An Underrecognized Cause of Recurrent Nephrolithiasis: Distal Renal Tubular Acidosis Progressing to Xanthogranulomatous Pyelonephritis and Nephrectomy

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Case Presentation

A 50-year-old male with a history of spina bifida complicated by neurogenic bladder status post ileal conduit, chronic kidney disease, and recurrent multidrug-resistant urinary tract infections presented with a two-week history of vomiting, decreased oral intake, and cloudy urine. Initial evaluation revealed leukocytosis, acute kidney injury, and urinalysis notable for pyuria and hematuria. Computed tomography of the abdomen and pelvis revealed multiple obstructing right renal calculi with severe cortical loss and inflammatory changes consistent with xanthogranulomatous pyelonephritis. Prior nuclear imaging demonstrated a nonfunctioning right kidney. The patient underwent right nephrectomy with drainage of associated retroperitoneal abscess. Further metabolic evaluation revealed a non-anion gap metabolic acidosis with urine pH of 7 and a positive urine anion gap. A 24-hour urine collection revealed hypocitraturia, confirming distal renal tubular acidosis (RTA) as the underlying driver of recurrent nephrolithiasis. Potassium citrate therapy was initiated for stone prevention. Following nephrectomy, renal function initially improved but remained labile, with recurrent episodes of acute kidney injury in the setting of a solitary functioning kidney, ongoing infection, and volume shifts.

Discussion

Distal RTA is an underrecognized metabolic disorder characterized by impaired hydrogen ion secretion, leading to alkaline urine and hypocitraturia, both of which promote calcium nephrolithiasis. In this patient, unrecognized distal RTA likely contributed to years of recurrent stone formation, ultimately leading to chronic obstruction, infection, and progression to xanthogranulomatous pyelonephritis with loss of renal function. This case demonstrates the compounding effect of metabolic and structural risk factors in progressive stone disease. While neurogenic bladder and urinary diversion predisposed this patient to urinary stasis and infection, the presence of distal RTA provided a critical metabolic driver of ongoing stone formation. Recognition of distal RTA occurred only after advanced disease had developed, representing a missed opportunity for early metabolic evaluation and prevention. Furthermore, this case highlights the heightened vulnerability of patients with a solitary kidney, in whom even minor physiologic stressors may precipitate recurrent kidney injury. This case underscores the importance of early metabolic evaluation in patients with recurrent nephrolithiasis, even when structural abnormalities are present.