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Nephrocalcinosis in primary Sjögren's syndrome

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A 29-year-old woman presented with a known nephrocalcinosis that had developed nine years earlier. Her past medical history was remarkable for a cardiac arrest secondary to a profound hypokalaemia (1.7 mEq/L) in the post-operative period following a complicated appendicular peritonitis at 19 years of age. Her on-going treatment included potassium citrate supplementation and increased fluid intake to induce hyperdiuresis. There was no relevant family history. On admission, pertinent laboratory data were as follows: hypokalaemia (3.4 mEq/L), normal anion gap hyperchloraemic metabolic acidosis (plasma HCO_3^- 21 mEq/L), inappropriately high urine pH (7.0), positive urinary anion gap (þ21 mEq/L), tubular proteinuria 0.5 g per 24 h and chronic kidney disease (serum creatinine, 1.4 mg/dL. CKD Epi MDRD creatinine clearance, 72 mL/min). A urine pH of >5.3 in the presence of metabolic acidosis suggests distal renal tubular acidosis. On bicarbonate loading, $FEHCO_3^-$ was <5%, with a low (urinary–blood) $DpCO_2$ (13 mmHg), suggesting defective H^+ secretion by alpha-intercalated cells. A CT scan imaging (Figure 1A) and plain X-ray of the abdomen (Figure 1B) showed macroscopic bilateral nephrocalcinosis. Biochemical analysis of the kidney stone revealed a calcium phosphate stone type suggestive of distal renal tubular acidosis (dRTA). She had polyclonal hypergammaglobulinaemia and positive antinuclear antibodies (1/1600) of anti-Ro/SS-A type. Accessory salivary gland biopsy showed Grade 2 inflammatory infiltrates on the Chisholm–Mason scale. The kidney biopsy revealed mild inflammatory cell infiltrates, with interstitial fibrosis.

Distal RTA is the most common cause of nephrocalcinosis due to hypercalciuria without hypercalcaemia. The reported prevalence of nephrocalcinosis in patients with dRTA ranges from 60 to 80%. Distal renal tubular acidosis (dRTA) related to Sjögren's syndrome occurred in up to 33%. Primary Sjögren's syndrome may manifest itself as dRTA-associated nephrocalcinosis by autoantibodies directed against the A-type intercalated cells in the collecting tubules leading to a defect in hydrogen ion secretion.

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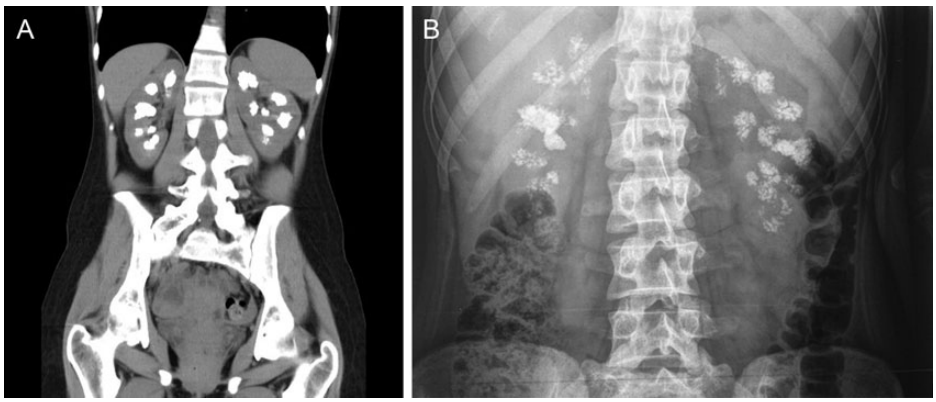

Fig. 1. (A) Bilateral macroscopic nephrocalcinosis. (B) Plain X-ray.