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The Clinical Picture

Recurrent pyelonephritis as a sign of ‘sponge kidney’



FIGURE 1. Radiograph, kidney-ureter-bladder view, showing multiple radiopaque densities (arrowheads) within bilateral renal shadows.



FIGURE 2. Computed tomographic scan of the abdomen showing bilateral renal medullary nephrocalcinosis (arrowheads).

A 39-YEAR-OLD WOMAN PRESENTED 1 month ago with left flank pain, chills, and spiking fever over the past day. She reported having an episode of acute pyelonephritis with similar manifestations 6 months previously.

Blood tests showed a leukocyte count of $12.2 \times 10^9/L$ (normal range $7.05\text{--}14.99 \times 10^9/L$) with predominant neutrophils, C-reactive protein 6.3 mg/dL (0.0–1.0 mg/dL), blood urea nitrogen 16 mg/dL (10–25 mg/dL), and serum creatinine 1.1 mg/dL (0.70–1.40 mg/dL). Radiographic study of the kidneys, ureters, and bladder showed multiple radiopaque densities in the calyces of both kidneys (FIGURE 1). Computed tomography of the abdomen without contrast revealed multiple punctuated calcifications aligned concentrically in the medulla

of both kidneys (FIGURE 2). Blood and urine cultures were positive for *Escherichia coli* that was sensitive to ceftriaxone (Rocephin). This antibiotic was started, and her acute pyelonephritis and bacteremia resolved.

Now, at a follow-up visit 1 month later, intravenous urography clearly shows unique linear and striated opacities and cupping of renal papillae, key features of medullary sponge kidney (FIGURE 3). Because medullary sponge kidney is strongly associated with kidney stones, potassium citrate is given to prevent medullary calculi formation.

KEY FEATURES

Medullary sponge kidney causes extensive cystic dilation of medullary collecting tubules.¹ It

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FIGURE 3. Intravenous urogram at 10 minutes demonstrating the bright brush-like or “paintbrush” pattern (arrowheads) of contrast collection along ectatic medullary collecting tubules uniformly directing the cupping of papillae.

More than 70% of patients with medullary sponge kidney develop stones

is usually an incidental finding in patients undergoing intravenous urography as part of the evaluation for infection, hematuria, or kidney stones.

The classic urographic appearance is linear striations with small brushes or “bouquets of flowers,” which represent the collection of contrast material in small papillary cysts.

Medullary sponge kidney has long been considered a congenital disorder, but the genetic defect has not yet been identified, and the pathogenesis is not yet known. It has only rarely been reported in children.²

It is usually asymptomatic, but complications may occur, including nephrocalcinosis or lithiasis, urinary tract infection, renal tubular acidosis, and impaired urine concentrating ability.

RISK OF LITHIASIS

Gambara et al³ estimated that medullary sponge kidney is found in up to 20% of patients with urolithiasis, and that more than 70% of patients with medullary sponge kidney develop stones.

Cystic dilation of medullary collecting tubules (an anatomic abnormality) inevitably causes urinary stasis. This, combined with hypercalciuria and reduced excretion of urinary citrate and magnesium (metabolic abnormali-

ties), contributes to lithiasis.⁴ Lithiasis in medullary sponge kidney is a well-known cause of urinary tract infection, and it tends to facilitate infective stone formation after episodes of urinary tract infection.⁵

The unique anatomic derangement of medullary sponge kidney contributes to the recurrence of pyelonephritis, in addition to the conventional risk factors—frequent sexual intercourse, avoidance of voiding because it is inconvenient, incomplete bladder emptying, ureteropelvic junction obstruction, ectopic ureter, and impaired immunity.⁶

DIAGNOSIS AND TREATMENT

Intravenous urography is the gold standard for the diagnosis of medullary sponge kidney; computed tomography and ultrasonography are generally limited in their ability to clearly show the tubular ectasia.⁷

Treatment includes antibiotics for acute pyelonephritis and thiazide diuretics and potassium citrate to prevent stone formation and renal tubular acidosis. Due to its silent course, medullary sponge kidney should be considered not only as a cause of nephrocalcinosis and nephrolithiasis, but also as a distinct entity complicating recurrent pyelonephritis.

REFERENCES

1. Gambara G, Feltrin GP, Lupo A, Bonfante L, D'Angelo A, Antonello A. Medullary sponge kidney (Lenarduzzi-Cacchi-Ricci disease): a Padua Medical School discovery in the 1930s. *Kidney Int* 2006; 69:663–670.
2. Kasap B, Soylu A, Oren O, Turkmen M, Kavukcu S. Medullary sponge kidney associated with distal renal tubular acidosis in a 5-year-old girl. *Eur J Pediatr* 2006; 165:648–651.
3. Gambara G, Fabris A, Puliatta D, Lupo A. Lithiasis in cystic kidney disease and malformations of the urinary tract. *Urol Res* 2006; 34:102–107.
4. O'Neill M, Breslau NA, Pak CY. Metabolic evaluation of nephrolithiasis in patients with medullary sponge kidney. *JAMA* 1981; 245:1233–1236.
5. Miano R, Germani S, Vespasiani G. Stones and urinary tract infections. *Urol Int* 2007; 79(suppl 1):32–36.
6. Scholes D, Hooton TM, Roberts PL, Gupta K, Stapleton AE, Stamm WE. Risk factors associated with acute pyelonephritis in healthy women. *Ann Intern Med* 2005; 142:20–27.
7. Maw AM, Megibow AJ, Grasso M, Goldfarb DS. Diagnosis of medullary sponge kidney by computed tomographic urography. *Am J Kidney Dis* 2007; 50:146–150.

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