SCIE DOI: 10.5505/jkartaltr.2015.02360 | 10.14744/scie.2017.02360
South. Clin. Ist. Euras. 2016;27(2):138-141

Retrocaval Ureter: A Rare Congenital Anomaly

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Submitted: 21.04.2015
Accepted: 28.04.2015

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Keywords: Hydronephrosis; inferior vena cava; ureter.

ABSTRACT

Objective: Described in this study is surgery to treat retrocaval ureter, a rare congenital anomaly, and relevant literature findings.

Methods: Data on patients who underwent surgery in the clinic between January 2003 and January 2015 were reviewed retrospectively. Three patients who were operated on for retrocaval ureter were included in the study. Age, sex, laterality, symptoms, and degree of hydronephrosis were analyzed. Open ureteroureterostomy with double J stent insertion was performed on all 3 patients. Double J stents were removed after 3 weeks and patients were evaluated at postoperative 3 months.

Results: Mean age of patients was 17.66 years (range: 13–24 years). Two patients were male, 1 was female. All cases had right-sided retrocaval ureter. Proximal ureteral dilatation and reverse J images were seen on intravenous pyelograms (IVPs) of all patients before surgery. On postoperative third month evaluation, ureteral drainage was normal, proximal ureteral dilatation had regressed and patients were asymptomatic.

Conclusion: Open surgery for retrocaval ureter is effective and preferred treatment modality with high success rate.

INTRODUCTION

Retrocaval (circumcaval) ureter is a rarely seen congenital condition. It is a developmental anomaly of ureter localized on posterolateral aspect of inferior vena cava (IVC). Incidence is 1/1000.[1–3] Although it is an inborn pathology, diagnosis is usually made at 30 to 40 years of age.[4,5] Generally, it is seen on the right side. Most patients demonstrate symptoms caused by ureteral obstruction.[6,7] Pain felt in right lumbar region, recurrent urinary system infection, and hematuria are frequently reported admission complaints. On intravenous pyelograms (IVPs), deformities seen in the form of inverted "J" or "S" and proximal ureteral dilatation with medial deviation should suggest presence of retrocaval ureter. Surgical treatment is required in the presence of retrocaval ureter so as to prevent urinary tract infection, stone formation, and loss of renal function.[4,8,9]

In this study, data of patients who had been operated on with indication of congenital retrocaval ureter were retrospectively evaluated and experience in diagnosis and treatment of retrocaval ureter is presented.

MATERIAL AND METHODS

Patients who underwent surgery in the clinic between January 2013 and January 2015 were retrospectively screened and patients operated on with diagnosis of retrocaval ureter were included in the study. Study protocol was approved by the Ethics Committee of Kartal Dr. Lütfi Kırdar Training and Research Hospital. Age, gender, side of affected kidney, admission symptoms, and grade of hydronephrosis were recorded.

Patients whose urinary system ultrasonographic (US) examination revealed hydronephrosis underwent IVP examination for further evaluation of urinary system. Patients whose IVP established diagnosis of retrocaval ureter underwent surgical treatment.

Standard open ureteroureterostomy was performed through an extended flank incision. Ureter with dilated proximal segment that crossed IVC from posterior aspect and coursed in medial direction was dissected. Suspension sutures were placed proximally and distally to the point where ureter crossed IVC, and it was brought in front of vena cava. Cut ends of ureter were spatulated, and end-to-end anastomosis was performed over double J catheter using 4–0 gauge Vicryl sutures (Figure 1). On postoperative third day, transurethral Foley catheters were removed, and double J catheters were removed at third week. Patient symptoms were evaluated at third month. Follow-up IVPs were obtained in order to evaluate grade of hydronephrosis and ureteral drainage.

RESULTS

A total of 3 patients (male: n=2; female: n=1) with mean age of 17.66 years (range: 13–24 years) who were operated on in the clinic with diagnosis of retrocaval ureter were included in the study. All cases had right-sided retrocaval ureters. Demographic findings are provided in Table 1.

IVPs of all patients demonstrated presence of hydronephrosis, dilatation of proximal ureteral segment, and typical inverted "J" image (Figure 2a). No urinary stones were observed. History of urologic surgery was not available.

It was observed during open ureteroureterostomy of all patients that dilated proximal segment of right ureter crossed IVC from behind and coursed medially (Figure 2b). No perioperative complications developed. All patients during preoperative period were asymptomatic at third postoperative month evaluation. IVPs revealed normal ureteral drainage and regression of dilatation (Figure 2c).

DISCUSSION

Retrocaval ureter is a rarely seen developmental abnormality of IVC resulting in deviation of ureter from its normal anatomical position. In normal embryological development, connection between right subcardinal vein and supracardinal vein regresses. Disruption of transformation of supracardinal vein into IVC has been held responsible for formation of the retrocaval right ureter.[10] Incidence is 1/1000, and it is seen 3 times more frequently in men.

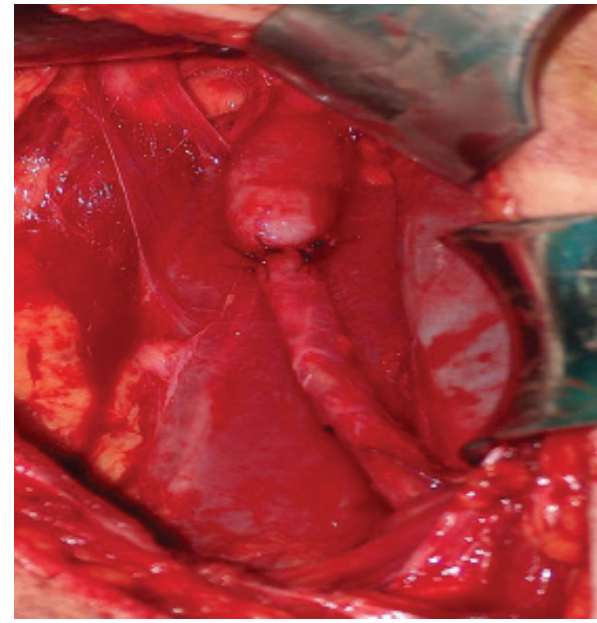

Figure 1. Appearance of ureter after anastomosis.

It is generally observed on the right side.[11,12] Left-sided retrocaval ureter is very rare. Association with Goldenhar syndrome and IVC duplication has been reported.[13] In all present cases (male: n=2; female: n=1), retrocaval ureter was localized on right side, consistent with most reports in the literature.

Diagnosis is usually made in the third or fourth decade of life. Time of diagnosis typically corresponds to gradual development of hydronephrosis, which results in diagnosis of these patients at an advanced age when they become symptomatic.[3,5] Though very rarely, symptomatic patients in the pediatric age group have been reported. Symptoms are related to hydronephrosis that develops due to ureteral obstruction; however, retrocaval ureter is not always associated with symptoms of obstruction. Frequent admission symptoms include abdominal pain localized in right lumbar region, recurrent urinary system infection, and hematuria.[14] Two patients in present study who were in second decade of life presented with lumbar pain, and the remaining patient was in third decade of life and presented with complaint of hematuria.

Dilated, tortuous, proximal ureter demonstrating medial deviation on US may indicate presence of retrocaval

Table 1. Demographic characteristics of the patients

No	Age	Gender	Side	Symptom	Grade of hydronephrosis
1	16	Female	Right	Lumbar pain	Grade 3
2	13	Male	Right	Hematuria	Grade 3
3	24	Male	Right	Lumbar pain	Grade 4

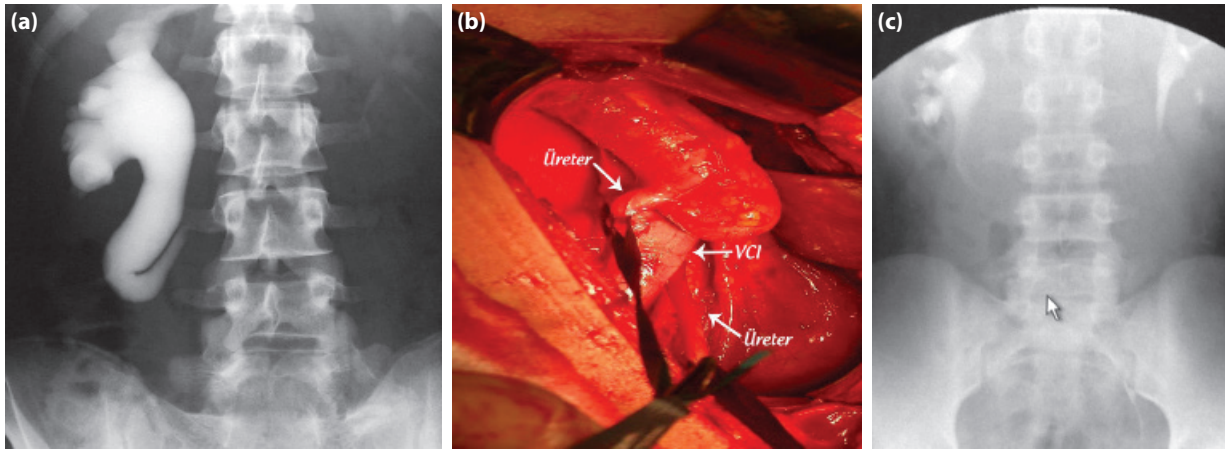

Figure 2. (a) Intravenous pyelogram image of right hydronephrosis, dilated proximal ureter, and inverted "J"; **(b)** Intraoperative appearance of dilated proximal ureter and pelvis (ureter coursing behind inferior vena cava); **(c)** Intravenous pyelogram at postoperative third month.

ureter. Diagnosis is usually made based on intravenous or retrograde pyelographic examinations. Contrast-enhanced computed tomography (CT) and magnetic resonance urography are other noninvasive methods that can be used in diagnosis of retrocaval ureter.[2] The advantage of retrograde pyelography is ability to demonstrate different areas of stenosis and obstruction that cannot be observed in preoperative radiological studies.[12] Since in present cases diagnostic inverted "J" images were seen on IVPs, retrograde pyelograms were not necessary; however, in cases where obstruction cannot be demonstrated and definitive diagnosis cannot be made, retrograde pyelograms should be obtained.

Bateson and Atkinson detected 2 types of retrocaval ureter based on intravenous urogram images. Type I (low loop) is more frequently seen. Inverted "J" fishhook appearance signifies severe or moderate degree of dilated proximal ureter. Type 2 (high loop) is less frequently seen and medial deviation is less severe. On intravenous urograms, sickle-shaped ureter and mild degree of hydronephrosis can be observed.[1,2] Present cases were all Type I retrocaval ureter.

Surgical intervention is required when patient is symptomatic or when function of affected kidney deteriorates. Long-term follow-up of asymptomatic patients is recommended with periodic US and diuretic renograms to allow for early intervention and prevention of further impairment of renal function.[11]

Retrocaval ureter may be repaired with either open or laparoscopic surgery. Surgical intervention involves resection of retrocaval segment and re-anastomosis of ureter in front of IVC. Open surgery has a higher success rate, but requires a wide skin incision, causes greater postoperative pain, and prolongs the healing process. Laparoscopic approach is minimally invasive treatment modality with less postoperative pain and shorter recovery period.[1,2,4] Laparoscopic surgery is especially preferred for young female patients because of its cosmetic advantages; however, longer operative time is a disadvantage of this procedure. Some clinics in Turkey do not use laparoscopic procedure in their daily routine practice. Therefore, open surgery is still the preferred method for management of retrocaval ureter.

REFERENCES

1. Rao J, Yang J, Liu Z, Wang L, Liu L, Yin Z, et al. Right retrocaval ureter and left nutcracker syndrome: a case report. Urology 2008;71:1226.e9–11.
2. Uthappa MC, Anthony D, Allen C. Case report: retrocaval ureter: MR appearances. Br J Radiol 2002;75:177–9.
3. Shigeta M, Nakamoto T, Nakahara M, Hiromoto N, Usui T. Horseshoe kidney with retrocaval ureter and ureteropelvic junction obstruction: a case report. Int J Urol 1997;4:206–8.
4. Tobias-Machado M, Lasmar MT, Wroclawski ER. Retroperitoneoscopic surgery with extracorporeal uretero-ureteral anastomosis for treating retrocaval ureter. Int Braz J Urol 2005;31:147–50.
5. Mitchell J, Stahlfeld KR, Cercone RG. Retrocaval ureter with intestinal malrotation. Urology 2003;62:142–3.
6. Ishitoya S, Arai Y, Waki K, Okubo K, Suzuki Y. Left retrocaval ureter associated with the Goldenhar syndrome (branchial arch syndrome). J Urol 1997;158:572–3.
7. Rubinstein I, Cavalcanti AG, Canalini AF, Freitas MA, Accioly PM. Left retrocaval ureter associated with inferior vena caval duplication. J Urol 1999;162:1373–4.
8. Wang LT, Lo HC, Yu DS, Sun GH, Wu CC, Fong CJ. Ureteral obstruction caused by a duplicated anomaly of inferior vena cava. Int J Urol 2005;12:842–4.
9. Salomon L, Hoznek A, Balian C, Gasman D, Chopin DK, Abbou CC. Retroperitoneal laparoscopy of a retrocaval ureter. BJU Int 1999;84:181–2.
10. Bass JE, Redwine MD, Kramer LA, Huynh PT, Harris JH Jr. Spec-

trum of congenital anomalies of the inferior vena cava: cross-sectional imaging findings. Radiographics 2000;20:639–52.
11. Soundappan SV, Barker AP. Retrocaval ureter in children: a report of two cases. Pediatr Surg Int 2004;20:158–60.
12. Dogan HS, Oktay B, Vuruskan H, Yavascaoglu I. Treatment of retrocaval ureter by pure laparoscopic pyelopyelostomy: experience on 4 patients. Urology 2010;75:1343–7.
13. Acharya SK, Jindal B, Yadav DK, Singha S, Bagga D. Retrocaval ureter: a rare cause of hydronephrosis in children. J Pediatr Surg 2009;44:846–8.
14. Pais VM, Strandhoy JW, Assimos DG. Pathophysiology of urinary tract obstruction. In: Wein AJ, Kavoussi LR, Novick AC, Partin AW, Peters CA, editors. Campbell-Walsh Urology. Vol 2. Philadelphia, PA: Saunders-Elsevier 2007. p. 1195–226.

Retrokaval Üreter: Doğuştan Nadir Bir Anomali

Amaç: Bu çalışmada, nadir bir doğuştan anomali olan retrokaval üreter nedeniyle kliniğimizde cerrahi tedavi uygulanan olgulardaki deneyimimizi literatür eşliğinde sunmayı amaçladık.

Gereç ve Yöntem: Kliniğimizde Ocak 2003 ve Ocak 2015 tarihleri arasında ameliyat edilen hastalar geriye dönük olarak tarandı. Çalışmaya retrokaval üreter tanısı ile ameliyat edilen üç hasta dahil edildi. Çalışmaya alınan hastaların yaş, cinsiyet, böbrek tarafı, başvuru semptomları ve hidronefroz dereceleri kayıt edildi. Tüm hastalara açık üreteroüreterostomi+double J stent uygulandı. Hastaların ameliyat sonrası üçüncü haftada double J kataterleri çekildi ve ameliyat sonrası üçüncü ayda hastalar tekrar değerlendirildi.

Bulgular: Hastaların yaş ortalaması 17.66 (dağılım, 13–24 yıl) idi. Hastaların ikisi erkek, biri kadındı ve tüm olgular sağ taraf yerleşimli idi. Tüm hastaların intravenöz pyelogramlarında proksimal üreterde dilatasyon ve tipik ters "J" görünümü mevcuttu. Ameliyat sonrası üçüncü ayda tüm hastaların semptomsuz olduğu, üreteral drenajın normal olduğu, dilatasyonun gerilediği tespit edildi.

Sonuç: Retrokaval üreter tedavisinde açık cerrahi günümüzde yüksek başarı oranları ile tercih edilen etkili bir yöntemdir.

Anahtar Sözcükler: Hidronefroz; vena kava inferior; üreter.