

Retrocaval Ureter

A Study of 13 Cases

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Introduction: The aim of this study was to report our 23-year experience in the diagnosis and treatment of retrocaval ureter.

Materials and Methods: Data from 13 patients with retrocaval ureter were reviewed. Intravenous urography and retrograde pyelography had been used for confirming the diagnosis. All of the patients had been symptomatic and undergone surgery. A control intravenous urography had been performed 6 months postoperatively.

Results: The mean age of the patients was 23 years (range, 12 to 37 years). Twelve patients (92.3%) were men. The clinical manifestations were pyelonephritis in 7 (53.8%), right flank pain in 4 (30.8%), gross hematuria in 1 (7.7%), and ureteral calculus in 1 (7.7%). All of the patients had type 1 right-sided retrocaval ureter. Associated anomalies were seen in none of the patients. The control intravenous urography showed improvement of renal function.

Conclusion: In our patients, the most common cause of referral was pyelonephritis. In symptomatic cases, operation is needed and can improve renal function.

Keywords: ureter, retrocaval, ureteroureterostomy, urogenital abnormalities

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INTRODUCTION

Retrocaval ureter is a rare congenital abnormality in association with upper urinary tract obstruction and usually has an S-shape or fishhook appearance on intravenous urography (IVU) that is due to the passage of the ureter posterior to the inferior vena cava (IVC). Congenital anomalies that result in the obstruction of the ureter are extremely rare; however, retrocaval ureter is the most common anomaly with a venous cause.⁽¹⁾ It is also called circumcaval or postcaval ureter.⁽²⁾ The anomaly is usually observed in the right side and in some cases (such as patients with situs inversus) it may be left sided. The prevalence of the disease is reported to be 1 in 1000 live births.⁽¹⁾ Although

the anomaly is congenital, patients become symptomatic in their 3rd or 4th decade of life.⁽¹⁾ In symptomatic cases, surgical intervention is often required.⁽¹⁻³⁾ We report 13 patients with this anomaly during our 23-year experience in the diagnosis and treatment of the retrocaval ureter.

MATERIALS AND METHODS

Medical records of 13 patients with retrocaval ureter who had been treated in Qaem Hospital (Mashhad) between 1983 and 2005 were reviewed. The definite diagnosis was made by IVU and retrograde pyelography. The type of the retrocaval ureter was identified according to the classification by Bateson and Atkinson.⁽³⁾ Intravenous urography had been performed for all

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patients 6 months postoperatively, as well. All patients were symptomatic and had undergone surgery. Age, sex, reason for referral, hospital staying, treatment modality, treatment outcome, and associated anomalies were collected from the patients' data sheets.

RESULTS

A total of 13 patients had retrocaval ureter. The median age of the patients was 23 years (range, 12 to 37 years). Twelve patients (92.3%) were men and 1 (7.7%) was a woman. The reason for seeking treatment was pyelonephritis in 7 patients (53.8%), right flank pain in 4 (30.8%), gross hematuria in 1 (7.7%), and ureteral calculus in 1 (7.7%). The mean hospital staying was 3.24 days (range, 3 to 4 days).

No associated anomaly was seen in these patients. The retrocaval ureter was type 1 and right-sided in all of the patients. We performed end-to-end ureteroureterostomy through an extraperitoneal incision on the 12th rib in all patients. On the control

IVU performed 6 months postoperatively, there were no remarkable findings and no complication occurred during the follow-up (Figures 1 and 2).

DISCUSSION

Retrocaval ureter was first reported by Hochstetter in 1893.⁽⁴⁾ Normally, IVC originates from the supracardinal and subcardinal veins inferior and superior to the kidney, respectively. If the IVC inferior to the kidney is formed by subcardinal vein, it will be located anterior to the ureter and will form a retrocaval ureter. There are two types of retrocaval ureter: type 1 which is more prevalent and has an S-shape or fishhook appearance, and type 2 which is sickle shaped.⁽³⁾ In radiographic studies, all of our patients had type 1 pattern of the retrocaval ureter.

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Abnormal development of the IVC is generally considered as the etiology of the retrocaval ureter;



Figure 1. **Left,** Intravenous urography before surgery for retrocaval ureter in a 12-year-old boy. Severe dilatation and S-shape pattern of the ureter is seen in the right side. **Right,** Six-month postoperative IVU in the same patient. Decreased hydronephrosis and correction of the ureter pathway is seen.

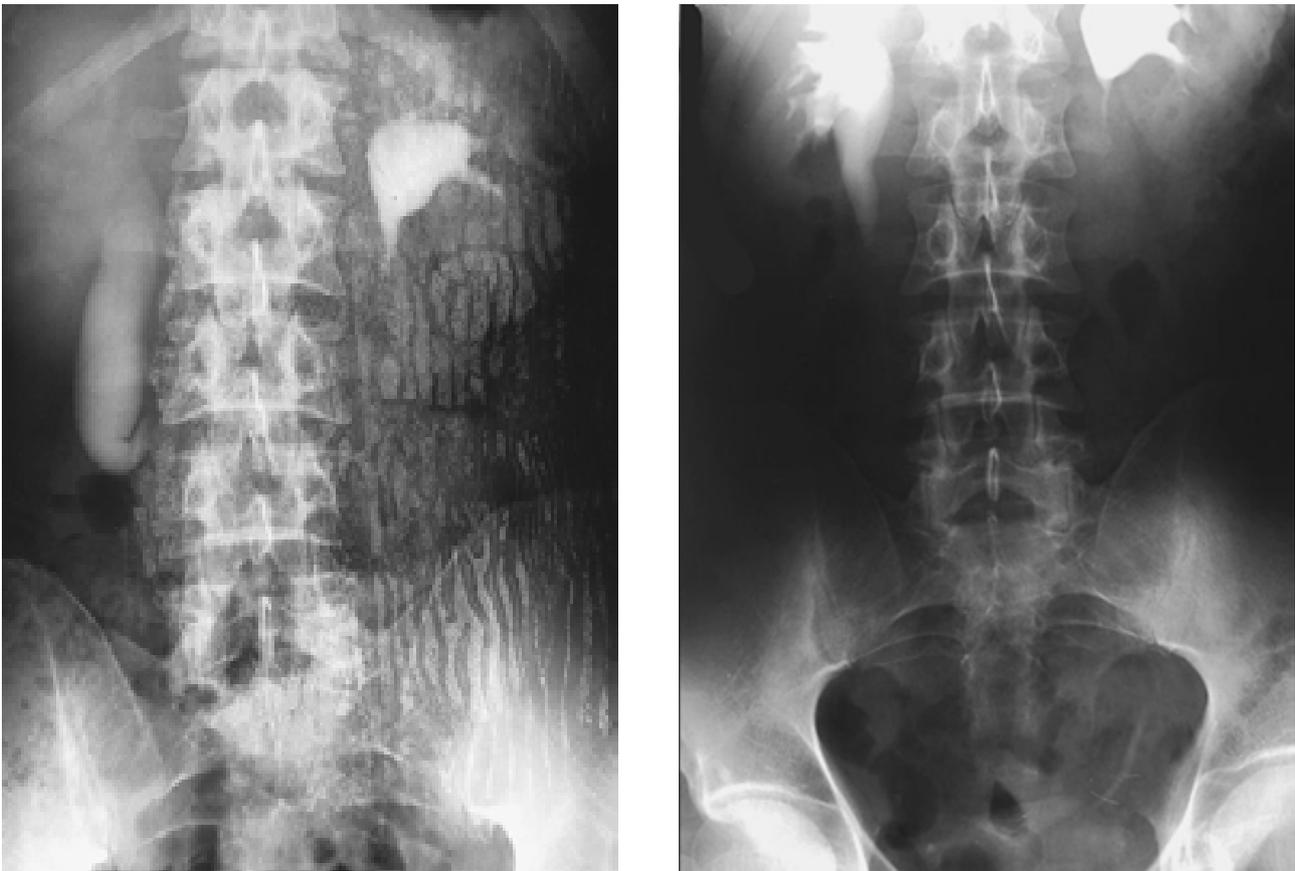


Figure 2. Left, Intravenous urography before surgery for retrocaval ureter in a 24-year-old man. Dilatation and hydronephrosis exist in the right side. Right, Postoperative IVU of the same patient. Decreased dilatation and improved renal function is seen.

however, maternal exposure to diethylene glycol monomethyl ether (an industrial solvent) during fetal period is proposed to be a probable cause.⁽⁵⁾ None of our patients had the history of such exposure. Retrocaval ureter is almost always right sided; however, in cases with situs inversus or duplication of the IVC, it may be seen in the left side.^(2,6) In our study, all of the patients had right-sided retrocaval ureters. The ratio of men to women is 2.8:1 in clinic.⁽⁷⁾ In our patients, however, this rate was 12:1. Patients usually present in their 3rd or 4th decades of life.⁽⁸⁾ The median age of our patients was 23 years, similar to the age mentioned in the literature. Retrocaval ureter may be asymptomatic or cause symptoms such as flank pain, urinary tract infection, hematuria, or calculus formation.^(4,9) Other disorders that have been reported to be associated with the retrocaval ureter are retroperitoneal fibrosis, carcinoma of the ureter, and renovascular hypertension.⁽¹⁰⁻¹²⁾ The referral reasons in our patients were pyelonephritis (the most common manifestation), right flank pain, gross hematuria,

and ureteral calculus. Associated anomalies with retrocaval ureter are reportedly up to 21% and are mainly related to the cardiovascular and urogenital systems (including horseshoe kidney, ureteropelvic junction obstruction, congenital lack of the vas deferens, hypospadias, extra vertebra, diverticulum, anterior urethral calculus, kidney agenesis, syndactyly in both feet, intestinal malrotation, and Goldenhar syndrome.⁽¹³⁻²⁰⁾ None of these anomalies was seen in our patients.

Retrocaval ureter has been previously diagnosed by IVU and retrograde pyelography, but nowadays, CT scan is the best modality for diagnosis.^(4,21) Diagnosis of the retrocaval ureter has also been reported by technetium Tc 99m diethylenetriamine pentaacetic acid scan, technetium Tc 99m methylene diphosphonate scan, and magnetic resonance imaging.⁽²²⁻²⁴⁾ In our series, the diagnosis was made based on IVU and retrograde pyelography.

Asymptomatic cases of retrocaval ureter do not need surgery,⁽⁹⁾ but symptomatic patients

generally need surgical intervention which is mainly ureteroureterostomy.⁽⁴⁾ In all 13 patients, a moderate to severe hydronephrosis was present and all of them were symptomatic. Thus, they all required surgical intervention. Intravenous urography, performed 6 months after ureteroureterostomy, revealed considerable improvement. Laparoscopic correction of the retrocaval ureter is also reported which may be transperitoneal or extraperitoneal.⁽²³⁻²⁵⁾ In case of renal dysfunction, nephrectomy is mandatory.⁽⁴⁾

CONCLUSION

Of the most common causes of referral in the patients with retrocaval ureter is pyelonephritis. In symptomatic cases, surgical intervention should be performed and renal function improves after the operation. Although the known associated anomalies must be considered, they seem not to be very common in retrocaval ureter.

CONFLICT OF INTEREST

None declared.

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EDITORIAL COMMENT

This is an interesting article demonstrating good number of cases with good results. It has recently been shown that during the management of the retrocaval ureter, removal of the retrocaval segment is not necessary.⁽¹⁾

Also today, laparoscopic approach to the retrocaval ureter without removing the retrocaval segment seems preferable which results in less morbidity with less pain, shorter hospitalization, and better cosmetic results.

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REPLY BY AUTHOR

The authors would like to express their gratitude to the editors of the *Urology Journal* for their meticulous appraisal of our paper. This series is a report of our experience within more than 2 decades. Laparoscopic approach is a brand new technique which was not available at our center; however, we have started laparoscopic surgeries and hope to report our experiences in the laparoscopic management of the retrocaval ureter regarding such novel approaches in the future.

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