Bilateral Giant Ureteric and Staghorn Calculi in a Patient with Incomplete Distal Renal Tubular Acidosis

İnkomplet Distal Renal Tübüler Asidozlu bir Hastada Bilateral Dev Üreter Taşı ve Staghorn Taş

Staghorn Taş ve Renal Tübüler Asidoz / Staghorn Calculi and Renal Tubular Acidosis

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Özet

Akut pyelonefrit ve inkomplet distal renal tübüler asidoza (idRTA) bağlı geliştiği düşünülen bilateral dev üreter taşı, nefrokalsinozis ve sol böbrekte staghorn taş ile prezente olan, 18 yaşında bir olguyu sunuyoruz. Kalsiyum taşı tespit edilen hastalardan, sistemik asidoz yokluğunda idrar pH'sının devamlı olarak yüksek seyrettiği olgularda, idRTA'nın akılda tutulması gerektiğini vurgulamak istiyoruz.

Anahtar Kelimeler

Bilateral Dev Üreter Taşı; İnkomplet Renal Tübüler Asidoz; Staghorn Taş; Nefrokalsinozis

Abstract

We report a 18-year-old adult presenting with acute pyelonephritis, bilateral giant ureteral stones, nephrocalcinosis and left staghorn calculi most probably due to the underlying incomplete distal renal tubular acidosis (idRTA). Particularly, we want to underline that, idRTA should be kept in mind in the setting of calcium stone disease where urinary pH is persistently high in the absence of systemic acidosis.

Keywords

Bilateral Giant Ureteral Stone; Incomplete Renal Tubular Acidosis; Staghorn Calculi; Nephrocalcinosis

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Introduction

Distal renal tubular acidosis is characterized by a wide spectrum of biochemical abnormalities depending on the degree of acidification defect. idRTA term is used to define those who had fasting urine pH>5.3 with normal baseline systemic pH and bicarbonate levels [1]. As a consequence of ongoing acidification defect of urine, secondary to hypercalciuria and hypositraturia, nephrocalcinosis and nefrolithiasis are frequently seen in patients with idRTA [2]. Ureteral stones are generally sympthomatic and particularly those larger than 10 mm in diameter usually need intervention. A small portion of these stones, may reach even 10 cm in length or more than 50 gram in weight, following an asymptomatic period. These are defined as giant ureteral stones and usually have been reported as unilateral cases [3]. To our knowledge, there is no report presenting bilateral giant ureteral stones in the literature. Hereby, we report a case presenting with bilateral giant ureteral stones, nephrocalcinosis and left staghorn calculi most probably due to the underlying idRTA.

Case Report

A 18-year-old adult referred with the complaints of right flank pain persists for three days, fever over 38.5°C and chills. Physical examination revealed right costovertebral angle tenderness. In laboratory investigations, white blood cell was 19.500/mm3, serum blood urea nitrogen (BUN) was 48 mg/dl, serum creatinine was 1.7 mg/dl and CRP was 150 mg/dl. Urinalysis demonstrated microscopic hematuria and pyuria. He was diagnosed as acute pyelonephritis and empirical parenteral cephtriaxone antibiotherapy was administered after obtaining blood and urine cultures. Renal ultrasound revealed grade 3 hydronephrosis with perirenal fluid collection surrounding right kidney and bilateral hydroureter. Computed tomography revealed a giant ureteral stone 10.5 x 1.6 cm in size in right ureter, four ureteral stones with sizes of 4.0 x 2.5, 2.7 x 2.1, 2.3 x 0.9 and 1.7 x 0.8 cm in left ureter, left renal staghorn calculus and right medullary nephrocalcinosis (Figure 1a and 1b). Urine and blood cultures were negative and the patient has showed improvement on the 5th day of cephtriaxone. Double J ureter stent was placed to right ureter and the empiric antibiotherapy was continued for 14 days. A right ureterolithotomy was performed as first operation and stone was removed through a longitudinal incision (Figure 2). During the same session left pneumatic ureterolithotripsy was also performed followed by insertion of a double J stent for both ureters. Stone analysis revealed a composition of calcium oxalate (60 %) and magnesium ammonium phosphate (40 %). The patient experienced spontaneous removal of many small calculi and control plain radiography demonstrated several small residual stone fragments in the left lower ureter 1 month after the intervention. Soon after, we hospitalized the patient and performed the second operation; residual stone fragments were removed via bilateral endoscopic approach. At the same session left percutaneous nephrolithotomy and antegrade ureterolithotripsy were performed. Bacterial cultures of specimen taken from the stone matrix grew nothing. Vesicoureteral reflux and bilateral ureter obstruction were ruled out via control venous urography and cystography assessments.

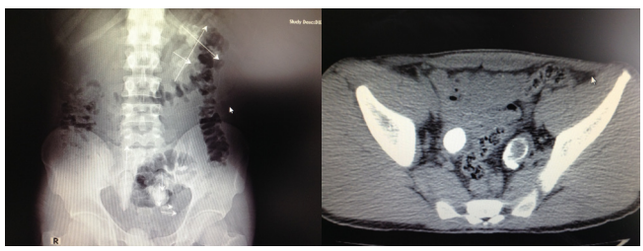

Figure 1. Plain radiography showing bilateral giant ureter stones (a). Abdominal computerized tomography demostrating right hydroureteronephrosis and perirenal reaction, left staghorn calculi and medullary nephrocalcinosis in right kidney. In addition, bilateral lower ureter stones exist in pelvic scans of tomography (b).

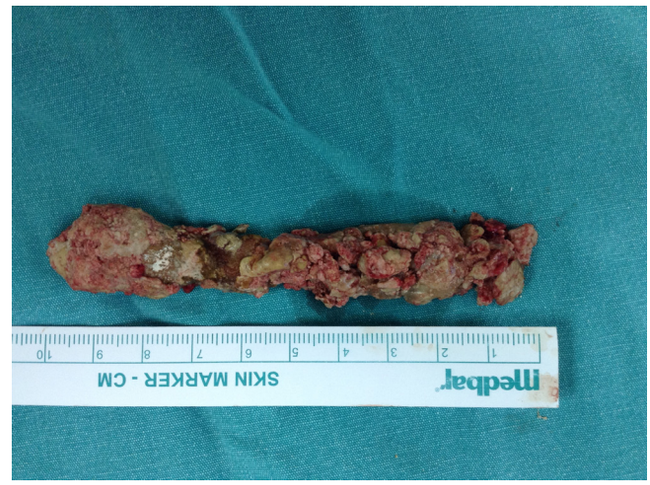

Figure 2. Removed right giant ureteral stone with a soft matrixt due to its struvite content.

Laboratory investigations were as follows; BUN: 34 mg/dl, serum creatinine: 1.4 mg/dl, sodium: 136 mmol/L, chloride: 99 mmol/L, potassium: 4.0 mmol/L, calcium: 9.2 mg/dl, phosphorus: 2.5 mg/dl, parathyroid hormone: 40 pg/ml, vitamin D level: 25.4 ng/ml. Venous blood gas analysis revealed pH: 7.40, PCO2: 42.9 mmHg, HCO3: 25.1 mmol/L. Metabolic evaluation of urine revealed a urine pH:7.0; with hypercalciuria (47.5 mg/dl) and hypocitraturia (72.5 mg/day). Additionally, a urinary acidification test [4] was performed with informed consent of the patient. After taking furosemide 40 mg and fludrocortisones 1 mg, the patient's urine pH remained over 5.3 (it was 6.1) and accordingly he was diagnosed as idRTA. Thereafter a prophylaxis treatment consisting of nitrofurantoin 100 mg/daily, indapamide 1.5 mg/daily, potassium citrate (60 mmol/day) and L-methionine (1000 mg/day) was started. However, the patient could not tolerate potassium citrate due to side effects, so fresh orange juice (500 ml/day) was recommended. One month after the prophylaxis, his laboratory results were as follows; serum potassium: 4.1 mmol/L, urine pH: 6.7, urinary citrate excretion: 365 mg/day (NR: 116-931) and urinary calcium excretion: 34.5 mg/dl (NR: 6.7-21.3). The patient is still under control and neither new stone formation nor urinary infection was observed during four month follow-up period.

Discussion

In patients with idRTA, a persistently high urine pH is generally observed but they are able to maintain net acid excretion under basal conditions probably due to increased ammonium secretion in proximal tubules unlike dRTA [5, 6]. They frequently suffer from recurrent nephrolithiasis, due to the combination of

hypercalciuria, low urine citrate, and high urine pH which favors crystallization [5, 6]. Although stone formation can be reduced to some extent by using potassium citrate, early diagnosis of idRTA is important. Due to side effects of short ammonium chloride test, we have used an easier, effective and well-tolerated alternative urinary acidification test by simultaneous administration of furosemide and fludrocortisones for diagnosis [4]. Consecutive urine samples were collected once an hour for the following four hours after drug intake and minimum pH was recorded to be 6.1 demonstrating the failure to acidify urine to pH less than 5.3. When normal serum pH was taken into consideration, the patient was diagnosed as idRTA which had probably led to hypercalciuria, hypocitraturia, and nephrocalcinosis. It also seems to have played a facilitator role on the recurrent urinary tract infections.

Unilateral giant ureteral stones have previosly been reported, however a concurrent renal stoghorn calculus is extremely rare [7]. To our knowledge, a case of idRTA presenting with bilateral giant ureteral stones accompanying with a staghorn calculi in kidney has not been reported previously. Unlike our case, renal stones seen in patients with idRTA are generally composed of calcium phosphate. We have observed a mixed stone composed of calcium oxalate and struvite in the setting of idTRA and we suggest that recurrent urinary infections had probably contributed to this unusual finding.

In recurrent stone formers with idRTA, increasing urinary citrate concentration could be aimed in order to prevent further deposition of calcium salts, without exceeding urinary pH of 7. Additionally, thiazide diuretics can also be used for management of patients in order to decrease renal calcium excretion [8]. Potassium citrate therapy may have an unfavorable effect on urinary infection rates by increasing urine pH. Actually, there is not a definite suggestion for the prophylaxis of struvite stones seen in idRTA patients, thus we have prescribed an individualized treatment for our patient. We added L-methionine [9] and nitrofurantoin to the treatment to avoid recurrent infections. It is encouraging that, we could achieve an increase in urinary citrate excretion without exceeding the threshold urine pH level and without recurrent stone formation or a new infection episode. The patient was not able to use potassium-citrate even in the initial dose (20 mmol/day) due to nausea and vomiting, so fresh orange juice was recommended as an alternative urinary citrate supplement [10]. Therefore we decided to continue to the prophylaxis regimen composed of fresh orange juice, indapamide, L-methionine and nitrofurantoin with previously mentioned daily doses.

We want to underline that, persistently high urine pH should not be accepted only as a consequence of recurrent urinary infections. idRTA should be kept in mind in the setting of calcium stones where urine pH is persistently above 5.5 in the absence of systemic acidosis.

Competing interests

The authors declare that they have no competing interests.

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