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Sjögren's Syndrome and Renal Tubular Acidosis Type 1

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Hypokalemia often accompanies certain types of renal tubular acidosis (RTA) as a result of potassium wasting. Distal RTA, also known as type 1 RTA, is typically associated with hypokalemia. The major causes of distal RTA in adults are autoimmune diseases and hypercalciuria. Distal RTA may be the presenting manifestation of autoimmune diseases such as Sjögren's syndrome.

Sjögren's syndrome is characterized by the presence of keratoconjunctivitis, xerostomia and chronic inflammatory sialadenitis. This immune process can also affect non-exocrine organs, including the kidneys, producing an interstitial nephritis and defects in tubular function. The mechanism by which Sjögren's syndrome leads to distal RTA remains to be completely elucidated. During the evaluation of a patient with hypokalemia, we diagnosed distal RTA associated with Sjögren's syndrome. A 31-year-old woman with no significant past medical or family history was admitted with complaints of general weakness. Laboratory tests revealed: serum potassium 3.0 mEq/L, arterial blood pH 7.28, serum bicarbonate 17.8 mEq/L and urinary pH 7.0. Double-labeling confocal fluorescence microscopy using H^+-ATPase and pendrin antibodies demonstrated a decreased expression of these proteins in the patient's renal collecting duct compared to normal controls. Anti-Sjögren's-syndrome-related antigen A (Anti-Ro/SS-A) and anti-Sjögren's syndrome type B (anti-La/SS-B) antibodies were strongly positive with very high titers, consistent with Sjögren's syndrome.

In conclusion, we present a case of distal RTA in Sjögren's syndrome with a defect in H^+-ATPase and pendrin in the renal collecting duct.