platelet count 16K/cmm) without schistocytes, and elevated liver enzymes. CT scan of the abdomen displayed splenomegaly. Urinalysis showed proteinuria and microhematuria without any casts.
- Serologies for ANA, ANCA, anti-GBM, and complements were normal. A renal biopsy was obtained and empiric doxycycline was started for possible tick-borne illness.
- Serum ferritin was found to be highly elevated to 12K ng/ml which prompted concern for HLH. Triglycerides & soluble CD25 were high at 659 mg/dl & 16230 IU/ml (normal <2400) respectively.
- Bone marrow aspirate did not show hemophagocytosis, however, he met diagnostic criteria for HLH so dexamethasone was started.
- Anaplasma phagocytophilum IgM titers came back elevated at 1:256 (normal <1:16). Renal biopsy revealed mesangial hypercellularity associated with mesangial C3 deposits, a picture suggestive of infection-related GN (IRGN).
- The patient showed rapid clinical and renal improvement after initiation of treatment with doxycycline and was discharged home in stable condition.

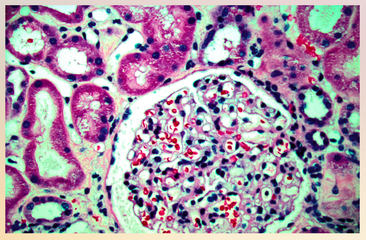
LABS

Blood Count		
Hgb	13.4	
WBC	2.3	L
Platelet count	16	L
CMP		
BUN	72	H
Creatinine	5.44	H
Sodium	131	L
Potassium	3.7	
Chloride	93	L
Carbon Dioxide	21	L
Anion Gap	17	H
GFR	10	L
Phosphorus	4.3	
Albumin	2.2	L
Calcium	7.1	L
Protein, total	5.6	L
Bilirubin, total	6.2	H
Bilirubin, direct	5.5	H
AST	181	H
ALT	103	H

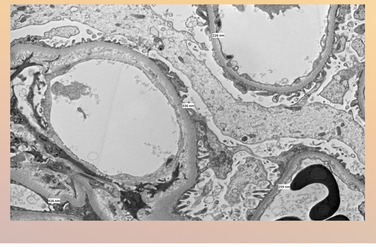
Urinalysis		
Specific Gravity	1.012	
pH	5	
Protein	30-70	I
Glucose	Negative	
Ketone	Negative	
Blood	>or=1.00	I
Leukocytes	Negative	
Nitrite	Negative	
WBC/HPF	Rare	
RBC/HPF	21-50	
Amorphous Material	Present	
Squamous Epithelial Cells	>10	
Hyaline Casts	0-2	
RBC Casts	3-5	
Urine Chemistry		
Creatinine	77.6	
Sodium	23	L
Potassium	24.2	
Chloride	<10	
Protein, Total	134.9	H
Protein/Creatinine Ratio	1.74	H

NATIVE KIDNEY BIOPSY





Light microscopy: Minimal mesangial hypercellularity, associated with mesangial C3 deposits. There is no evidence of endocapillary proliferation, crescents, or necrotizing lesions. Most suggestive of post-infection glomerulonephritis.



Electron microscopy: No significant ultrastructural findings. There is no evidence of electron dense, immune complex-type deposits or protein deposition disorder.

DISCUSSION

In the course of HLH, kidney damage has been reported to be related to ischemic lesions, high levels of nephrotoxic tumor necrosis factor and or underlying infection or malignancy. Mesangial proliferative GN pattern is reported in 8% of all IRGN cases. In our case, AKI was attributed to HGA itself and secondary HLH.

Thorough history with high clinical suspicion for possible tick-borne illness led to the initiation of correct treatment within 24 hours of presentation. This resulted in improvement in AKI and allowed the patient to avoid unnecessary potential toxic effects of chemotherapy for HLH.

CONCLUSION

Early identification and initiation of treatment of the underlying trigger for AKI and HLH can result in a greater chance of renal recovery.

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