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Fanconi syndrome in Patient with Primary Sjogren's syndrome

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Case Study: Introduction

Primary sjogren's syndrome is a systemic autoimmune disease. Characteristic symptoms include dryness of the mouth and eyes, fatigue and joint pain. Extraglandular involvement may occur. Renal involvement in primary sjogren's syndrome may be associated with morbidity and mortality. Herein, we present Fanconi syndrome in Patient with Primary Sjogren's syndrome.

Case Reports

A-67-year-old woman was admitted with fatigue. On admission, level of potassium and phosphate was low and normal anion gap metabolic acidosis was identified. The results of the urinary potassium loss and transtubular potassium gradient confirmed the renal potassium excretion. She also showed the positive urine anion gap. The patient was diagnosed with renal tubular acidosis. In addition to these results and considering glucosuria with a normal serum glucose concentration, hypouricemia and aminoaciduria, she was diagnosed Fanconi syndrome. Additional tests were conducted to determine the cause of the renal tubular acidosis. The patient complained of dry mouth and dry lips, and was positive for ANA (> 1:1280, speckled nucleolar pattern) and anti-SS-A (Ro) Ab. Schirmer test was positive, and the result of the salivary gland scan was that the function of left parotid and left submandibular glands was decreased. Based on the above results, primary Sjogren's syndrome was diagnosed. Because of azotemia, renal biopsy was performed and acute tubulointerstitial nephritis was diagnosed. Based on the above results, the patient was diagnosed with acute tubulointerstitial nephritis and Fanconi syndrome due to renal involvement of primary Sjogren's syndrome. She has been treated with methylprednisolone and oral supplementation with sodium bicarbonate, and electrolyte imbalance and renal function were improved.

Conclusion

If metabolic acidosis and hypokalemia are present in patients with primary sjogren's syndrome, the possibility of renal involvement and the early diagnosis is important for preventing complications and improving prognosis.

Tubules reveal focal severe necrosis and loss with infiltration of mononuclear cells and neutrophils in interstitium as with fibrosis

