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View Abstract

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TITLE: Sjögren's syndrome with nephrogenic diabetes insipidus as the first manifestation: A case report

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ABSTRACT BODY:

Learning Objective 1: Recognize that nephrogenic diabetes insipidus can be a manifestation of Sjögren's syndrome

Learning Objective 2: Diagnosis Sjögren's syndrome with nephrogenic diabetes insipidus as the first manifestation

Case: A 36-year-old female was admitted to hospital because of "polydipsia, polydipsia, polyuria, and fatigue for more than 4 years" The patient developed polydipsia and polydipsia without obvious cause 5 years ago. The daily water intake was about 8000-10000ml, accompanied by general weakness. No family history of similar diseases. Physical Examination: T: 36 °C, P: 86 bpm, R: 21 /min, BP 138/87 mmHg. No abnormality of heart, lung and abdomen. Laboratory examination: 24-hour urine output 4000-5850ml. Urine specific gravity: 1.002-1.005, urine pH6.5, urine protein (+), WBC qualitative 4+, WBC quantitative, bacteria 2571/μl, WBC (high power) 463/HPF. ESR 45mm/h. Blood gas analysis: pH 7.41, PaO_2 70 mmHg, PCO_2 32mmHg, HCO_3 20.3 mm/L, BE 3.4 mmol/L. Anti-SSA (+), Anti Ro-52(+), antinuclear antibody (+). Water deprivation-vasopressin test consistent with nephrogenic diabetes insipidus. After 1 week of potassium supplementation, regular urine test: Urine pH 6.5, specific gravity 1.004. Diagnosis: primary Sjögren's syndrome, renal tubular acidosis, nephrogenic diabetes insipidus. After treatment with potassium titanate, polydipsia and polyuria have reduced, serum potassium was normal.

Discussion: Sjögren's syndrome is a diffuse connective tissue disease characterized by a high degree of lymphocytic infiltration that invades exocrine glands such as lacrimal and salivary glands. The clinical manifestations are mainly keratitis sicca, conjunctivitis sicca, xerostomia, and other internal organs such as lung, liver, pancreas, kidney, blood system, nervous system, etc. Among them, the incidence of renal involvement is about 30%-50% in US according to reports. Renal damage caused by primary Sjögren's syndrome, mainly manifested as renal tubular dysfunction, prominently manifested as distal renal tubular acidosis, renal tubular hydrogen secretion and ammonia secretion dysfunction, hypokalemia, hyperkalemia. Some patients may also have reduced urine concentration, hypotonic urine, fasting of drinking and injection of vasopressin, and the urine osmolality and urine specific gravity cannot be increased, manifesting as nephrogenic diabetes insipidus. In this case, the patient presented with dry mouth, polydipsia, and polyuria. Water deprivation-vasopressin test results suggest of nephrogenic diabetes insipidus. Renal tubular damage leads to the failure of renal tubular water reabsorption, which makes nephrogenic diabetes insipidus as the first presentation in this patient. After the correction of serum potassium and acidosis, the patient's symptoms of polydipsia and polyuria were significantly relieved, which was consistent with the pathological process of renal tubular damage leading to nephrogenic diabetes insipidus. Sjögren's syndrome with nephrogenic diabetes insipidus as the first manifestation is relatively rare and should be paid attention to by clinicians.

PRESENTATION TYPE: Clinical Vignettes

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Shengzhou Wen : Non-Member - Medical Student

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