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Mark E. Rosenberg

On the Cover

What is the diagnosis?
A 32-year-old male presented for initial evaluation of CKD with a history significant for Fabry disease. Past medical history was notable for irritable bowel syndrome, painful neuropathy of his extremities, and anhidrosis. On examination, blood pressure was 110/74 mmHg; there was no flank tenderness. Serum creatinine was 0.64 mg/dl. Urine dipstick was positive for protein of 100mg/dl but otherwise bland.

Image Description:
A screening renal ultrasound showed multiple anechoic lesions in the renal sinus with no connection to the ureter, which was initially characterized as moderate bilateral hydronephrosis (Left). Magnetic resonance imaging (MRI) with gadolinium was performed to further evaluate the presence of obstruction. This revealed multiple bilateral T2 hyperintense parapelvic cysts without communication with the collecting system (Center). These were non-enhancing cystic lesions along the renal sinuses without evidence of contrast filling on T1 delayed postcontrast imaging (Right). There was no obstructing calculus or hydronephrosis, and the corresponding findings on the ultrasound were attributed to the bilateral renal sinus cysts discovered on MRI.

Teaching Points:
Parapelvic cysts are common in Fabry disease, with an estimated prevalence of 29%–43%, significantly higher than the approximately 1% estimated prevalence in the general population; but they are not specific to this condition. They are often misdiagnosed as hydro nephrosis due to the hypoechoic character by ultrasonography and the location in the renal sinus. When asymptomatic hydronephrosis is noted by ultrasonography without identifiable obstructive lesions in patients with Fabry disease, the possibility of parapelvic cysts should be considered.

Disclaimer:
The views expressed herein are those of the authors and do not reflect the official policy of the Departments of Army, Navy, Air Force, Department of Defense, or US Government.

(Images and text provided by Richard Plasse, DO, Megha Joshi, DO, Robert Nee, MD, and Maura Watson, DO, MPH, Nephrology Service, Walter Reed National Military Medical Center, Bethesda, MD, and Department of Medicine, Uniformed Services University, Bethesda, MD; and Nathan Bumbarger, Radiology Service, Walter Reed National Military Medical Center, Bethesda, MD.)