P98

쇼그렌증후군과 원발성 담관성간경화를 가진 환자에서 병발한 판코니증후군과 원위부 신세뇨관산증 1예

울산대학교 의과대학 서울아산병원 내과학교실

백주은 · 석현정 · 김태희 · 장재원 · 김순배 · 박수길 · 이상구 · 박정식 · 양원석

Fanconi Syndrome and Distal Renal Tubular Acidosis in a Patient with Sjögren's Syndrome and Primary Biliary Cirrhosis

Joo Eun Baek, Hyun Jung Seok, Tae Hee Kim, Jai Won Chang
Soon Bae Kim, Su-Kil Park, Sang Koo Lee, Jung Sik Park, Won Seok Yang

Department of Internal Medicine Asan Medical Center College of Medicine University of Ulsan

Introduction : Sjögren's syndrome (SS) and primary biliary cirrhosis (PBC) are mutually associated autoimmune diseases characterized by lymphocytic infiltration of the exocrine glands or portal regions. Tubulointerstitial nephritis (TIN) is the most common renal complication in SS and typically manifested by distal renal tubular acidosis (RTA). Distal RTA is also the main feature of renal involvement in patients with PBC. In contrast to distal RTA, proximal RTA or Fanconi syndrome has been reported only in a few cases of either disease, and was associated with severe pathologic changes such as tubular atrophy and extensive fibrosis. We report a 37-year-old woman with SS and PBC who exhibited both proximal and distal tubular dysfunction.

Case report : The patient had suffered from xerostomia and xerophthalmia. Laboratory tests revealed hypokalemia, hypouricemia and low normal phosphate. In liver function tests, AST, ALT and bilirubin were normal, but alkaline phosphatase and & γ-GT were mildly increased. Urinalysis demonstrated normoglycemic glucosuria, low-grade proteinuria and hematuria. Antibodies to Ro/SS-A and La/SS-B antigens, anti-mitochondrial antibody and antinuclear antibody were positive. Blood gas analysis revealed metabolic acidosis with normal serum anion gap (AG) and positive urine AG. Urine pH was constantly $\geq$7.0. Schirmer's test was positive, and salivary scintigraphy showed decreased uptake of both submandibular glands. The patient was diagnosed with SS, and we proceeded with further evaluation of suggested RTA, and kidney and liver biopsies. The diagnosis of distal RTA was suggested by alkaline urine and positive urine AG in hyperchloremic metabolic acidosis. An intravenous bicarbonate loading test revealed a high fractional excretion of bicarbonate indicating the presence of proximal RTA. Fractional excretions of phosphate and uric acid were increased. Fanconi syndrome was diagnosed based on hypokalemia, hypouricemia, normoglycemic glucosuria, phosphaturia, uricosuria together with proximal RTA. Kidney biopsy showed TIN with mild tubulointerstitial lymphocytic infiltrate, and liver biopsy demonstrated typical features of PBC. Cellular infiltrate was composed mainly of CD4+ and CD8+ lymphocytes in both tissues.

Conclusion : This is the first report of Fanconi syndrome and distal RTA in a patient with SS and PBC. Proximal RTA was present without severe histologic changes. The mechanisms underlying proximal RTA need further investigation.