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Takashi Kawahara, Hiroki Ito, Hideyuki Terao, Hiroji Uemura, Masahiro Yao, Junichi Matsuzaki

ABSTRACT

Introduction: Ureteroscopy-assisted retrograde nephrostomy (UARN) makes it is possible to continuously visualize dilation of the ureter from the site of puncture to insertion of the nephroaccess sheath, with minimal complications and effective for percutaneous nephrolithotripsy (PCNL).

Case Report: A 43-year-old male with cerebrotendinous xanthomatosis (CTX) was referred to our department to undergo PCNL for a diagnosis of a right kidney stone. In July 2012, the patient was admitted to our department for PCNL to treat a right renal staghorn calculus. We performed PCNL using the technique of UARN. The operative time was 195 minutes, and the chemical composition of the stone was calcium oxalate.

Conclusion: UARN is effective for the case who have large renal stone with complications of CTX.

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CASE REPORT

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Keywords: Cerebrotendinous xanthomatosis, Percutaneous nephrolithotripsy (PCNL), Renal stone, Ureteroscopy-assisted retrograde nephrostomy (UARN), Ureteroscopy

How to cite this article

Kawahara T, Ito H, Terao H, Uemura H, Yao M, Matsuzaki J. Ureteroscopy-assisted retrograde nephrostomy (UARN) for the patients with cerebrotendinous xanthomatosis. Int J Case Rep Images 2015;6(12):777–779.

doi:10.5348/ijcri-2015126-CR-10587

INTRODUCTION

We previously described the use of ureteroscopy-assisted retrograde nephrostomy (UARN), which makes it is possible to continuously visualize dilation of the ureter from the site of puncture to insertion of the nephro access sheath, with minimal complications [1, 2]. We herein report the first case in which UARN was applied for percutaneous nephrolithotripsy (PCNL) in a patient with cerebrotendinous xanthomatosis (CTX).

CASE REPORT

A 43-year-old male with CTX was referred to our department to undergo nephrolithotripsy for a diagnosis of a right kidney stone. In July 2012, the patient was admitted to our department for PCNL to treat a right renal staghorn calculus. We have previously reported the technique of UARN and performed this procedure in the present case, as described below [1].

Briefly, under general and epidural anesthesia, the patient is placed in a modified Valdivia position (Galdakao modified Valdivia position). Flexible URS (Flex-X2, Tuttlingen, Karl Storz, Germany) insertion is carried out under an inserted ureteral access sheath in the ureter after ensuring that the rigid URS (Uretero-roscope, Karl Storz, Tuttlingen, Germany) does not encounter either ureteral stenosis or stones. The UAS device includes a

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Received: 07 July 2015

Accepted: 22 September 2015

Published: 01 December 2015

12/14-Fr (inner/outer) 35-cm ureteral access sheath (UAS) (Flexor®, COOK Urological, IN, USA) or an 11/13-Fr 46-cm UAS (Navigator® 11 Fr 46 cm, Boston Scientific, MA, USA) placed in the ureter. We carefully observe the target calculi and select the appropriate renal calyx to puncture. Thereafter, a Lawson retrograde nephrostomy puncture wire (Lawson Retrograde Nephrostomy Wire Puncture Set, COOK Urological, IN, USA) is carefully placed into the flexible URS. The URS approaches the desired renal calyx a second time, and the route from the renal calyx to the exit site on the skin is then confirmed under fluoroscopy.

In order to avoid injury to the spleen, liver, intestines or pleural cavity, a puncture is then performed under ultrasonography after rechecking the preoperative computed tomography (CT) scan. The puncture wire is easily passed through the muscle and “tents” the skin at the posterior axial line. The skin is then incised, and the needle is delivered. Next, the dilator is placed using the puncture wire. A 24-Fr percutaneous nephro-access sheath (X-Force®N30 Nephrostomy Balloon Dilation Catheter, BARD, USA) is passed over the balloon into the calyx under ureteroscopic and fluoroscopic guidance, after which the balloon is removed. Calculus fragmentation is performed using the Swiss LithoClast® pneumatic lithotripter (EMS, Nyon, Switzerland) through a rigid nephroscope (percutaneous nephroscope, Karl Storz, Tuttlingen, Germany).

The operative time was 195 minutes, and the chemical composition of the stone was calcium oxalate.

DISCUSSION

Cerebrotendinous xanthomatosis (CTX) is a rare neurological disease characterized by the accumulation of cholesterol and cholestanol in the brain and tendons due to a mutation in the sterol 27-hydroxylase gene (CYP27A1). More than 400 cases have been previously described worldwide in medical literature [3]. The clinical signs of CTX develop gradually during the patient's lifetime. Some patients exhibit mental impairment in childhood, whereas others do not display any neurological symptoms, even at an older age, with the only sign of the disease being Achilles tendon xanthoma formation [4]. Premature atherosclerosis and cardiac complications have been reported to be systemic manifestations of CTX [5]. In some cases, myocardial infarction is the cause of sudden death.

The mean age of symptom onset is estimated to be between 14 and 19 years of age, with a mean delay in diagnosis of 17–19 years. Early typical signs include chronic diarrhea, juvenile bilateral cataracts, Achilles (or other) tendon xanthomas, psychomotor retardation, cognitive impairment with learning difficulties or mental retardation, cerebellar ataxia and epilepsy, which can affect up to 50% of patients [3]. Other late-stage neurological features comprise pyramidal tract signs

with extensor plantar responses, progressive spastic paraplegia, progressive cerebellar ataxia and dysarthria, nystagmus, peripheral polyneuropathy with distal muscle wasting and, more rarely, movement disorders, such as parkinsonism and palatal myoclonus, and, in patients with advanced disease, pseudobulbar and bulbar syndromes [3]. As the disorder progresses, the patient can become incapacitated due to motor dysfunction, with premature death often occurring as a result of advancing neurological deterioration. Treatment is based on the administration of chenodeoxycholic acid. The early detection of CTX is essential, as drug therapy can limit further damage to the CNS and prevent progression of tendon xanthomas, as well as lead to regression in early cases.

Although the effectiveness of shock wave lithotripsy (SWL) and flexible ureteroscopic lithotripsy has been reported, patients with a greater stone burden are thought to be better candidates for PCNL. Previously, nephrostomy was usually transcutaneously performed under ultrasonographic and/or fluoroscopic guidance. With the arrival and development of flexible ureteroscopy (URS), both observation and manipulation in the renal pelvis can be easily achieved. We previously described a new technique for performing UARN. This procedure involves less radiation exposure and a shorter operative time than the previous percutaneous nephrostomy method. Our technique represents an additional new option for performing PCNL in patients with a non-dilated intrarenal collecting system.

Due to the continuous visualization offered by URS, the use of UARN facilitates PCNL to be performed more safely and easily [2]. This report describes the first case of nephrolithiasis treated with UARN in a patient with CTX.

CONCLUSION

Ureteroscopy-assisted retrograde nephrostomy (UARN) is effective for the case who have large renal stone with complications of cerebrotendinous xanthomatosis (CTX).

Author Contributions

Takashi Kawahara – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Hiroki Ito – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Hideyuki Terao – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Hiroji Uemura – Analysis and interpretation of data,

Revising it critically for important intellectual content,
Final approval of the version to be published
Masahiro Yao – Analysis and interpretation of data,
Revising it critically for important intellectual content,
Final approval of the version to be published
Junichi Matsuzaki – Analysis and interpretation of data,
Revising it critically for important intellectual content,
Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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REFERENCES

1. Kawahara T, Ito H, Terao H, et al. Ureteroscopy assisted retrograde nephrostomy: a new technique for percutaneous nephrolithotomy (PCNL). *BJU Int* 2012 Aug;110(4):588–90.
2. Kawahara T, Ito H, Terao H, et al. Effectiveness of ureteroscopy-assisted retrograde nephrostomy (UARN) for percutaneous nephrolithotomy (PCNL). *PLoS One* 2012;7(12):e52149.
3. Fraidakis MJ. Psychiatric manifestations in cerebrotendinous xanthomatosis. *Transl Psychiatry* 2013 Sep 3;3:e302.
4. Björkhem I. Cerebrotendinous xanthomatosis. *Curr Opin Lipidol* 2013 Aug;24(4):283–7.
5. Dotti MT, Mondillo S, Plewnia K, Agricola E, Federico A. Cerebrotendinous xanthomatosis: evidence of lipomatous hypertrophy of the atrial septum. *J Neurol* 1998 Nov;245(11):723–6.

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