doi: 10.5262/tndt.2015.1001.23

A Case of Medullary Sponge Kidney Diagnosed at an Older Age

İleri Yaşta Tanı Konan Medüller Sünger Böbrek Hastalığı Olgusu

ABSTRACT

Medullary sponge kidney (MSK) is known as a congenital disesase with dilatation of collecting tubules. MSK is usually asymptomatic but various clinical manifestations may occur including nephrocalcinosis, renal tubular acidosis, impaired urine concentrating ability, urinary tract stones and infections. Although MSK is a congenital disorder, it is usually diagnosed in second or third decade. A 53-year-old woman presented with recurrent dysuria and bilateral costovertebral pain radiating to the groin region. Pyuria and hematuria were found. On ultrasonography, hyperechogenic densities of multiple calculi in the calyces of both kidneys, and minimal pelvicaliectasis were detected. Computed tomography revealed multiple punctate calcifications aligned concentrically in the medulla of both kidneys. On clinical and radiological grounds, the diagnosis of MSK was established. The patient's complaints were resolved with antibiotherapy for urinary tract infection. Thiazide or potassium citrate was not prescribed since hypercalciuria or hypocitraturia was not present. In conclusion, MSK should be considered in the differential diagnosis of nephrocalcinosis, nephrolithiasis and recurrent urinary tract infections in older patients, even in the fifth decade.

KEY WORDS: Medullary sponge kidney, Nephrocalcinosis, Nephrolithiasis, Urinary tract infection

ÖZ

Medüller sünger böbrek (MSB), renal papilladaki toplayıcı kanalların dilatasyonuyla karakterize konjenital bir hastalıktır. MSB genellikle asemptomatiktir fakat nefrokalsinozis, renal tübüler asidoz, idrarı konsantre etme kapasitesinde bozulma, böbrek taşları ve enfeksiyonları içeren çeşitli klinik durumlarla ortaya çıkabilir. MSB konjenital bir hastalık olmasına rağmen, genellikle 2. veya 3. dekada kadar tanı konulamaz. Bu olgu bildiriminde, 53 yaşında MSB tanısı konan bir kadın olguyu tartışmayı amaçladık. Hastamız, tekrarlayan idrarda yanma ve kasık bölgesine yayılan çift taraflı yan ağrısı şikayetiyle başvurdu. Tetkiklerinde piyüri ve hematüri saptandı. Ultrasonografide her iki böbrek kalikslerinde birden fazla taşla uyumlu hiperekojen dansiteler ile birlikte minimal pelvikaliektazi tespit edildi. Bilgisayarlı tomografide, her iki böbrek medüllasında çok sayıda noktasal kalsifikasyonlar saptandı. Hastaya klinik ve radyolojik kriterlerle MSB tanısı kondu. Uygun antibiyotik tedavisiyle idrar yolu enfeksiyonu tedavi edildi ve hastanın şikayetleri geriledi. Hiperkalsiüri veya hipositratüri saptanmadığı için tiazid veya potasyum sitrat tedavileri başlanmadı. Sonuç olarak, hastalar ileri yaşta olsa bile, MSB, nefrokalsinozis, nefrolitiazis ve tekrarlayan idrar yolu enfeksiyonu ayırıcı tanısında düşünülmelidir.

ANAHTAR SÖZCÜKLER: Medüller sünger böbrek, Nefrokalsinozis, Nefrolitiazis, İdrar yolu enfeksiyonu

Erhan EKEN
Abdullah ÖZKÖK
Fatoş ARSLAN
Satılmış BİLGİN
Kübra AYDIN BAHAT
Ali BAKAN
Ömer Celal ELCİOĞLU
Ali Rıza ODABAŞ

İstanbul Medeniyet University,
Göztepe Training and Research Hospital,
Department of Nephrology,
İstanbul, Turkey

Received : 18.12.2013
Accepted : 10.01.2014

Correspondence Address:
Abdullah ÖZKÖK
İstanbul Medeniyet Üniversitesi, Göztepe Eğitim ve Araştırma Hastanesi,
Nefroloji Bölümü, İstanbul, Turkey
Phone : +90 216 570 92 97
E-mail : abdullahozkok@yahoo.com

INTRODUCTION

Medullary sponge kidney (MSK) is known as a congenital disesase with dilatation of collecting tubules. The exact pathophysiology has not yet been identified (1, 2). MSK is usually asymptomatic but various clinical manifestations may occur including nephrocalcinosis, renal tubular acidosis, impaired urine concentrating ability, and urinary tract stones and infections (3-5). Although MSK is a congenital disorder, it is usually diagnosed in second or third decade (2,6). Herein, we present a case of MSK who presented with recurrent urolithiasis and urinary tract infections in the 5th decade.

CASE PRESENTATION

A 53-year-old woman presented with dysuria and bilateral costovertebral pain radiating to the groin region. She had also similar complaints 2 months ago. Pyuria and hematuria were found on urine analysis. Urinary ultrasonography revealed hyperechogenic densities of multiple calculi in the calyces of both kidneys, and minimal pelvicaliectasis. Baseline biochemical analysis was presented in Table I. Accordingly, basic biochemical parameters including serum creatinine, calcium, phosphorus and parathormone levels were in the normal range. On 24-hour urine analysis, she had proteinuria of 160 mg/day, urinary calcium of 232.07 mg/day (100-300 mg/day) and uric acid of 374.90 mg/day (250-750 mg/day). Abdominal computed tomography was performed to better delineate the renal pathology and revealed multiple punctate calcifications aligned concentrically in the medulla of both kidneys (Figure 1A,B). On clinical and radiological grounds, the diagnosis of MSK was established. Escherichia coli sensitive to ciprofloxacin was grown at more than 100.000 colonies In urine culture. The patient's complaints were resolved with antibiotic treatment for the urinary tract infection. Thiazide and potassium citrate were not prescribed since hypercalciuria or hypocitraturia was not present. She is currently followed up by the Nephrology outpatient clinics.

Table I: Baseline biochemical parameters of the patient.

	Values	Reference Values
Urea (mg/dL)	33	17–43
Creatinine (mg/dL)	0.95	(0.7-1.4)
Uric acid (mg/dL)	5.1	(2.6-6)
Sodium (mEq/L)	139	(134-146)
Potassium (mEq/L)	5.2	(3.5-5.2)
Calcium (mg/dL)	9.7	(8.5-10.5)
Phosphorus (mg/dL)	3.0	(2.7-4.5)
Magnesium (mg/dL)	2.11	(1.6-2.6)
ALT (U/L)	17	(5-45)
25-OH Vitamin D (ng/mL)	9.80	(11-43)
PTH (pg/mL)	51.60	(12-88)
Total cholesterol (mg/dL)	317	(130-200)
Triglyceride (mg/dL)	193	(<30)
Total protein (g/dL)	7.43	(6.0-8.0)
Albumin (g/dL)	4.2	(3.2-5.5)
ESR (mm/h)	15	(0-20)
CRP (mg/dL)	0.46	(0-0.8)
WBC (/uL)	6800	(4000-11.000)
Hb (g/dL)	13.4	(12-18)

ALP: Alkaline phosphatase, **ALT:** Alanine aminotransferase, **CRP:** C-reactive protein, **ESR:** Erythrocyte sedimentation rate, **GGT:** Gamma glutamyl transferase, **Hb:** Hemoglobin, **LDH:** Lactate dehydrogenase, **PTH:** Parathyroid hormone, **WBC:** White blood cells

DISCUSSION

Medullary sponge kidney is a congenital disease characterized by extensive cystic dilatation of the medullary collecting tubules (6). MSK is usually an incidental finding in patients who are evaluated for infection, hematuria, or kidney stones. Genetic defect and pathogenesis of MSK has not been defined. MSK is usually clinically silent, however complications such as nephrocalcinosis, nephrolithiasis, urinary tract infection, renal tubular acidosis, and polyuria casued by impaired urine concentrating ability may occur (4,5-7). Cystic dilation of medullary collecting tubules causes urinary stasis which contributes to nephrolithiasis (8). MSK is found in 20% of patients with kidney stones and more than 70% of patients with MSK develop stones (7,9). Nephrolithiasis in MSK facilitates urinary tract infection(3).Dilatation of collecting tubules in MSK also plays role in recurrent pyelonephritis (10).

The classic radiologic appearance in intravenous pyelogram is the "bouquets of flowers" representing the collection of contrast material in dilated collecting tubules (7). CT scanning is also good at revealing papillary calcifications or hyperdense papillae (11,12).

Treatment of MSK includes antibiotics for acute pyelonephritis. A 24-hour urine tests (eg. calcium, citrate, uric acid, magnesium, sodium, oxalate, phosphate) for the evaluation of risk factors for nephrolithiasis may be very helpful in the management of metabolic factors contributing to nephrolitiasis. Thiazide diuretics together with potassium citrate may be prescribed to prevent the formation and progression of nephrocalcinosis/nephrolithiasis if hypercalciuria and hypocitraturia are present. In our case, since hypercalciuria or hypociraturia was not present, thiazide or potassium citrate was not prescribed. Patients with MSK are recommended to drink water in order to have a daily urinary output of about 2 liters (2,13).

In conclusion, although medullary sponge kidney is a congenital disease, it should be considered in the differential diagnosis of nephrocalcinosis, nephrolithiasis and recurrent urinary tract infections in older patients, even in the fifth decade.

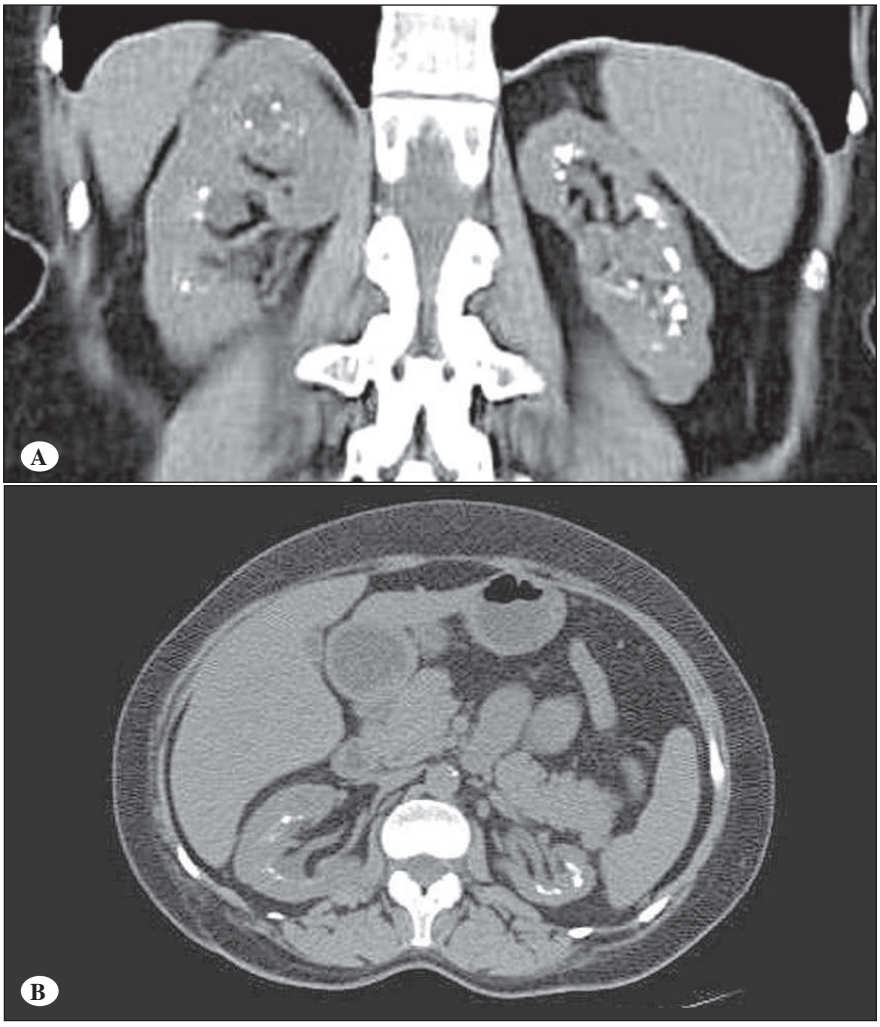

Figure 1: A,B) *Abdominal computed tomography sections of the patient with medullary sponge kidney revealing multiple punctuated calcifications aligned concentrically in the medulla of the kidneys.*

REFERENCES

1. 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
2. Fick GM, Gabow PA: Hereditary and acquired cystic disease of the kidney. Kidney Int 1994;46(4):951-964
3. Miano R, Germani S, Vespasiani G: Stones and urinary tract infections. Urol Int 2007;79(Suppl 1):32-36
4. Higashihara E, Nutahara K, Tago K, Ueno A, Niijima T: Medullary sponge kidney and renal acidification defect. Kidney Int 1984;25(2):453-459
5. Fabris A, Lupo A, Bernich P, Abaterusso C, Marchionna N, Nouvenne A, Gambaro G: Long-term treatment with potassium citrate and renal stones in medullary sponge kidney. Clin J Am Soc Nephrol 2010;5(9):1663-1668
6. Gambaro 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
7. Chu HY, Yan MT, Lin SH: Recurrent pyelonephritis as a sign of sponge kidney. Cleve Clin J Med 2009;76(8):479-480
8. O'Neill M, Breslau NA, Pak CY: Metabolic evaluation of nephrolithiasis in patients with medullary sponge kidney. JAMA 1981; 45:1233-1236
9. Gambaro G, Fabris A, Puliatta D, Lupo A: Lithiasis in cystic kidney disease and malformations of the urinary tract. Urol Res 2006; 34:102-107
10. 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
11. Levine E, Hartman DS, Meilstrup JW, Van Slyke MA, Edgar KA, Barth JC: Current concepts and controversies in imaging of renal cystic diseases. Urol Clin North Am 1997;24(3):523-543
12. Lang EK, Macchia RJ, Thomas R, Watson RA, Marberger M, Lechner G, Gayle B, Richter F: Improved detection of renal pathologic features on multiphasic helical CT compared with IVU in patients presenting with microscopic hematuria. Urology 2003;61(3):528-532
13. McPhail EF, Gettman MT, Patterson DE, Rangel LJ, Krambeck AE: Nephrolithiasis in medullary sponge kidney: Evaluation of clinical and metabolic features. Urology 2012;79(2):277-281