51-Year-Old Female with Epistaxis and Generalized Weakness for 2 Months Diagnosed with RPGN

Meihuan Lin MD R3

Background

Rapidly progressive glomerulonephritis (RPGN)'s clinical presentation including acute onset of macroscopic hematuria, decreased urine output, acute onset of hypertension, and edema. Other clinical findings include nephritis presentation on urine analysis with proteinuria, micro and macroscopic hematuria, red blood cells, dysmorphic RBC and RBC casts. Pathological findings characterized extensive crescent formation. Untreated patients can rapidly progress to end-stage kidney disease over a very short periods (days to months). Renal biopsy can further differentiate the types of crescentic glomerulonephritis.

Objective

To highlight the clinical, laboratory and histologic studies that support the diagnosis of RPGN and management options to improve clinical outcome.

Case Presentation:

Patient is a 51-year-old female with history of hypothyroidism and iron deficiency anemia presented with nosebleed and generalized weakness for 2 months. Her baseline creatine was 0.6 back in 2022. On admission, her creatine was 17.9 along with hyperkalemia which required urgent hemodialysis. Serological studies and renal biopsy were done and confirmed with Pauci Immune Glomerulonephritis with positive P-ANCA, negative anti-GBM antibodies. The patient was treated with IV methylprednisolone 1 gram daily. Hem/Onc was consulted, and patient was transferred to another facility for emergent chemotherapy.

Vitals: Temp: HR 86, RR:18, BP:163/93, SaO2: 100% (Room Air)
Physical examination: Positive for CVA tenderness/ back pain.

Results

Pertinent Laboratory Studies

Admission Creatinine and eGFR	BUN 134/ CR 17.9 eGFR 2
Anti-Nuclear Antibody (ANA)	Negative
Serum complement	C3 117 (normal) C4 35 (normal)
UR Eosinophil	High
Anti-Glomerular Basement MRBN AB	<0
Urinalysis	UA GLUCOSE < 50 , UA BILIRUB... < 1 , UA KETONES < 5 , UA SPEC GR... 1.015 , UA BLOOD >=250 H, UA PH 5.0 , UA PROTEIN 100 H, UA UROBILI... < 1 , UA NITRITE Negative , UA LEUK ES... < 25 , RBC >100 H, WBC 16-25 H, BACT Few H, SQUAM CELL... 16-25 H, BUDDING YE... Few
DNA Double strand AB	1 (normal 0-9)
C-ANCA	<1:20
Anti-MPO	>8.0
P-ANCA	1:640 (high)
KIDNEY, CORE NEEDLE BIOPSY:	
- NECROTIZING AND CRESCENTIC GLOMERULONEPHRITIS WITH LOW-GRADE MESANGIAL IgG DEPOSITS (SEE COMMENT). - DIFFUSE ACUTE TUBULAR INJURY WITH FOCAL TUBULOINTERSTITIAL NEPHRITIS. - ARTERIOSCLEROSIS, SEVERE. - INTERSTITIAL FIBROSIS AND TUBULAR ATROPHY, MODERATE (30%).	

Discussion

There are three major types of RPGN: 1. Anti-glomerular basement membrane (GBM) disease is caused by anti-GBM antibodies. It is also associated with pulmonary components with cross-reaction of the antibodies with membrane of the pulmonary capillaries. 1. Immune complexes RPGN associated with immune complexes in the glomeruli such as IgA deposits, antistreptococcal antibodies and subepithelial humps, and antinuclear antibodies and subepithelial deposits in lupus nephritis. 3. Pauci immune RPGN is associated with small vessel vasculitis, a necrotizing GN with few or negative immune deposits. It is often antineutrophil cytoplasmic antibody (ANCA) positive.

An urgent diagnosis is important in the patient who is presenting with clinical findings of RPGN. Serologic tests such as a ANCA, anti-BGM antibodies, ANA, and others with biopsy results. Prompt diagnosis with kidney biopsy and serological test with early therapy is the key to minimize the kidney injury. Specific treatment can be given once the diagnosis is made.

Conclusion

Prompt diagnosis with kidney biopsy and serological test with early therapy is the key to minimize the kidney injury.

References

Couser WG. Rapidly progressive glomerulonephritis: classification, pathogenetic mechanisms, and therapy. Am J Kidney Dis 1988; 11:449.

Falk RJ, Hogan S, Carey TS, Jennette JC. Clinical course of anti-neutrophil cytoplasmic autoantibody-associated glomerulonephritis and systemic vasculitis. The Glomerular Disease Collaborative Network. Ann Intern Med 1990; 113:656.

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