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TUBULOINTERSTITIAL NEPHRITIS ASSOCIATED WITH IGG4-RELATED DISEASE IN A FEMALE PATIENT WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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BACKGROUND

The disease related to immunoglobulin G4 (IgG4-RD) is a fibroinflammatory condition capable of affecting multiple organs. The characteristics of IgG4-related disease (IgG4-RD) are dense lymphoplasmacytic infiltrations with a predominance of IgG4-positive plasma cells in the affected tissue, usually accompanied by some degree of fibrosis and often obliterative phlebitis and an increased number of eosinophils. Serum IgG4 levels are elevated in approximately two thirds of patients. It is more common in middle-aged and older men, but it is also seen in women and has been reported in children. A good initial therapeutic response to glucocorticoids is characteristic and corroborates the diagnostic hypothesis.

CASE REPORT

Female patient, 40 years old, with previous history of subarachnoid hemorrhage (SAH) and systemic lupus erythematosus (SLE), hospitalized with nephrotic syndrome in 2016. Systemic lupus erythematosus started with photosensitivity, serositis, presence of anti-DNA, consumption of supplements and FAN 1:320 NPF. At the time, the patient was using hydroxychloroquine, with good disease control. She referred to xerostomia. Initial laboratory data on admission included normochromic anemia, proteinuria (+++/1.22 g in 24 h) and elevated creatinine. Renal biopsy showed an interstitial plasmacytic infiltrate, with more than 30 plasmacytes per high magnification field, areas with mild tubulitis and focal fibrosis, with normal glomeruli. Immunofluorescence revealed granular deposition of IgG (++) and C3c (++) and C1q (++) in the tubular basement membrane. Immunohistochemistry was positive for IgG4 (EP4420), IgG (RWP49), with Kappa and Lambda light chains and an IgG4/IgG ratio greater than 40%, showing a polyclonal plasmacytic inflammatory infiltrate, indicative of IgG4 tubulointerstitial nephritis. The serum IgG4 level was increased by 6410 mg/dL. The diagnosis of IgG4 tubulointerstitial nephritis was then established. The patient received corticosteroid therapy (methylprednisolone), with significant improvement in symptoms and renal function.

CONCLUSION

Tubulointerstitial nephritis is the most common renal manifestation of IgG4-RD, with different degrees of renal dysfunction and variable clinical findings. The diagnosis of NT by IgG4 can be established based on the clinic, radiological images, laboratory tests and morphological parameters. In addition, it should be noted that IgG4-RD may overlap with other autoimmune diseases, and other causes of nephritis should be excluded. The main differential diagnoses include: lupus nephritis, diabetic nephropathy, cytoplasmic antibody-associated glomerulonephritis, Sjögren's syndrome, chronic pyelonephritis, membranous glomerulonephritis, drug-induced interstitial fibrosis, granulomatous lesions, and idiopathic tubulointerstitial nephritis.