

A RARE CASE OF MGRS - PGNMID WITH LIGHT CHAIN RESTRICTION ONLY

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INTRODUCTION

- Proliferative glomerulonephritis with monoclonal immunoglobulin G deposition (PGNMID) is characterized by glomerular monoclonal IgG subclass deposition with a single light chain subtype.
- It is a rare disease with an autologous kidney biopsy rate of 0.17% to 0.21%.
- PGNMID as such has only 30% clone detection rate while the subtype of PGNMID with light chain restriction only was found to have higher clone detection rates.

CASE REPORT

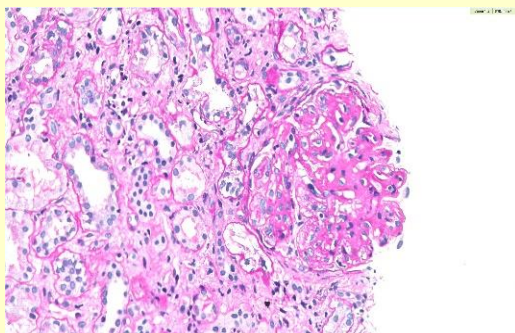
- A 60-year-old male patient presented with progressive swelling of lower limbs, shortness of breath and decreased urine output since 1 week. On examination he had bilateral pitting pedal edema and pallor was present.

LAB INVESTIGATIONS

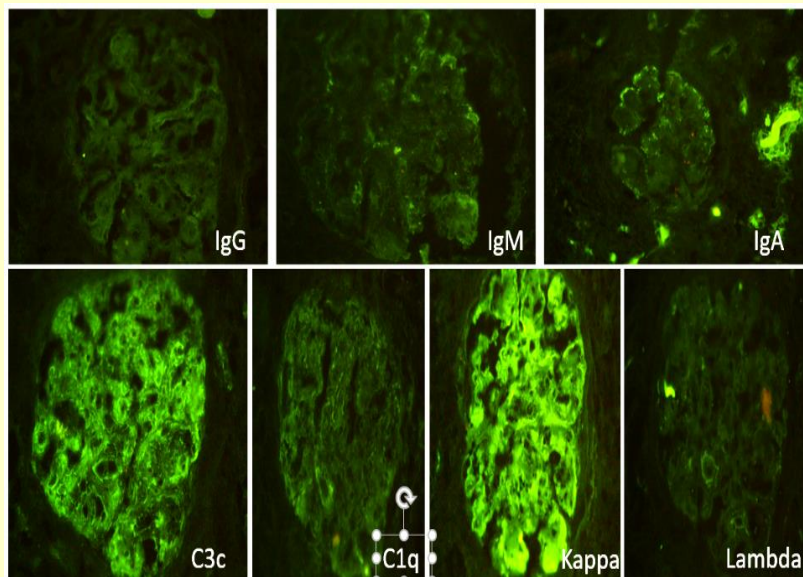
- Hemoglobin 8.9 gm/dl
- Serum urea - 151mg/dl
- Serum creatinine - 7.7 mg/dl
- Serum albumin - 2.9 gm/dl.
- Urine routine examination – RBC – 10-12 cells, Protein 4+
- USG - Kidneys were normal size, CMD maintained

RENAL BIOPSY:

- LM - Significant peripheral and mesangial coarse granular deposits of C3c and kappa light chains with insignificant deposits of IgA, IgM, C1q.
- IF - Deposits of kappa light chain and C3c was found to be present.
- Diagnosis - Proliferative glomerulonephritis with monoclonal immune deposits - light chain type (PGNMID-LC) with evidence of chronicity with global glomerulosclerosis (10/14) and mild IFTA (20%).



- The underlying pathogenic plasma cell clone could not be identified despite all the workup hence the patient was started on empirical chemotherapy for a hypothetic plasma/B cell clone.



CONCLUSION

- LC only variant of PGNMID is generally associated with a high detection rate of pathogenic plasma cell clone and has a good prognosis
- Monoclonal Immunoglobulin is identified by serum and urine IF in 65% and 73 % respectively, with abnormal serum free light chain in 83% and detectable BM plasma cell clone in 88%.
- Our patient continued to be on dialysis in spite of the empirical chemotherapy