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CASE STUDY

Medullary Sponge Kidney

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An 18-year-old man was evaluated for recurrent urinary tract infections (UTIs). His first infection was at age 15 with Escherichia coli; before this occurrence he had experienced an episode of reddish urine; had no episodes of stones, and had no other contributory past or family history. His serum creatinine was 0.9 mg/dL and his urinalysis was negative. His voiding cystourethrogram was negative. Because of his history, there was a suspicion for some structural disorder or stones. Subsequently his intravenous pyelogram (IVP) showed medullary sponge kidney (MSK) (Figure 1).

Another patient with similar symptoms was evaluated with a computed tomographic urogram (CTU) (Figures 2 and 3).

MSK is a congenital disorder that involves malformation of the terminal collecting ducts of renal tubules, which leads to formation of microscopic to large cysts in the medullary portion of both kidneys mostly, though it can be unilateral.[1] The true prevalence of this disorder is not clear, though estimates range from 1 in 5000 to 1 in 20,000 people.[2] Most patients have no family history, although a rare autosomal dominant form has been reported and MSK also can rarely occur in conjunction with other rare congenital abnormalities. It is more common in patients who are recurrent stone formers (mostly calcium oxalate and calcium phosphate) and in women.

Most patients are asymptomatic. If patients do have clinical manifestations, they are caused by kidney stones, hematuria, and urinary tract infections. These mostly start presenting in the second and third decade of life. Other signs may also be renal tubular defects in acidification and urinary concentration.

This disorder is mostly diagnosed during work-up for hematuria, recurrent stones, recurrent urinary tract infections, or incidentally discovered during work-up of urologic disorders, after radiologic study such as an IVP or CTU.[3,4]

There is no specific therapy that can treat the cysts of MSK. Once this diagnosis is made, it is imperative to follow recommendations for prevention and treatment of UTIs and stones. There are various guidelines for prevention of recurrent UTIs. Regarding stones, prevention methods include oral citrate supplements depending on 24-hour urine collection results and adequate hydration so as to maintain a urine output of greater than 2 liters. Although there is no guideline for surveillance or specialty referral once MSK is diagnosed, an

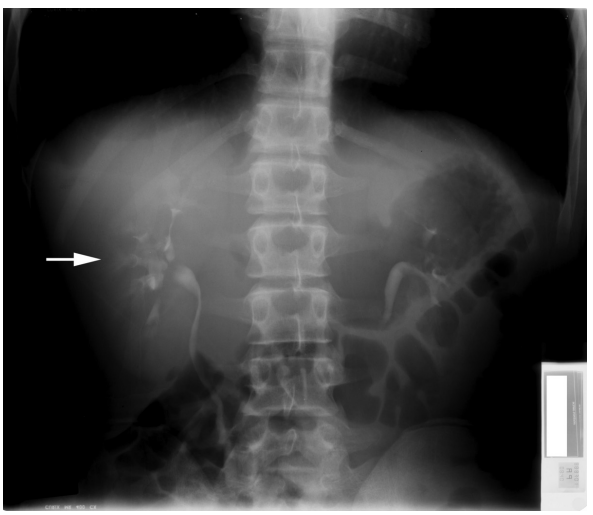

Figure 1. Intravenous pyelogram showing the classic "paintbrush" (arrow) appearance of dilated contrast-filled tubules within the renal medulla, characteristic for medullary sponge kidney.

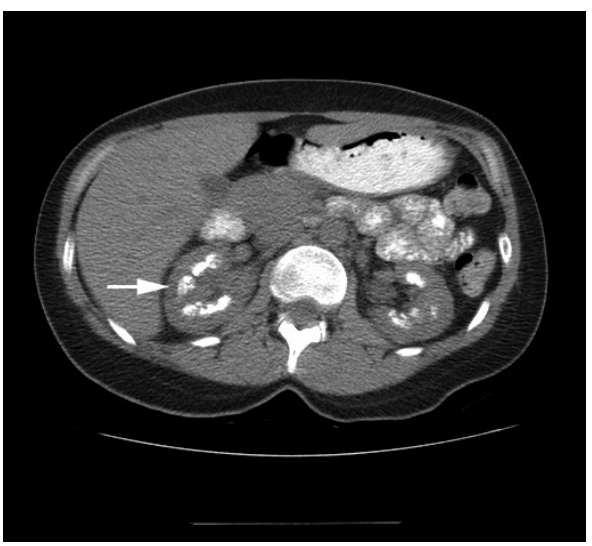

Figure 2. Computed tomographic urogram (precontrast) axial view demonstrating clusters of pyramidal medullary calcification (arrow), characteristic for medullary sponge kidney.

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initial nephrology referral can be useful. However, in cases of complications from infection or stone, specialty referral will be mandated.

Prognosis is generally good. However, in cases of recurrent infections and stones, chronic kidney disease can develop. ❖

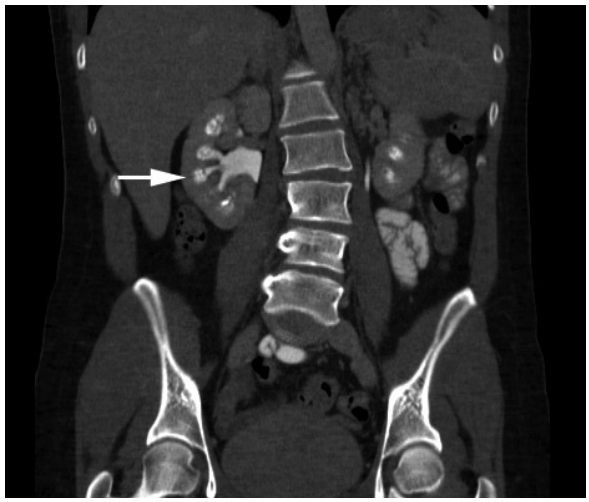

Figure 3. Computed tomographic urogram (postcontrast) coronal view demonstrating dilated contrast-filled tubules within the renal medulla and pyramidal medullary calcifications (arrow).

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