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Original Research Paper

General Medicine

CASE REPORT: A RARE CASE OF COLLAGENOFIBROTIC GLOMERULOPATHY IN A 45-YEAR-OLD FEMALE

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ABSTRACT Collagenofibrotic glomerulopathy (CG) is a rare renal disorder characterized by the accumulation of type III collagen in the glomeruli. This report presents the case of a 45-year-old female with CG, detailing her clinical presentation, diagnostic workup, and response to treatment.

KEYWORDS :

INTRODUCTION

Collagenofibrotic glomerulopathy is a rare renal disease marked by the deposition of type III collagen in the glomerular mesangial and subendothelial areas. First described by Arakawa in 1979, it was classified by the World Health Organization in 1995. It can manifest sporadically in adults or as a familial form in children. Typical presentations include nephrotic-range proteinuria and microscopic hematuria, with progressive renal function decline leading to end-stage kidney disease. Diagnosis is based on renal biopsy findings, including electron microscopy and immunohistochemical assays specific for type III collagen.

Case Presentation

A 45-year-old perimenopausal female, recently diagnosed with hypertension, hypothyroidism, and anemia, presented with bilateral lower limb swelling and facial puffiness for 15 days. Her medical history was negative for diabetes and significant family history.

Physical Examination

- Conscious and oriented
- Blood Pressure: 170/100 mmHg
- Pulse Rate: 80 bpm
- Respiratory Rate: 20 cpm
- Oxygen Saturation: 94% on room air
- Pallor present
- Bilateral pitting pedal edema up to the knees
- No icterus, cyanosis, clubbing, or lymphadenopathy

Systemic Examination:

- Cardiovascular: Normal heart sounds, no murmurs
- Respiratory: Normal vesicular breath sounds, no adventitious sounds
- Abdominal: Soft, non-tender, no organomegaly
- Neurological: No functional deficits

Investigations

- Hemoglobin: 10.3 g/dL
- Platelets: 1.74 lakhs
- White Blood Cells: 8260
- Serum Creatinine: 1.3 mg/dL
- Urine Protein: 4+
- Urine RBC: 15-20 cells
- Urine Protein Creatinine Ratio: 5.3 g/day
- Complement levels: Normal
- Virus Screen: Negative for HIV, HBsAg, and anti-HCV
- Renal Biopsy: Showed characteristic findings of collagenofibrotic glomerulopathy, including type III collagen deposits.

Management

The patient was started on Ramipril 5 mg/day, Telmisartan 40 mg/day, and Atorvastatin 20 mg/day. At the six-month follow-up, partial remission of proteinuria (reduced from 5.3 to 2.8 g/day) was noted, along with controlled blood pressure (140/80 mmHg) and stable renal function (serum creatinine 1.09 mg/dL, eGFR 75 mL/min).

DISCUSSION

Type III collagen is typically absent in normal glomeruli but can be seen in certain glomerular diseases. Collagenofibrotic glomerulopathy is distinct from conditions like nail–patella syndrome due to its unique clinicopathologic features. Histopathology shows mesangial expansion and PAS-negative material in the capillary loops, with electron microscopy revealing characteristic type III collagen fibers.

CONCLUSION

Collagenofibrotic glomerulopathy is a rare and poorly understood renal disease with limited treatment options focused on managing symptoms. Further research is essential to uncover the pathogenesis and develop targeted therapies to improve patient outcomes.

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