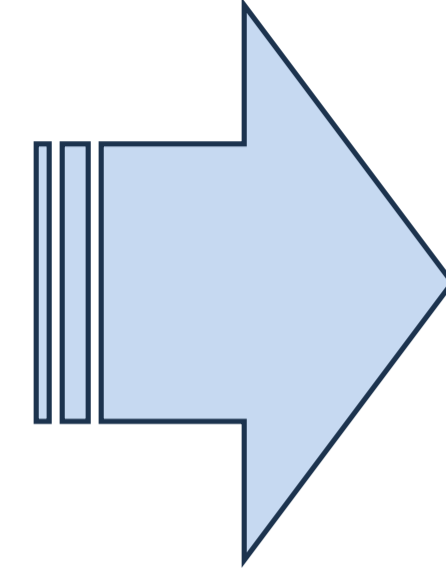


Introduction

Proliferative Glomerulonephritis with Monoclonal Immunoglobulin Deposits (PGNMID) is :

- ❖ Rare kidney-limited disease within the spectrum of MGRS.
- ❖ **Clinical presentation:** proteinuria, impaired renal function, sometimes nephrotic syndrome; acute nephritic syndrome uncommon.
- ❖ **Diagnosis:** kidney biopsy showing **glomerular deposits of monotypic immunoglobulins**.
- ❖ **Distinctive feature:** circulating monoclonal protein or B-cell clone identified in only ~30% of cases.



Implication: therapeutic decision-making remains **challenging** due to limited clonal identification.

References

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- Gumber R, Cohen JB, Palmer MB, et al. A clone-directed approach may improve diagnosis and treatment of proliferative glomerulonephritis with monoclonal immunoglobulin deposits. Kidney Int. 2018;94(1):199–205.
- Nasr SH, Sethi S. PGNMID: update on diagnosis and management. Clin Kidney J. 2021;14(1):28–37.

Cases

We report two young patients diagnosed with proliferative glomerulonephritis with monoclonal immunoglobulin deposits (PGNMID), both presenting with a membranoproliferative glomerulonephritis (MPGN) pattern and monotypic light-chain deposits, in the absence of detectable circulating monoclonal protein or hematologic clone.

The first patient, a 28-year-old man, presented in September 2021 with nephrotic syndrome. Laboratory investigations revealed proteinuria of 7 g/24 h, anemia (hemoglobin 8 g/dL), and hypoalbuminemia (18 g/L), with preserved renal function. Kidney biopsy showed MPGN with C3 deposits and lambda light-chain restriction. All etiological investigations were negative. Despite the absence of an identifiable clone, the patient received four cycles of bortezomib, cyclophosphamide, and dexamethasone (VCD), achieving complete renal remission with a progression-free survival of 54 months.

The second patient, a 32-year-old man, presented in November 2022 with severe renal failure (eGFR 13 mL/min/1.73 m²), requiring dialysis. Kidney biopsy demonstrated MPGN with C3 deposits and kappa light-chain restriction. Hematologic evaluation did not identify any monoclonal protein or clonal disorder and an extensive etiological workup was negative. Treatment with four cycles of VCD resulted in no clinical response.

Discussion

PGNMID

- Rare entity at the interface of nephrology and hematology.
- Diagnostic challenge: absence of circulating monoclonal protein or identifiable clone → “renal-limited” disease.
- Case observations:
 - * Patient 1 (preserved renal function) → complete durable remission after Bortezomib.
 - * Patient 2 (advanced renal failure, dialysis) → primary refractoriness despite similar histology.
- Key insight: timing of intervention and baseline renal function are critical determinants of outcome.
- Early treatment: may reverse renal injury before irreversible damage. Late treatment: limited efficacy in advanced disease.

Conclusion

Diagnostic & therapeutic challenge.

Clinical heterogeneity.

Treatment response variability