IgG4-related disease with kidney infiltration mimicking multiple myeloma

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Introduction: Immunoglobulin G4-related disease (IgG4-RD) is an immune-mediated disorder, characterized by chronic inflammation and multiple organ infiltration of IgG4. IgG4-RD can form tumefactive lesion or involve multiple organs, making its diagnosis difficult in clinical practice. We report a case of IgG4-RD mimicking multiple myeloma (MM).

Case: A 68-year-old man with diabetes and hypertension presented with general weakness. Laboratory findings revealed that elevated Blood Urea Nitrogen [40.8 mg/dl (6.0~20.0mg/dl)], Creatinine (Cr) [2.94mg/dl (0.7~1.2 mg/dl)], decreased albumin-globulin ratio [0.54, (1~2)], bicytopenia [Hemoglobin 10.1 g/dl (13.0~18.0 g/dl), Platelet 59 x 109/L (150~450 x 109/L)]. Although serum and 24 hours urine electrophoresis (EP) showed no monoclonal gammopathy, there was serum IgG elevation [5266 mg/dl (700~1600 mg/dl)] and abnormal ratio of kappa/lambda [2.92, (0.26-1.65) with serum kappa of 629.74mg/L and serum lambda of 215.94 mg/L]. Bone marrow biopsy showed the 12% of plasma cell among all nucleated cell population. X-ray and PET CT imaging didn't show typical MM-related signs like bone lytic lesion; however, multiple enlarged lymph nodes were observed. Accordingly, we performed renal and abdominal fat biopsy to distinguish MM-related renal disease from other disease-related renal involvement. Abdominal fat biopsy showed no evidence of amyloidosis and pathologic findings. However, renal biopsy indicated IgG4-related renal disease rather than hematologic clonal diseases: immunohistochemical staining showed polyclonally based on CD20 (+), CD3 (+) whereas IgG4+ plasma cells was significantly observed with IgG4+/IgG+ ratio of 0.5. Also, serum IgG subclass study supports the diagnosis of IgG4-RD by marked elevation of IgG4 level to 15 times of upper normal limit. The patient started high-dose steroid therapy, and serum Cr, platelet and serum EP results were improved. This clinical course indicates the diagnosis of IgG4-RD, rather than MM.

Conclusion: Although the bone marrow and serum light chain levels shows suspicious findings for MM, a proactive approach involving organ biopsy may be essential when MM-related signs are insufficienctf