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RENAL INVOLVEMENT IN PRIMARY SJOGREN'S SYNDROME

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BACKGROUND

Primary Sjögren's syndrome (pSS) is an autoimmune disorder characterized by lymphoplasmacytic infiltration of the exocrine glands (salivary and lachrymal) that results in sicca symptoms (dryness of the eyes and mouth). However, it is a systemic disease, and renal involvement can reach up 30% of cases, with tubular and/or glomerular disorders.

CASE REPORT

A serie of 11 cases of distal renal tubular acidosis (dRTA), 10 female and 1 male, mean age of 39 years old, being: 8 patients with recurrent nephrolithiasis (NL), of these 5 had nephrocalcinosis (NC), and 3 patients with hypokalemic paralysis (HP).

We performed a propaedeutic study with the following findings: hypocitraturia in 10 of 11 cases (10/11), urinary concentration deficit 7/11, tubular proteinuria (TP) 7/11, hypercalciuria 1/11, ANA positive in 11/11, anti-Ro 9/11, Rheumatoid Factor 6/8, hypergammaglobulinemia 6/11. Sicca symptoms were reported on 9/11. Five of the 8 patients with NL had previous pSS diagnosis and in all 3 cases of HP the diagnosis of pSS was made during the secondary investigation. Of dRTA, 4 were complete, with systemic acidemia, and 8 were incomplete, confirmed by urinary acidification test, with urinary pH do not acidifying with fludrocortisone and furosemide challenge.

Renal changes in pSS may precede or follow by sicca syndrome. In general, they begin 2-8 years after the diagnosis of the disease. The pathophysiology includes: Tubulointerstitial lymphocytic inflammatory infiltrate (TIN); immunocomplex deposition; and antibodies against specific tubular transporters. The most common histopathological finding is TIN (approximately 80%), followed by cryoglobulinemic membranoproliferative glomerulonephritis. The clinical presentation spectrum is variable, usually expressed by dRTA, TP and CKD. In dRTA, present in 30-70% of the cases, may be accompanied by NC, NL, hypocitraturia and hypokalemia, and results from autoantibodies against H-ATPase of the intercalated cells of the collecting duct. In incomplete dRTA, serum bicarbonate is normal despite alkaline urine, being diagnosed by the urinary acidification test. Severe hypokalemia with HT occurs in up to 7% of cases. Immunosuppressive treatment may prevent progression to CKD.

CONCLUSION

Adults with a history of recurrent nephrolithiasis and nephrocalcinosis should be screened for distal Renal Tubular Acidosis and primary Sjögren Syndrome, even in the absence of sicca symptoms and specific autoantibodies. As well patients with hypokalemic paralysis has seen the high mortality and misdiagnosis in these cases in the emergency rooms. There is ample and subclinical renal involvement, and potential evolution for CKD. Screening for renal manifestations in patients with SS is mandatory.