ACR Open Rheumatology
Vol. 3, No. 7, July 2021, pp 496

DOI 10.1002/acr2.11288

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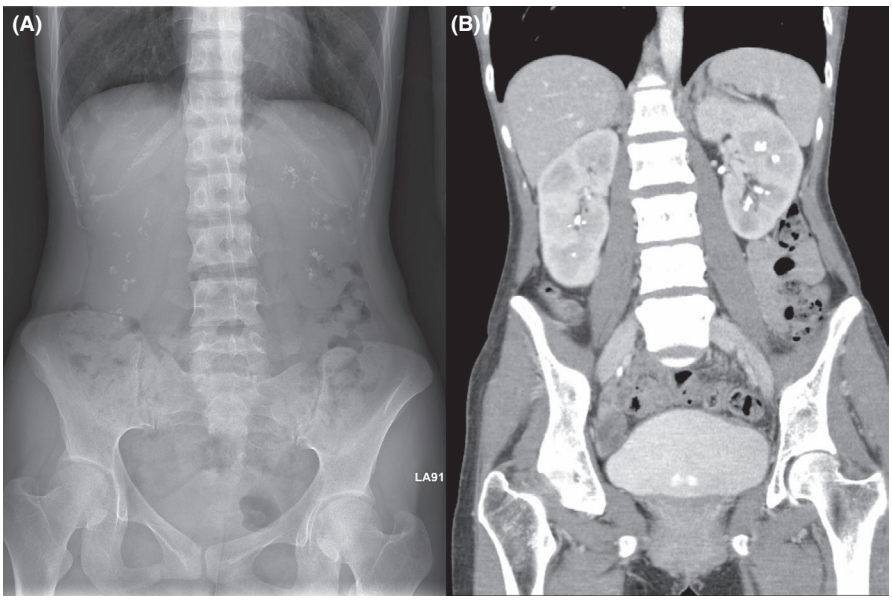
Clinical Images: Medullary nephrocalcinosis in Sjögren syndrome



Sjögren syndrome is a chronic autoimmune disease that can involve multiple organs, especially the exocrine system (1). In addition to xerostomia and xerophthalmia, it can also attack extraexocrine organs, including pulmonary, digestive, mucocutaneous, and urinary systems (such as type 1 renal tubular acidosis [RTA]–induced nephrocalcinosis) (2,3). RTA is a condition in which systemic acidosis occurs together with alkaline urine and hypercalciuria. The alkaline environment of the renal tubules reduces the solubility of the calcium deposition; hence favoring calcium phosphate precipitates (4,5) in the renal calyx, it forms the shape of the calyces, which resemble clusters of flowers. Herein, we demonstrated that one female patient with Sjögren syndrome–related type I RTA presented with bilateral medullary nephrocalcinosis with the flower cluster–like appearance of multiple tiny renal stones and profound hypokalemia. The patient, 31-year-old woman, had persistent weakness, polyarthralgia, and recurrent urinary tract infection. A laboratory survey disclosed profound hypokalemia (3.43 mmol/L) with acidemia (serum CO_2: 15 mmol/L), normal anion gap, and high transtubular potassium gradient. Image studies showed bilateral nephrocalcinosis with multiple tiny stones with flower cluster–like appearances (abdominal radiograph; **A**) in the renal medullary region (pelvic computed tomography with contrast enhancement; **B**). She had polyclonal hypergammaglobulinemia with positive antinuclear antibody (640×; speckle pattern) and high serum antibodies level of anti-SSA/Ro Ab (>240 U/mL) and anti-SSB/La Ab (>320 U/mL). Sialoscitingraphy revealed impaired salivary gland function, and Schirmer's test showed inadequate tear secretion (<5 mm/5 minutes; bilateral). Therefore, Sjögren syndrome–related type I RTA with nephrocalcinosis and hypokalemia was diagnosed (6,7). She was under control with taking oral potassium citrate monohydrate, low-dose prednisolone, hydroxychloroquine, and azathioprine.

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