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A case of rare MGRS associated renal lesion

Ruimin Hu
The First Affiliated Hospital of Zhengzhou University, China

Monoclonal gammopathy of renal significance (MGRS), was introduced by the International Kidney and Monoclonal Gammopathy Research Group (IKMG) in 2012 and refined and updated in 2017, to describe any B-cell or plasma-cell clonal disorder that does not fulfill the criteria for cancer yet produces a nephrotoxic monoclonal immunoglobulin that leads to kidney injury or disease. MGRS associated renal lesions are classified by the presence or absence of monoclonal immunoglobulin deposits on IF and further subcategorized by EM into organized categories (fibrillar, microtubular and inclusions or crystalline) and non-organized.

Here, we report a case of 32-year-old Chinese man diagnosed MGRS by renal biopsy, with history of ankylosing spondylitis for 6 years, and treated by adalimumab for about 20 weeks 2 years earlier. Laboratory examination: Urinalysis: protein(+++), RBC 14/HPF(0-5), WBC 462.80/HPF, glucose(±). Urine protein:14.445 g/24h. Serological examination: WBC 6.40×10^9/L, RBC 3.27×10^{12}/L, Hb 95g/L, PLT 198×10^9/L, BUN 13.33 (2.9-8.2) mmol/L, Scr 251.00 (44-97) μmol/L, UA 271μmol/L, ALB 19.9 g/L, GLB 20 g/L, CHOL 4.78 mmol/L, TG 0.99 mmol/L, GLU 4.40 mmol/L. Electrolyte: K^+ 4.5 mmol/L, Na^+ 145.8 mmol/L, Cl^- 110.4(96-108) mmol/L, Ca^{2+} 1.81 (2.0-2.5) mmol/L. ESR 68 mm/h, CRP 0.5 mg/L,C3 0.60 g/L, C4 0.20 g/L,CFH 382.3(247-1010.8) ng/ml, CFB 286.2 (100-400) ng/ml , CFI 55.8 ng/ml, ADAMTS13 1203.5 pg/ml, TSH 3.96 mIU/l, FT3 4.29 pmol/L, FT4 11.81 pmol/L. ANA(-), anti-dsDNA (-), pANCA (-), MPO (-), cANCA (-), PR3 (-), anti-GBM (-), HLA-B27(+), RF 3.5 IU/ml. ASO 78.5 IU/ml, HBV(-), HCV(-), HIV (-). M proteinemia screening: IgA 110.00 (82-453) mg/dL, IgM 52.6 (46-304) mg/dL, IgG 318.00 (751-1560) mg/dL, FLCκ387 (639-1350) mg/dL, FLC λ 123.00(313-723) mg/dL, κ/λ3.15, urine Bence-Jones protein(-), SPEP (-). Renal biopsy revealed MPGN, immunofluorescence microscopy (IF) identified positive staining for κ light chain (2+) and C3 (2+) in the capillary wall and mesangium, and confined to glomeruli only, while no staining was observed for IgG, IgA and λ light chain. IgM, C4 and C1q were all trace. IF staining for IgG, IgA, C3, C1q, κ, and λ on paraffin tissue after proteinase K digestion was performed, showing κ light chain (2+) and C3 (2+), while IgG, IgA,C1q, and λ all negative. On electron microscopy (EM), granular electron-dense deposits were observed in subepithelial, subendothelial, mesangial and intramembranous, without substructure.