A Case of Necrotizing IgA Nephropathy Presenting as Rapidly Progressive Glomerulonephritis in a Young Adult

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INTRODUCTION

- IgA nephropathy (IgAN) is the most common GN globally, but glomerular necrosis is uncommon.
- Necrotizing IgAN represents a severe variant with accelerated functional decline.
- This case describes a case of necrotizing IgAN without crescents, highlighting the diagnostic uncertainty due to limited guidance.

CASE PRESENTATION

- A 32 year old man with long standing hypertension and hyperlipidemia was referred for incidental creatine elevation to 2.8 mg/dL from a baseline of 0.9 mg/dL.
- Labs showed BUN 48 mg/dL, eGFR 30 mL/min/1.73 m2, albumin 3.9 g/dL. Urinalysis revealed protein 300 mg/dL, 2+ blood, RBC 60/HPF Serologies were negative except for a low-titer ANA. Renal ultrasound showed increased cortical echogenicity.
- Renal biopsy revealed mesangial expansion with a necrotizing lesion with segmental sclerosis (Oxford MEST C score was M1, E0, S1, T1, C0), and IFTA 40%, consistent with necrotizing IgA nephropathy with chronic fibrosis and no evidence of crescent formation. (**Figure 1**)

FIGURE

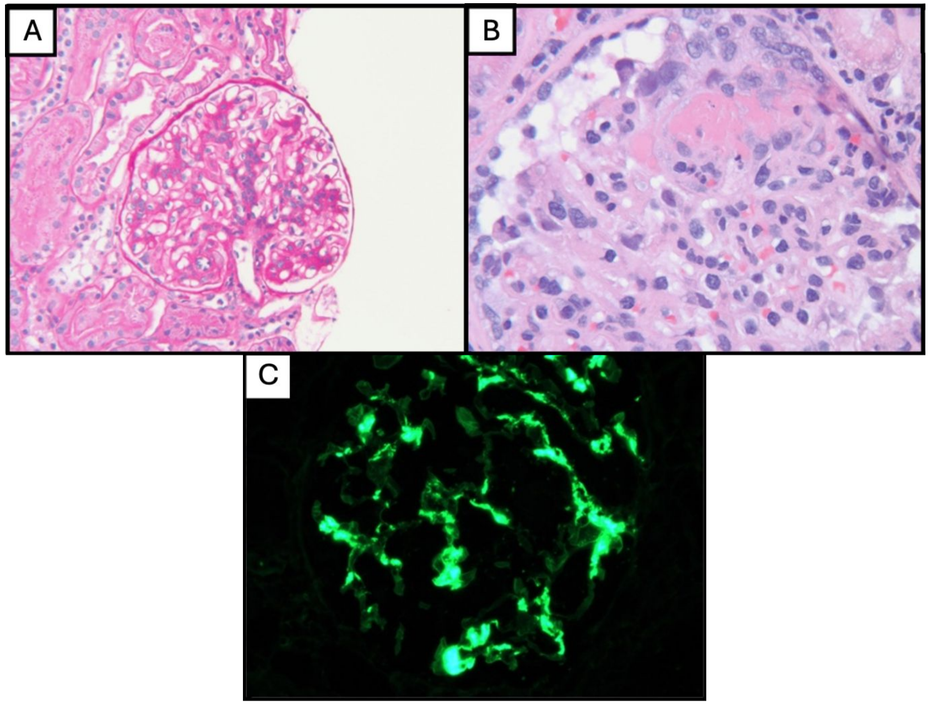

Figure 1: Renal biopsy demonstrating: **(A)** PAS stain and **(B)** H&E stain with mesangial expansion and a necrotizing lesion with segmental sclerosis **(C)** Immunofluorescence with IgA mesangial staining.

DISCUSSION

- Necrotizing lesions are rare in IgAN and signify active fibrinoid injury associated with rapid functional decline.[1]
- Dedicated treatment guidelines are lacking though these findings often lead clinicians to consider aggressive therapy.[2]
- In the absence of systemic features or crescents, superimposed vasculitis is unlikely, yet the necrotizing focus indicates a more aggressive phenotype than typical chronic IgAN.
- Early recognition and individualized management are crucial.

CONCLUSION

- Necrotizing IgAN is rare, poorly characterized, and lacks therapeutic guidance.
- It is important to consider early biopsy to identify and individualize management in the setting of limited evidence-based guidelines.

REFERENCES

1. Tan, Min, et al. Outcomes of IgA Nephropathy with Segmental Glomerular Necrosis but Without Crescent Formation. Am J Nephrol 43.5 (2016): 341-347.
2. Gleeson PJ et al. IgA Nephropathy in Adults—Treatment Standard. Nephrol Dial Transplant (2023): 2464-2473.