

Bilateral pyelolitotomy for cystine stones encircling the bilateral D-J stents in an infant presenting with anuria

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SUMMARY

Cystinuria is a hereditary disorder of cystine and dibasic amino acids (lysine, arginine, ornithine) transport across the luminal membrane of renal tubules and intestine, resulting in recurrent nephrolithiasis. Predominant clinical finding of this disease is the occurrence of recurrent cystine stones. Cystine stones frequently occur in the second or third decade of life with an occasional occurrence in infancy. Atypical symptoms of renal stones as anuria can become apparent in infancy. Bilateral cystine stones causing acute anuria in infants are extremely rare. We report the first case of bilateral pyelolitotomy performed for cystine stone and impacted bilateral double-J (DJ) stents in an infant presenting with anuria. In our patients DJ stents were removed, and a stone-free state was successfully achieved.

Key words: Anuria, cystine stone

ÖZET

Sistin taşları ile sarılmış bilateral D-J stentli anürik infanta bilateral pyelolitotomi yapılması

Sistinüri, sistin ve dibazik amino asitlerin (sistin, lizin, arjinin, ornitinin) böbrekler ve gastrointestinal sistemden taşınmasındaki defekt sonucu ortaya çıkan kalıtsal bir bozuktur. Bu hastalığın başlıca klinik bulgusu üriner sistemde rekürren sistin taşlarının oluşmasıdır. Sistin taşları genellikle yaşamın ikinci ve üçüncü dekadında ortaya çıkarken infantil dönemde ender olarak görülmektedir. Anüri gibi atipik böbrek taşı semptomları infantil dönemde ortaya çıkabilir. Infantil dönemde akut anüriye neden olan bilateral sistin taşı oldukça enderdir. Olgumuz, literatürde anüri ile başvuran sistin taşları ile sarılmış bilateral DJ stentli infanta bilateral pyelolitotomi yapılan ilk olgudur. Hastamızda DJ stentler çıkarılmıştır ve taşsızlık başarı ile sağlanmıştır.

Anahtar kelimeler: Anüri, sistin taşı

Normally developed 18-month-old boy referred to another hospital 9 month ago with 1 week history of vomiting and low urine output. Catheterisation released only 10 mL of urine. Serum creatinine was 4.8 mg/dL. X-ray examination of the abdomen showed one semi-opaque stone overlying the right kidney, near the renal pelvis, and two semi-opaque stones in the left kidney, one in the lower renal calyx and the other in the renal pelvis. Ultrasound (USG) reported bilateral multiple renal calculi with bilateral hydronephrosis, and 6 mm calculi in the left distal ureter. He had then become listless, vomited frequently, and for 24 hours did not pass urine. There was a known family history of cystinuria in his brother. In that hospital, bilateral DJ stent insertion under general anesthesia was tried but failed.

Bilateral percutaneous nephrostomy tubes were inserted, diuresis commenced, and the infant was stabilized with no need for dialysis. One month later, bilateral percutaneous nephrostomy tubes were removed after insertion of bilateral DJ stents. Then the patient was followed up monthly with urinary system USG. During the follow-up period of 3 months, size and number of stones increased, and bilateral DJ stents were tried to be removed without success due to stone encrustation. Eight months after DJ stent insertion, patient was referred to our institution for further treatment. Serum creatinine (0.4 mg/dL), potassium (4.1 mg/dL), and hematocrit values (31.5 %) were determined. Urine culture was sterile. Kidney-ureter-bladder (KUB) (Fig 1) X-ray revealed bilateral DJ stents, bilateral semi-opaque

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Figure 1. Kidney-ureter-bladder X-ray revealed bilateral DJ stents.

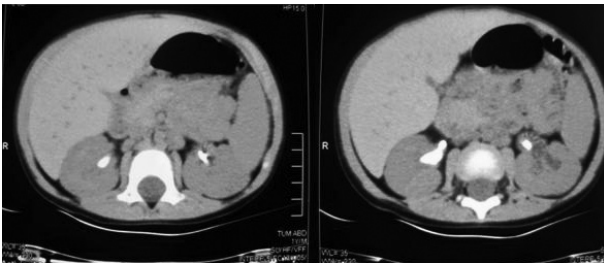


Figure 2. Abdominal computerized tomography revealed bilateral DJ stents.

renal pelvic stones, left proximal and distal ureter stones encircling the DJ stent. Abdominal computerized tomography (CT) (Fig 2) revealed bilateral DJ stents, bilateral renal pelvic stones, proximal and distal left ureter stones. Under general anesthesia, while the patient was in the right flank position, intercostal incision was made to reach renal pelvis. then through a pyelotomy incision stone in the right renal pelvis encircling the DJ stent was seen and pulled out from renal pelvis with a clamp. Intact stent with encircling stones were pulled out (Fig 3-4). The same procedure was applied for the left kidney after 1 month from the discharge. US examination was performed 1 week later and demonstrated bilateral normal-sized kidneys, without any residual stones or hydronephrosis. Stone analysis revealed cystine stones. A regimen of high-fluid intake and Thiola®

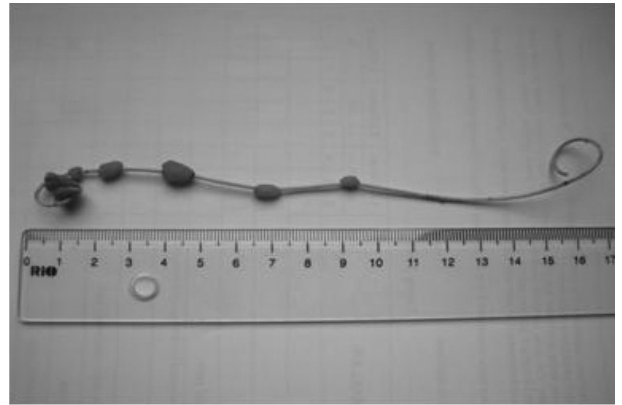


Figure 3. Intact stent with encircling stones were pulled out.

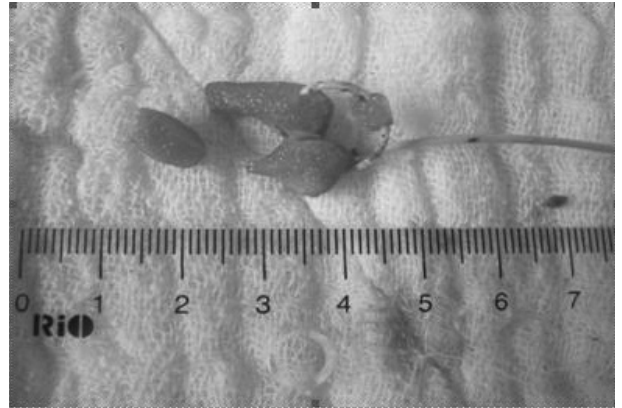


Figure 4. Intact stent with encircling stones were pulled out (closer view).

(tiopronin) was initiated. Follow-up ultrasound was performed at 6 months post-operatively and revealed bilateral normal kidneys without any evidence of stones. His creatinine was 0.5 mg/dL.

DISCUSSION

Atypical complaints of renal stones such as anuria can be more common in younger children. Bilateral cystine stones causing acute anuria in infants are extremely rare ⁽¹⁻³⁾. Arguments on the most efficient management of forgotten stents still remain unsolved. In general, combining endourologic approaches, even with open surgical management, is necessary to remove encrusted or fractured stents. Although retrograde intracorporeal stone disintegration is a current choice of this situation, few studi-

es have introduced algorithms for the management of retained indwelling ureteral stents with encrustation. Bultitude et al. and Sancaktutar AA et al. advised using endourological techniques to remove impacted stents with mild encrustation ⁽⁴⁻⁶⁾. In cases of ureterorenoscopic failure or marked encrustation, initial adjunctive use of extracorporeal shock wave lithotripsy (ESWL) with ureteroscopic lithotripsy for the proximal portion of D-J stent may be efficacious ⁽⁷⁾. Percutaneous nephrolithotomy (PCNL) is often used as a second-line treatment, particularly in cases with a large proximal stone burden or ESWL failure ^(8,9). In fact, the size of the stone burden may determine the specific approach. Nevertheless, forgotten stents shouldn't be force for removal with traction, because severe ureteral injury or stent fragmentation may occur during forced traction ⁽¹⁰⁾. It is believed that ESWL may offer less help in cases with a large stone burden and cystine stones bigger than a 1.5 cm ⁽¹¹⁾. Therefore, ESWL was not used as a initial treatment in our patient for renal stones. Due to the need of very small calibre equipments and large stone burden, we preferred thee open technique to pull the stents out completely in one session.

CONCLUSION

Although minimally invasive techniques such as PCNL, flexible ureteroscopy and laser lithotripsy or ESWL has been successfully used for the management of impacted DJ stents, open surgery is also a good treatment choice and an effective treatment modality for seleceted patients. We believe that for small infants with a large stone burden not amenable to extracorporeal shock wave lithotripsy or ureteroscopy, open surgery is a safe and efficacious modality, and should be an alternative method to percutaneous approach in selected patients.

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