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RESEARCH ARTICLE

PRIMARY SJOGREN'S SYNDROME WITH RENAL TUBULAR ACIDOSIS AND HYPOKALEMIC QUADRIPARESIS IN MIDDLE AGED FEMALE: A CASE REPORT

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ABSTRACT

Sjögren's syndrome is an autoimmune lymphocytic infiltrative disease that leads to chronic inflammatory changes to exocrine glands and extra-glandular systemic organs. It rarely affects children and adolescents. In cases where adolescents are affected, symptoms include xerostomia, xerophthalmia often leads to a missed diagnosis. Consequently, the first presenting sign of Sjögren's syndrome in adolescents may be heterogeneous, with varying clinical symptoms related to parotitis or systemic organ involvement. Renal involvement occurs in 18–67% of cases, with chronic tubulo-interstitial nephritis being the most frequent pathology which can lead to distal renal tubular acidosis characterized by normal anion gap acidosis with hypokalemia and alkaline urinary pH. Hypokalemic periodic paralysis can be primary or secondary to potassium deficiency which can arise from several causes In our case report, we discuss a 47-year-old female with distal renal tubular acidosis (RTA), who had experienced severe hypokalemic episodes since last 6 month; the patient was eventually diagnosed with Sjögren's syndrome. She was managed and maintained on potassium and alkali repletion therapy.

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INTRODUCTION

Distal renal tubular acidosis is characterized by a failure to acidify the urine in the distal parts of the nephron[1]. Frequently, patients present with minimal or no symptoms, which can lead to a delay in diagnosis. Progressively, it can lead to marked acid-base abnormalities, including hyperchloremic metabolic acidosis and severe hypokalemia, which can be fatal. In children, distal RTA is usually associated with a genetic defect or anatomic abnormality of the urinary system[2]. In contrast, distal RTA in adults is frequently related to acquired conditions such as infections, drugs, and autoimmune diseases. We describe a case of a woman with severe hypokalemia and weakness as the main reason for admission. Renal tubular acidosis (RTA) is characterized by renal tubular impairment in balancing physiologic acid-base. It often results from a defect in tubular transporters, which participate in the secretion or uptake of specific ions, due to congenital causes, exposure to nephrotoxic drugs, diuretic abuse, autoimmune disease, or malignancy (e.g., multiple myeloma).

There are three major types of RTA: distal or type 1, proximal or type 2, and hyperkalemic or type 4. All three types of RTA are characterized by a positive urine anion gap, hyperchloremic non-anion gap metabolic acidosis, alkalotic or acidotic urine pH, and serum potassium derangements (hypo- or hyperkalemia).

CASE VIGNETTE

A 47-year-old woman presented with an history of limb weakness and pain since last 1 day, she had noticed suddenlimb weakness and pain and experienced instability while standing. The patient's medical history included symptoms such as dry mouth and dry eyes that had appeared in last 8 months. On physical examination, the patient's vital signs were within the normal limits, and her higher mental functions were intact. Her oral cavity was dry, and she had no lymphadenopathy. The proximal muscle strength of the upper limbs was grade 1, and the distal muscle strength was grade 2; the muscle strength of the bilateral lower limbs was grade1.

Biochemical and hematological investigations

CBC	**Hb - 13.6 g/dl, TLC – 10.7×10^9/L, Plt - 256×10^9/L**
Serum electrolytes	Sodium - 139 mmol/L, potassium - 1.3 mmol/L, chloride – 115.40 mmol/L
ABG	pH - 7.30, pCO_2-37 mmHg, HCO_3-16.6 mmol/L, potassium –1.8 mmol/L, serum osmolality - 298 mos/kg
Urinalysis	pH - 7.45, urinary Na^+ - 80.0 mmol/L, K^+ - 23 mmol/L, urine chloride - 44 mmol/L, urine osmolality -312 mmol/kg
Serum anion gap	08 mmol/L
Urinary anion gap	59 mmol/L (positive)
LFT	ALT - 24 U/L, AST - 14 U/L, total protein/albumin –8.3/3.83 mg/dl
KFT	Urea - 34 mg/dl, creatinine –0.8 mg/dl
Viral markers	HIV - negative, HBsAg - negative, Anti-HCV – negative
Autoimmune profile	ANA - >10 byELISA, anti-SS-A/Ro - 240H U/ml, SS-b –147 H U/ml

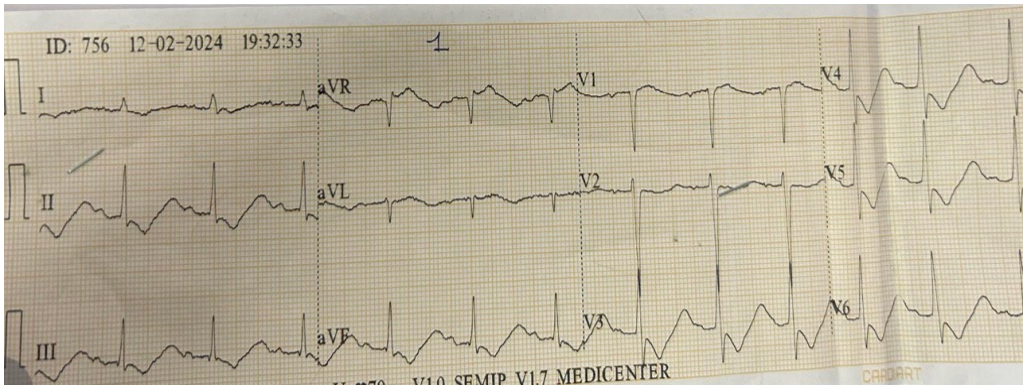

Fig 1. electrocardiograph show T-inversion, prolong PR interval, U-wave in precordial lead and slurring of T-wave into U wave

The tendon reflex was negative, and the muscle tension of the limbs was weak. No sensory deficit was present, and cranial nerve examination findings were unremarkable. Cardiovascular, respiratory, gastrointestinal, and thyroid examination findings were normal. The patient presented to the emergency department, and initial investigations revealed severe hypokalemia of 1.3 mmol/ L (reference range, 3.5–5.5 mmol/L), hyperchloremia of 115.40 mmol/L (reference range, 98–111 mmol/L), and hypophosphatemia of 1.4mmol/L (reference range2.5–4.5 mmol/L). An electrocardiogram showed ST-T-U changes. The patient was intravenously administered 1.5 g of potassium chloride, and her serum potassium level was 2.0 mmol/L the next morning. The intravenous injection of potassium chloride was repeated, and oral potassium chloride liquid were administered. After a series of treatments, her clinical condition substantially improved and her muscle power normalized. We repeated the relevant examinations after the patient's condition had stabilized and found a urinary pH of 6.0, with no history of diuretic use, vomiting, or diarrhea. The basic metabolic panel suggested low potassium, high chlorine, and hypophosphatemia, and an arterial blood gas analysis indicated metabolic acidosis with severe hypokalemia (serum potassium of 2.1 mmol/L). A diagnosis of RTA was made. SS was suspected based on the patient's history ofxerostomia/xerophthalmia without secondarycauses, and SS was later confirmed by antinuclear antibody test which was positive by ELISA method, SSA/Ro-52 strongly positive and SS-B/La also positive. Some related analyses were also conducted. Abdominal ultrasound study was normal. Opticalcoherence tomography showed that the retinal structure of the macular area of the eyes was almost normal. Schirmers test revealed tear secretion, 4 mm in the right eye and 4 mm in the left eye; tear breakup time, 2 seconds in the right eye and 3 seconds in the left eye. static imaging of salivary glands, severe impairment of bilateral salivary gland secretion. Ultrasound bilateral parotid gland enlargement with heterogeneous echogenicity, bilateral submandibular gland

DISCUSSION

Sjögren syndrome is chronic systemic autoimmune disorder characterized by lymphocytic infiltration of exocrine glands. The disease can present as alone (primary Sjögren syndrome) or associated with other autoimmune conditions such as SLE, rheumatoid arthritis, and scleroderma (secondary Sjögren syndrome). The prevalence of primary Sjögren's syndrome is ~0.5%–1% and middle-aged women (female-to-male ratio is 9:1) are primarily affected[3] Most patients presentwith sicca symptoms such as xerophthalmia (dry eyes), xerostomia (dry mouth), and parotid gland enlargement. In addition, various extraglandular features may develop. This includes arthralgia, arthritis, vasculitis, lymphoma, renal involvement, and Raynaud's phenomenon[4] Renal involvement includes proximal and distal tubular acidosis, tubular proteinuria, and nephrogenic diabetes insipidus[5]. mechanism of hypokalemia in Sjögren syndrome is because of distal RTA brought about by chronic interstitial nephritis which in turn leads to decrease tubular sodium delivery. Distal RTA, also known as type 1 RTA or classic RTA, is a complex entity characterized by an inability to acidify the urine; a process that occurs in the distal parts of the nephron, including the connecting tubule and the collecting duct[6]. Little is known about the prevalence of this condition in the general population the prevalence of distal RTA was 2.8%, concerning for an endemic form of this disease[7]. However, the prevalence of distal RTA can be as high as 22–25% in specific populations, such as in patients with osteopenia and SS[8].The exact mechanism by which SS causes distal RTA is incompletely understood. The complete absence of H-ATPase pump in cortical collecting duct is one of the proposed theories described in some studies. Another possible mechanism is carbonic anhydrase 2 inhibition caused by the high titer of autoantibodies, which leads to defective H^+ secretion[9].Hypokalemia is common in RTA, but severe hypokalemia mimicking periodic paralysis is rare[10]. This case presentation of hypokalemic periodic paralysis,

hyperchloremic metabolic acidosis and alkaline urine pH all pointed to the diagnosis of distal RTA.This case report describes a patient with recurrent hypokalemia. Her clinical manifestations included severe muscle weakness in upper and lower extremities, and marked acid-base disorders such as hyperchloremic metabolic acidosis and severe hypokalemia. Her urine studies showed persistent alkaline urine, a positive UAG, and urinary eosinophils, suggestive of distal RTA and an underlying nephritis. On further evaluation, the patient was diagnosed with SS and acute tubular injury. She was treated with prednisolone, hydroxychloroquine daily and MTX 7.5 mg weekly. Potassium chloride two tablets daily were added to the above regimen and the dose of prednisolone was increased to 15 mg daily to control the RTA. The level of serum potassium was increased and her weakness was gone after the replacement therapy.

CONCLUSION

We report this rare case presentation of Sjogren's syndrome in a middle-aged female who suffered from recurrent hypokalemic paralytic episodes due to distal RTA with sicca symptoms. A high index of suspicion is therefore required in females who present with hypokalemia with distal renal tubular acidosis for early diagnosis and treatment of Sjogren's syndrome so that complications can be averted

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