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A Case of IgG4-related Tubulointerstitial Nephritis (TIN) Mimicking Renal Infarction

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Introduction: Immunoglobulin G4-related disease is a systemic immune-mediate disorders shared particular clinicopathologic feature. Tubulointerstitial nephritis, most common form of IgG4-related kidney disease, present characteristic radiologic findings may be confused other renal disease. We report a case of IgG4 related tubulointerstitial nephritis, misdiagnosed as a renal infarction.
Case presentation: A 51-year-old chronic pancreatitis patient was consulted to the nephrology because of decreased eGFR to 29.6mL/min/1.73m^2. On computed tomography, multifocal cortical low attenuation lesions with atrophic change in both kidneys were demonstrated. However, renal arteriography revealed no abnormal finding in both main renal artery and intrarenal arteries. Laboratory findings disclosed: ANA positive, complement C3: 70mg/dL, C4: 3mg/dL, serum IgG4: >340mg/dL, Anti-phospholipid antibody IgM/IgG: 162.7/18.2U/mL, BUN/Cr: 23/2.22mg/dL. Renal biopsy was performed. Glomeruli near the subcapusular area show ischemic change. The tubules show severe atrophy, focal degeneration and regenerative change with neutrophilic and lymphocytic infiltration. The interstitium is diffusely expanded with focal edema and diffuse fibrosis with dense infiltration of lymphocytes, plasma cells and neutrophils. On immunohistochemistry, there are diffuse positive for CD138 with 102 IgG4+ cells/HPF. We started steroid therapy. 30mg oral prednisolone was administered initially and tapered. Serum creatinine level was decreased and eGFR recovered to 50 mL/min/1.73m2. C3, C4 and IgG4 levels were normalized progressively.
Conclusion: In our case, CT findings were suspected renal infarction at first. The well demarcated, wedge-shaped poor enhanced cortex areas are typical of renal infarction, although they may mimic the patchy involvement of IgG4-TIN. We conclude that renal biopsy is gold standard to differentiate such lesion.