

Renal tubular acidosis and primary polydipsia in Sjögren's syndrome treated with leflunomide

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Introduction: Sjögren's syndrome is an autoimmune disorder characterized by the infiltration of B-lymphocytes in epithelial glandular and extra-glandular tissues. Lymphocyte infiltration in renal tubular epithelium can lead to various kidney diseases, including tubulointerstitial nephritis, renal tubular acidosis (RTA), nephrogenic diabetes insipidus, and Fanconi syndrome. However, to date, distal RTA and primary polydipsia in a single Sjögren's syndrome patient treated with leflunomide has not been reported in literature. **A case report:** A 47 year-old female was referred to the nephrology department from the gynecology department for hypokalemia with polyuria, polydipsia, and generalized weakness during preoperative screening. She had a history of Sjögren's syndrome and rheumatic arthritis, being treated with leflunomide. At presentation, hyperchloremic metabolic acidosis with hypokalemia and elevated urine pH, which is shown in Table 1, revealed the existence of distal RTA. Furthermore, she had a normal serum osmolality and sodium levels, with a very low urine osmolality of 186 mOsm/kg. Water deprivation test showed no significant body weight change after a 12-hour dehydration and increase in urine osmolality from 186 to 423 mOsm/kg, compatible with a diagnosis of primary polydipsia. The patient was commenced the potassium citrate and sodium bicarbonate tablet for correction of metabolic acidosis and hypokalemia. **Discussion:** Sjögren's syndrome is one of the primary disorders resulting in RTA and primary polydipsia. Thus, RTA and primary polydipsia can be primarily attributed to Sjögren's syndrome in this patient. Of note, she was treated with leflunomide, an immunosuppressive disease-modifying antirheumatic drug, which was suggested a possible association with RTA. From this case, we surmise that close monitoring of electrolyte and acid-base balance is mandatory in Sjögren's syndrome and specific attention is needed in patients who were treated with leflunomide.

Table 1. Basic metabolic panel and urinalysis

Serum Na	Serum K	Serum HCO ₃	Serum Cl	Serum osmolality	Urine pH	Urine Na	Urine K	Urine Cl	Urine Ca	Urine osmolality
140 mEq/L	2.6 mEq/L	14 mEq/L	114 mEq/L	292 mOsm/kg	7.5	113 mEq/L	43 mEq/L	111 mEq/L	7.4 mg/dL	186 mOsm/kg