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Retrocaval Ureter: A Case Series of Three Cases Managed with Uretrouretrostomy

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Abstract

Retrocaval ureter is a very rare congenital malformation. We report a 10 years' experience in the diagnosis and treatment of retrocaval ureter, a case series of 3 cases in two different countries of the Middle East. This is a retrospective study that included 3 cases of retrocaval ureters in Egypt and Saudi Arabia. Standard open ureteroureteric anastomosis was performed through a flank incision for each case. Patients' symptoms were re-evaluated after two to four months. Complete recovery from symptoms occurred, and hydroureter and hydronephrosis regressed in all cases. Early diagnosis and treatment are the keys to prevent hydronephrosis and deterioration of renal functions.

Keywords

Loin Pain, Hydroureter, Hydronephrosis, Retrocaval Ureter, Preureteric Vena Cava Inferior, Uretroureteric Anastomosis

1. Introduction

Since its first description by Hochstetter in 1893 [1], approximately 250 cases of retrocaval ureter have been reported all over the world. The retrocaval ureter is a very rare congenital malformation; the incidence of 1 in 1000 live births has been reported [2], with a prevalence of around 0.06% [3]. It is more common in males than females (ratio = 3:1) and appears more commonly in the right ureter [3]. But it may be seen in the left side on cases with situs inversus or duplication of the Inferior Vena Cava (IVC) [4].

Retrocaval ureter (or circumcaval, postcaval ureter) is all misleading names, as the anomaly affects the IVC and not the ureter. So, "preureteric vena cava inferior" is aetiologically the most correct one.

During the embryonic development of the IVC, the Posterior Cardinal Vein (PCV) undergoes a complete regression, allowing the ureter to have an anterior position to the definitive IVC [5]. If there is abnormal development, the ureter is forced to surround the vein; initially located posteriorly, then anteriorly to it at a lower level [5].

According to Huntington and McClure [6], there is a theoretical probability of fifteen different forms of preureteric IVC, only five variants have been described in humans (Table 1, Figure 1), and the other twelve have been observed in animals.

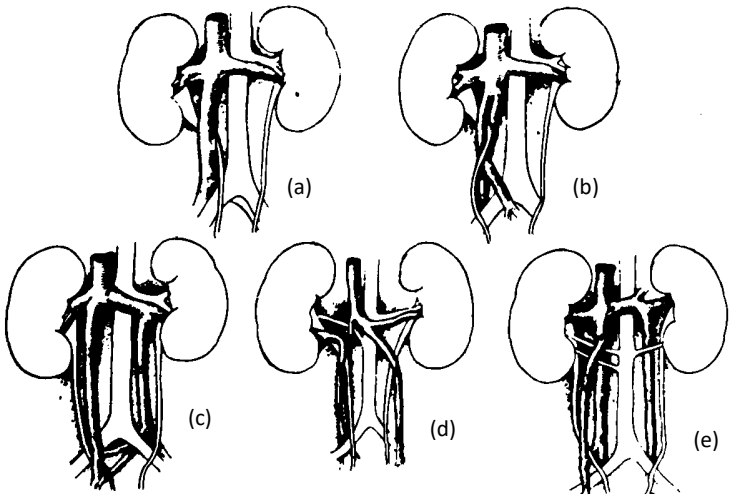

Figure 1. Graphic representation of the five types of retrocaval ureter found in man (after GOYANNA).

Table 1. Variants of preureteric Inferior Vena Cava in human.

		Possible mechanism
Group I	Figure 1(a)	Unilateral right-sided single preureteric vena cava. Persistence of the right postcardinal vein, disappearance or failure of development of the right supracardinal vein.
Group II	Figure 1(b)	Unilateral right-sided double Inferior Vena Cava. Ureter between the two veins. Persistence of the right supracardinal vein and of the right postcardinal vein.
Group III	Figure 1(c)	Bilateral, single Inferior Vena Cava, the right being preureteric and the left postureteric. Persistence of the right postcardinal vein and the left supracardinal vein.
Group IV	Figure 1(d)	Bilateral single preureteric Inferior Vena Cava. Persistence of the right and left postcardinal veins.
Group V	Figure 1(e)	Double right vena cava, ureter between the two veins, single postureteric left vena cava. Persistence of the right supracardinal and postcardinal veins as well as of the left supracardinal vein.

The etiology of retrocaval ureter remains uncertain, but some authors have reported that maternal exposure to substances such as monomethyl ether could be related to the development of this anomaly [7]. Simultaneous congenital anomalies are sometimes associated, such as horseshoe kidney, ureteropelvic junction obstruction, double IVC, congenital lack of the vas deferens, hypospadias, extra vertebra, diverticulum, anterior urethral calculus, renal agenesis, syndactyly in both feet, intestinal malrotation, and Goldenhar syndrome [8] [9].

Patients typically remain asymptomatic until their thirties, when the disease usually manifests itself with attacks of right loin pain, hematuria, lithiasis, and/or recurrent Urinary Tract Infection (UTI) [2]. An earlier diagnosis is not common except if it was associated with another symptomatizing condition, e.g. hydronephrosis [10]. Abdominal ultrasound is the first investigation that raises the suspicion of this pathology, which usually shows right hydronephrosis and right hydroureter at its upper part. Classically, this is followed by Intravenous Pyelogram (IVP), Computed axial Tomography (CT) abdomen with contrast, or Magnetic Resonance Urogram (MRU). All will identify a "fish hook", inverted "J" or "S"-shaped ureter, and a dilation of the collecting system [3] [8]. Retrocaval ureters are classified into two types [11], shown in **Table 2**. Care must be taken to diagnose any possible associated malformations [9].

We report a 10-year experience in the diagnosis and treatment of retrocaval ureter, a case series of three cases in two different countries of the Middle East.

2. Methods

A retrospective study included 3 cases of retrocaval ureters from 2 countries; Egypt and Saudi Arabia, in the period between 2013 and 2023. The study protocol was approved by the Ethics Committee of the Faculty of Medicine, Alexandria University.

Age, gender, side of the affected kidney, admission symptoms, radiological examinations, and grade of hydronephrosis were recorded. All patients underwent surgical treatment. A standard open ureteroureterostomy was performed through a flank incision. Ureter with a dilated proximal segment that crossed the IVC from the posterior aspect and coursed in the medial direction was dissected. Stay sutures were placed proximal and distal to the crossing point, and an oblique cut was done. Then, the ureter was brought in front of vena cava. Cut ends were spatulated, and end-to-end anastomosis was performed over a double J catheter

Table 2. Classification of Bateson and Atkinson for retrocaval ureter.

Type 1	Type 2
Most common (90% of cases)	Less common (10% of cases)
The ureter crosses at the height of the third lumbar vertebra	The ureter crosses at the level of the renal pelvis
Deformity in the form of fish hook or "S"	Sickle-shaped deformity
Marked hydronephrosis	Minimal hydronephrosis

(6 Fr) using 4 - 0 gauge Vicryl sutures, and a retroperitoneal non-suction drain was inserted before wound closure. Transurethral Foley catheter was removed on the third day, drain was removes on the fifth day, and the double J catheter was removed after 6 weeks postoperatively. Patients' symptoms were re-evaluated after two to four months. A follow-up CT scan was obtained in order to check the grade of hydronephrosis, hydroureter, and ureteric relation to the IVC.

2.1. Case 1 (Kafr El-Shikh, Egypt, 2013)

A 22-year-old man presented to the outpatient urology clinic at Kafr El-Shikh General Hospital with complaints of dull intermittent right loin pain interfering with his daily activity for the past year. Clinical examination of the abdomen was free. A laboratory evaluation was done. It included a urinalysis, complete blood picture, urea, creatinine, and electrolytes; all were within normal limits. Ultrasonography (US) of the kidney, ureters, and bladder showed Grade II right hydroureteronephrosis until the upper third of the ureter. CT scan with IV contrast revealed a dilated right renal pelvicalyceal system and upper ureter with the fish-hook sign (**Figure 2**). The diagnosis of retrocaval ureter was confirmed, the patient was operated in **Figure 3** and **Figure 4**, and no intra- or post-operative complications occurred. After 3 months, the patient was totally symptom-free, and the hydronephrosis decreased significantly.

2.2. Case 2 (Kafr El-Shikh, Egypt, 2017)

A 39-year-old married woman complained of right sided lower back pain for four years. She had no history of fever, dysuria, hematuria, or weight loss. She had an Ultrasonography (US) examination that revealed moderate right hydronephrosis and upper part hydroureter. Then, she had a failed ureteroscopy in private sector before she was presented to Kafr El-Shikh General Hospital's outpatient urology clinic. An Intravenous Pyelogram (IVP) and Magnetic Resonance Urorogram (MRU) were performed, which revealed the classic fish-hook sign of the right ureter associated with hydronephrosis Grade II (**Figure 5**). A complete laboratory evaluation, including urinalysis, complete blood picture, kidney function tests, and electrolytes, were within normal limits. She was operated on and no intra- or post-operative complications were reported. After 6 weeks, the symptoms regressed and the hydronephrosis decreased dramatically.

2.3. Case 3 (Hail, Saudi Arabia, 2022)

A 29-year-old man had complained of intermittent right flank pain for 7 years. His US and IVP (**Figure 6**) showed hydronephrosis and hydroureter Grade II. He was treated conservatively for a year by inserting a double "J" catheter (**Figure 7**) followed by intermittent courses of antibiotics and analgesics to relieve pain and prevent recurrent UTI. In 2022, he came for consultation in the outpatient urology clinic at King Khalid hospital. A CT with IV contrast was ordered and showed mild hydronephrosis of the right kidney, right proximal ureteric dilatation with medial deviation, and an abrupt change in the mid ureteric caliber

without detectable stones, likely a retrocaval ureter (Figure 8). All laboratory tests were normal, and the patient was operated on with smooth post-operative period. After 4 months, CT showed right the proximal ureter running in its normal course lateral to the IVC (Figure 9). Patient was totally symptom-free.

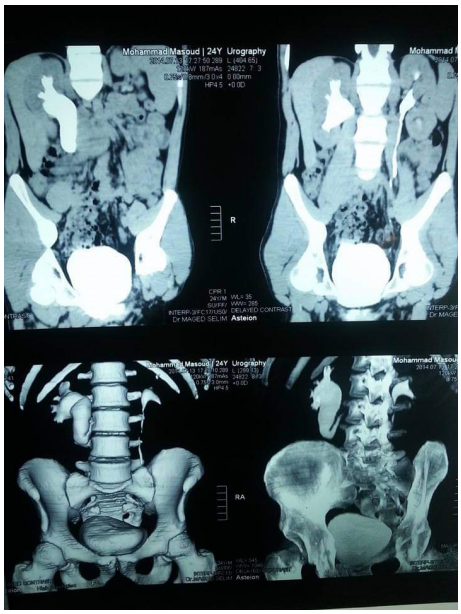

Figure 2. CT scan with IV contrast of Case 1 revealed a dilated right renal pelvicalyceal system and upper ureter with the fish-hook sign.

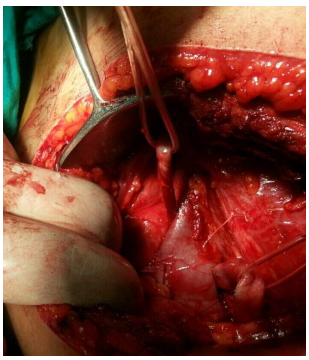

Figure 3. Case 1 before repairing retrocaval ureter, IVC crossing in front.

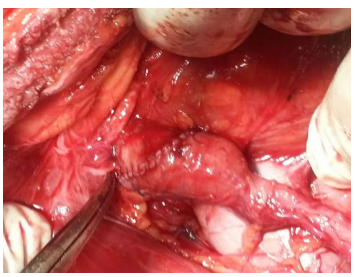

Figure 4. Case 1 after suturing retrocaval ureter in a correct anatomical position anterior to the IVC.

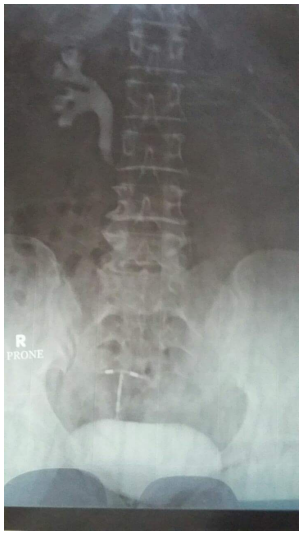

Figure 5. An Intravenous Pyelogram (IVP) of Case 2 revealed the classic fish-hook sign of the right ureter associated with hydronephrosis Grade II (Intra-uterine devise is apparent in the lower part).

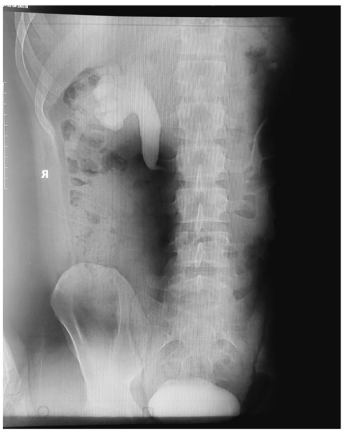

Figure 6. IVP of Case 3 showed right hydronephrosis and hydroureter Grade II.

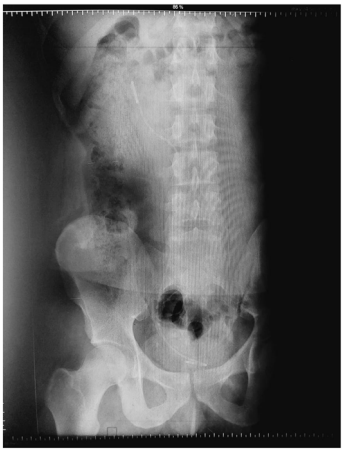

Figure 7. Plain X-ray of Case 3 showing a double "J" catheter in the right ureter.

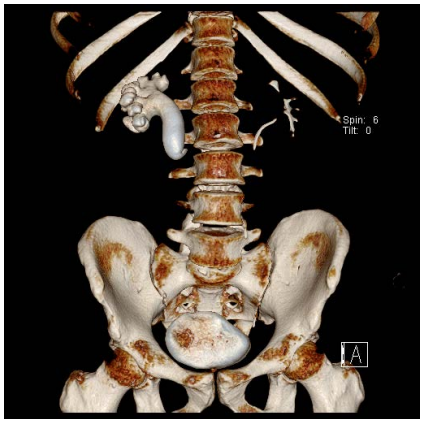

Figure 8. A CT with IV contrast of Case 3 showed mild hydroureter and hydronephrosis of the right kidney.

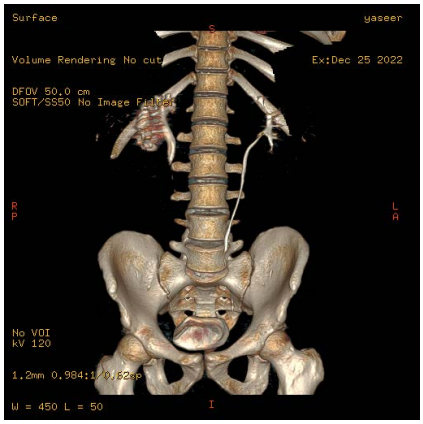

Figure 9. Post-operative CT with IV contrast of Case 3 showed right the proximal ureter running in its normal course lateral to the IVC.

3. Discussion

Although surgical treatment of the retrocaval ureter is not indicated except in symptomatic patients, the natural course of conservative management is not known in the literature. Almost all patients are diagnosed with this congenital anomaly due to their symptoms, which start later in the 3rd or 4th decade of life, and thus require intervention.

Surgical reconstruction can be done through open or laparoscopic approaches. Open surgery is the first line treatment described. However, laparoscopic surgery (transperitoneal or retroperitoneal) is associated with decreased postoperative pain and hospital stay time [12]. Recently, Laparoendoscopic Single-site Surgeries (LESSs) have been used to improve the cosmetic outcome and decrease the number of ports needed [13].

There are multiple techniques for open correction of retrocaval ureter. Some authors reported resection of the dilated renal pelvis, transposition, and re-anastomosis [14]. Others suggested the ureteropelvic anastomosis (Harril method), by which

a section is made at the level of the pelvis just above uretropelvic junction. This technique has the advantage of decreasing postoperative stricture at the anastomotic site due to good vascular supplies of the pelvis and upper ureter [14]. Rarely, nephrectomy for the nonfunctioning kidney because of severe hydronephrosis and infection could be proposed.

In this study, we described the successful repair of three cases of retrocaval ureters by open ureteroureterostomy with anteriorization of ureter. The prognosis and post-operative follow-up were satisfactory. Complete recovery from symptoms occurred, and hydroureter and hydronephrosis regressed in all cases. Early diagnosis and prompt treatment can preserve renal functions and prevent future complications.

4. Conclusion

Although a rare clinical presentation, retrocaval ureter should be suspected whenever its radiological signs are present. Surgical correction can be done by open or laparoscopic approaches, both with satisfactory results. Early diagnosis and treatment are the keys to prevent hydronephrosis and deterioration of renal functions.

Authors' Contribution

Abdullah Elrashidy: main surgeon, processing, revising manuscript, and critical analysis. Emad Ibrahim, Mohab Eleiba, Ayman Elshazly: assistant surgeons in different cases. Rasha Mattar: radiological examination and reporting of Case 3. Suzan Elsharkawy: data collection, writing manuscript, and proofreading.

Patient Consent

Obtained.

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Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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