Al-Mukhtar Journal of Sciences 36 (3): 194-201, 2021

Circumcaval Ureter: An Extremely Rare Vascular Anomaly Causing Hdroureteronephrosis: Case Report

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Received: 29 March 2021/ Accepted: 23 June 2021
Doi.: https://doi.org/10.54172/mjsc.v36i3.328

Abstract: Circumcaval (retrocaval) ureter is a very rare vascular (venous) anomaly caused by an error in the embryonic development of the inferior vena cava in which an embryologically normal ureter becomes entrapped behind the inferior vena cava causing varying degrees of upper urinary tract hydronephrosis. Herein, the current study presents a case report of a 19-year-old Libyan male, single patient, who presented with right flank pain and dysuria of one-year duration. On abdominal examination, he had tenderness on the right renal angle. His laboratory investigations (Hematological and biochemical) were within normal limits. Urine analysis showed *Escherichia coli* infection, which was treated with antibiotics. His workup, including abdominal ultrasonography, showed a moderate degree of hydronephsis. Intravenous urogram showed typical reverse "J" Shaped (fish-hook) deformity and dilatation of the proximal ureter and hydronephrosis on the right side. He underwent the right ureteroureterostomy through a conventional midline approach. He passed an uneventful post-operative course and had a good outcome.

Keywords: Congenital Anomalies; Circumcaval Ureter; Hydroureteronephrosis; Uretero-Ureteral Anastomosis.

INTRODUCTION

Circumcaval (retrocaval) ureter is a congenital anomaly in which the right ureter passes behind the IVC, leading to a varying degree of ureteral compression. This rare embryologic developmental anomaly was first reported by Hochstetler in 1893.(Resnick, 1998). Since its first description, approximately 200 cases have been reported all over the world (Uthappa et al., 2006). The etiology of this congenital disorder is assumed to be abnormal embryologic development of the IVC as a result of atrophy failure (persistence) of the right posterior cardinal vein in the lumbar portion. In this condition, the ureter deviates medially behind the IVC, winding about and crossing in front of it from medial to a lateral direction to resume a normal course distally to the bladder (Lin et al., 2003).

Aim of the study. The rarity of this congenital anomaly and its typical radiological finding necessitates its presentation and inclusion in the differential diagnosis of upper urinary tract dilatation even in the younger age group.

CASE SUMMARY

A 19-year-old Libyan male patient presented with right lumbar pain and dysuria of one-year duration. There were no other complaints related to the urinary tract. He had a herniotomy for congenital inguinal hernia at the age of 4 years. His medical history was unremarkable. No abnormality was detected on general physical examination. Abdominal examination revealed the scar of the previous herniotomy that healed

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by primary intention, and there is tenderness in the right flank region. The examination of the external genitalia showed no abnormality. All routine laboratory investigations were within normal range, except urinalysis, which revealed positive nitrite, 8-10 RBCs/HPF, 16 -20 WBCs/HPF, and urine cultures grew *Escherichia coli* sensitive to ciprofloxacin, furadantine, and gentamycin. Abdominal ultrasonography scan showed a moderate degree of right hydro-uretero-nephrosis with preservation of parenchymal thickness (Figure 1)..

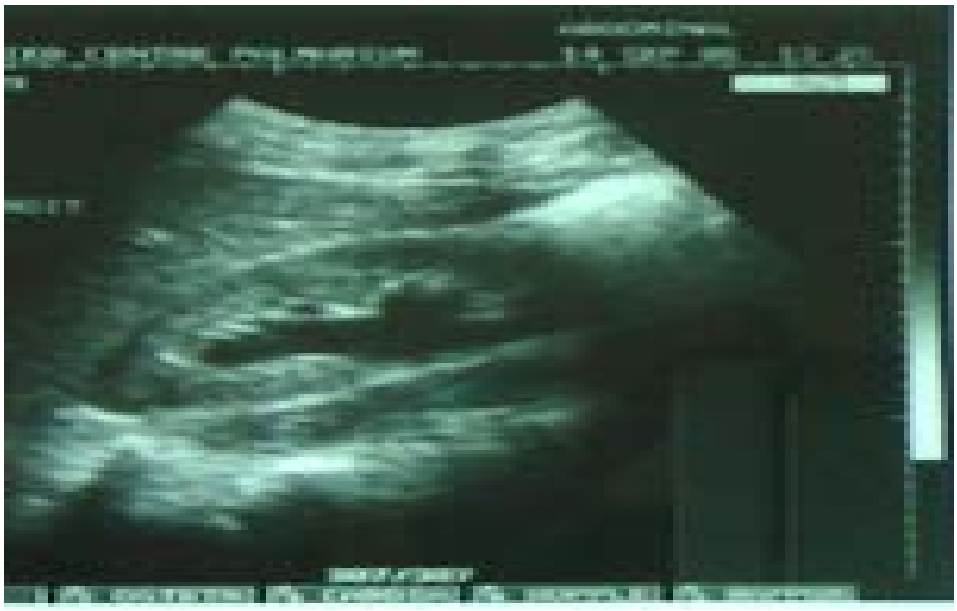

Figure: (1). Transverse renal sonogram showed a moderate degree of hydronephrosis of the right kidney with preservation of parenchymal thickness.

IVU showed delayed function of the right kidney and subsequently right hydronephrosis and hydro-ureter of the proximal ureter up to the level of L3 with typical fish hook /reverse "J" shaped deformity with non-visualization of the rest of the right ureter (Figure 2).

A retrograde pyelogram was attempted but was unsuccessful because of the resistance encountered within 1cm from the entrance of the right ureteric orifice.

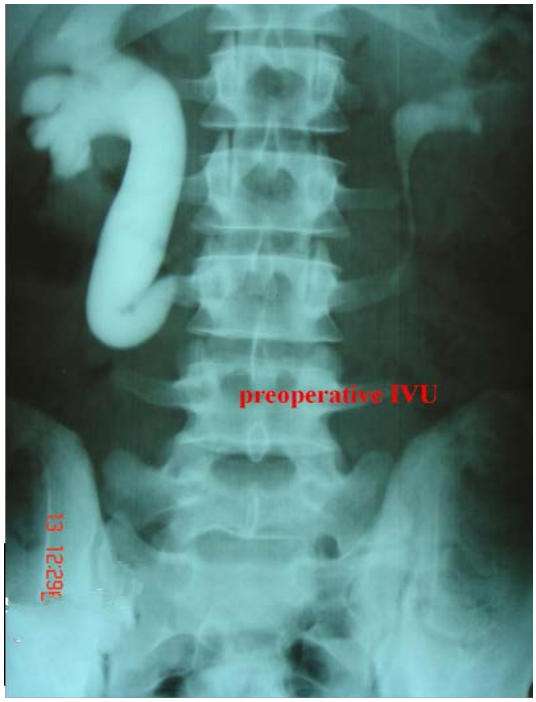

Figure: (2). 30- min. Excretory urography showed right-sided hydronephrosis & dilatation of proximal ureter with characteristic reverse "J" shaped (fish hook deformity) at L3 level.

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PROCEDURE

Through a midline incision, the proximal ureter was found to be moderately dilated, deviating medially dorsal to IVC and winding about and crossing it from medial to a lateral direction to resume a normal course distally to the urinary bladder which confirmed the presumptive diagnosis (Figure 3).

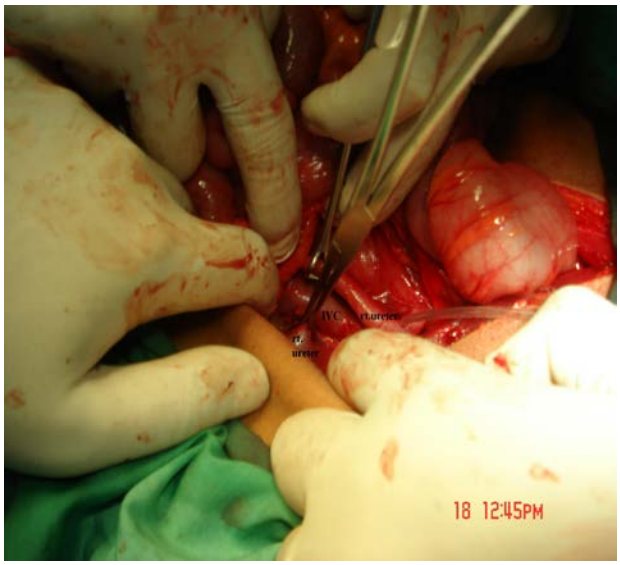

Figure: (3). Intraoperative view showed obstructed spiraling right proximal ureter held with Babcock, IVC and distal ureter with catheter around

After confirmation of the diagnosis; the ureter was carefully dissected anteriorly from the IVC and divided at the medial and lateral borders of the IVC. A dynamic (fibrotic) retrocaval segment was left in situ. The ureter was then relocated (anteriorized) and after confirmation of distal ureteric patency, a uretero-ureteral anastomosis (end-to-end) of the right ureter with a "fish mouth" manner was carried out using 4/0 vicryl over a "DJ" stent crossing the anastomosis.

A tube drain was placed, and the wound was closed in layers. The patient passed an uneventful postoperative course, and tube drain was removed on the fourth post-operative day. The patient left the hospital on the 7th postoperative day to return after 6-8 weeks for the removal of double J "DJ" stent and re-evaluation; where his flank pain disappeared and the DJ removed and IVU was ordered to disclose a regression in the degree of hydronephrosis and hydroureter and patency of the anastomosis site as evidence by visualization of the lower ureter (Figure 4 red arrow).

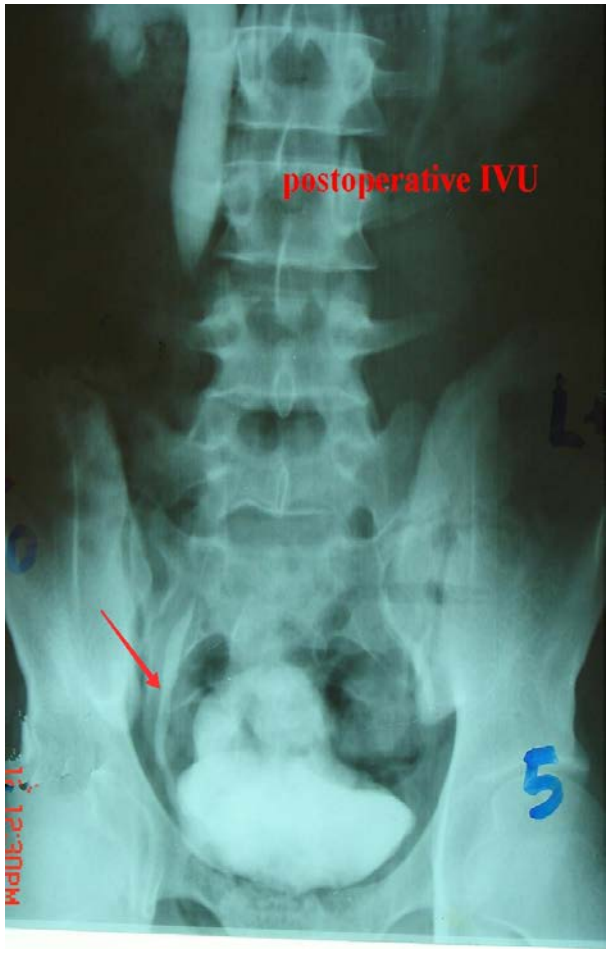

Figure: (4). 30 min .Excretory urography showed regression of hydronephrosis and hydroureter with patency of anastomotic site and visualization of distal ureter (red arrow).

Six months later, he returned for follow up and, he reported to be completely asymptomatic, and IVU showed near-complete regression of hydro-uretero-nephrosis (Figure 5)

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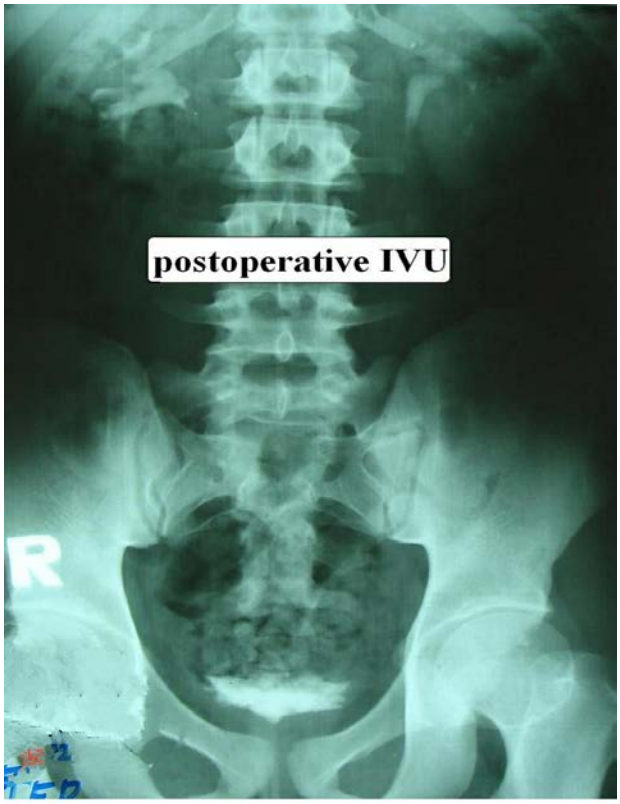

Figure: (5). Post-voidal IVU film showing near normal pelvicalyceal system

DISCUUSION

Circumcaval ureter is a rare congenital anomaly described for the first time by Hochstetter in 1893 (Kyei et al., 2011). The incidence of Circumcaval ureter is about 1 in 1500 cadavers; male to female ratio is 3 or 4:1. One explanation for this male preponderance is the fact that there is a higher rate of male autopsies performed (Richard & Schlussel, 1998).

Circumcaval ureter or retrocaval ureter as known to a urologist are terms that are anatomically descriptive but misleading in regards to development. It is not the result of an abnormality in ureteral development but rather an anomaly in the development of IVC. The term preureteral vena cava may thus emphasize that the circumcaval ureter results from altered vascular, rather than ureteral development (Lesma et al., 2006). The IVC normally develops from the posterior cardinal, sub-cardinal and supracardinal veins, which undergo sequential development, anastomosis, and regression to become the IVC and azygous venous system. Normally, the right sub-cardinal veins form the prerenal IVC, the sub-cardinal and supracardinal anastomosis form the renal segment, and the right supra-cardinal vein forms the postrenal IVC. The left supra-cardinal and lumbar portion of right posterior cardinal vein become atrophic.

If the sub-cardinal vein in the lumbar portion fails to atrophy and becomes primary the right side vein, the ureter is trapped dorsal to it (Akhtar M, 2011; Resnick, 1998). The anomaly predominantly involves the right ureter, as was observed in these reported cases. If it involves the left ureter, then it is usually associated with either partial or complete situs inversus or duplication of the IVC (Kyei et al., 2011; Rubinstein et al., 1999; Watanabe et al., 1991) Although it is a congenital anomaly, it presents in the third and fourth decade of life as typified by the ages of the presented cases. The majority of patients presenting with symptoms, present with flank or abdominal pain that can be intermittent, dull, and aching and is commonly due to ureteric obstruction and associated hydronephrosis. Some patients may present with recurrent urinary tract infections and hematuria. Renal calculi and pyonephrosis may complicate the condition. Some cases are found incidentally during radiographic imaging for other conditions (Kyei et al., 2011). Circumcaval ureter is classified into two types based on its radiographic appearance and the site of narrowing of ureter (Akolekar & Dharap, 2011; Bateson & Atkinson, 1969; Kenawi & Williams, 1976).

Type 1 (low loop);

- Most common (90%),
- Ureter cross behind the IVC at the level of Levertebra and has a fish-hook-shaped deformity (seahorse sign, or shepherd's crook appearance) of the ureter at the point of obstruction.
- Marked hydronephrosis is seen in 50% of the patients.

Type 2 (high loop).

- Less common (10%)

ISSN: online 2617-2186 print 2617-2178

- Cross-over occurs higher at the level of the renal pelvis.
- Lesser degree of hydronephrosis or none at all.
- Renal pelvis and upper ureter lie nearly horizontal before encircling the vena cava in a smooth curve (sickle-shaped curve).

Associated anomalies with retrocaval ureter are reportedly up to 21% (Aliasghar et al., 2006). The various anomalies associated are Horse-shoe kidney, double IVC (Kokubo et al., 1990), and left retrocaval ureter with Branchial arch syndrome (Goldenhar syndrome) (Ishitoya et al., 1997). Concurrent congenital non-vascular abnormalities have also been reported frequently in humans with a retrocaval ureter, including glandular hypospadias, supernumerary lumbar vertebrae, syndactylia, and intestinal malrotation (Rao et al., 2008).Different diagnostic modalities, including IVU, RGP, inferior vena cavography, abdominal USS, abdominal CT, and MRI, have been utilized. With regard to cost-effectiveness and invasiveness, abdominal CT may be the diagnostic procedure of choice (Resnick, 1998).

Abdominal USS can be the only screening radiologic investigation for those with circumcaval ureter which shows ipsilateral moderate hydronephrosis. IVU is the most commonly used radiologic investigation to diagnose circumcaval ureter, but has its limitation in that ureter distal to obstruction is often not visualized (Ratkal et al., 2016).

Pienkny *et al*. suggested that a 3D, volume-rendered CT scan in the excretory phase combined with diuretic renography could be a radiologic investigation of choice in these patients (Pienkny et al., 1999).

Spiral CT, which can simultaneously outline the ureter and IVC, could be another choice to confirm the diagnosis after the ultrasound. The presence of vena cava lateral to the right pedicle of the third lumbar vertebra, which is seen in 94% of patients with circumcaval ureter and only 6% of normal individuals, is said to be the pathognomonic feature of circumcaval ureter on CT (Singh et al., 2001).

In 2002, Uthappa *et al*. reported the potential of MRI to diagnose circumcaval ureter with equal effectiveness of spiral CT but without the radiation risk (Uthappa et al., 2002). The same has been confirmed by (Muthusami & Ramesh, 2013). MRI can also be used in those patients with poor renal function as MR urogram in pregnant women and children.

The main causes of hydronephrosis are lumen stenosis, torsion, and adhesion of the retrocaval segment. This segment is compressed by the psoas muscle, spinal column, and vena cava, which leads to inflammation and fibrosis. Treatment depends primarily on clinical presentation, the severity of the hydronephrosis, and impairment of renal function. Conservative treatment and periodic examinations are necessary for those patients who have mild hydronephrosis without obvious symptoms, infection, worsening renal function, or stone formation (Richard & Schlussel, 1998). In 1935 Kimbrough performed the first successful surgical correction (Huang et al., 2005; Kimbrough, 1935).

Treatment is usually surgical correction; uretero-ureteral re-anastomosis anterior to IVC with transection of the fibrotic retrocaval segment is the favored surgical treatment modality. The retrocaval potion of the ureter can either be excised or left alone. If there is severe hydronephrosis, Anderson Hynes pyeloplasty with precaval transposition of the ureter has been advocated. Occasionally nephrectomy may be required in the presence of thinned out cortex, poor function, or severe infection (Harrill, 1940). The surgical management in the last decade has shifted from open surgical repair to minimally invasive surgeries. Both transperitoneal and retroperitoneal approaches have been used, and both have their advocates (Ratkal et al., 2016). The treatment outcomes

ISSN: online 2617-2186 print 2617-2178

were evaluated by clinical symptoms and imaging studies (USS and IVU) about 6 to 12 months post-operatively. The successful treatment was defined as an improvement of hydronephrosis in imaging studies and clinical symptoms about 6- 12 months post-operatively (Iqbal & Ansari, 2006).

CONCLUSION

Although circumcaval ureter is a rare congenital anomaly and, patients usually present in the third to fourth decade of life, it should be suspected when imaging studies reveal the typical reverse "J"' deformity of the ureter. Recently spiral computed tomography scan (CT) is considered the tool of choice for the diagnosis of IVC abnormalities and circumcaval ureter. Even more recently, the potential use of magnetic resonance imaging (MRI) to diagnose circumcaval ureter was suggested to have equal effectiveness of a spiral CT scan and without risk of radiation. Associated anomalies are not very common in circumcaval ureter but must be considered.

ACKNOWLEDGEMENTS

We would like to express our deepest appreciation to our colleague Dr. Hassan A. Shembesh for his immense technical assistance during the preparation of this manuscript.

ETHICAL APPROVAL

(Administrative approval)The director's approval was taken before the collection of data from the medical records.

Consent for publication: A written informed consent was obtained from the patient for publication of this case report and any accompanying images which are ready to be sent on request by the editor-in-chief.

Conflict of interest: There are no conflicts of interest regarding the publication of this paper**.**

Funding: This work is not funded.

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ISSN: online 2617-2186 print 2617-2178

الحالب المحيط بالوريد الأجوف السفلي، شذوذ وعائي نادر جدا مسبب موه (استسقاء) الكلية، والحالب دراسة حالة

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تاريخ الاستلام: 29 مارس 2021/ تاريخ القبول: 23 يونيو 2021
Doi: https://doi.org/10.54172/mjsc.v36i3.328

المستخلص: يعد الحالب المحيط بالوريد الأجوف السفلي أحد الشذوذ الوعائية (الوريدية) نادر الحدوث، وسببه خطأ في التطور الجنيني (البدائي) للوريد الأجوف السفلي، ينتج عنه أن الحالب الطبيعي يصبح ماراً خلف الوريد الأجوف السفلي مسببا درجات مختلفة من موه (استسقاء) الجهاز البولي العلوي. هذه الدراسة تقدم تقريرا لمريض واحد، ليبي الجنسية، وهو ذكر يبل غ م ن العمر 19 عام والذي كان يشتكي ألما في الخاصرة اليمنى، وحرقة في التبول لمدة عام. فحص البطن اظهر بان لديه الم خفيق في الزاوية الكلوية اليمنى. كانت تحاليله المعملية (الدم والكيمياء الحيوية) ضمن الحدود الطبيعية. أظهر تحليل البو ل ع دوى الإشريكية القولونية والتي تم علاجها بالمضادات الحيوية. تم إجراء الفحوصات الطبية متمثلة في فح ص الـ بطن والحـ وض بالموجات فوق الصوتية، والتي أظهرت وجود موه (استسقاء) بالكلية اليمني من الدرجة المتوسطة، وكذلك أوضـ حت صـ ورة الصبغة الوريدية للجهاز البولي وجود تشوه نموذجي في شكل سنارة صيد الأسماك (الشص)، أو الشـ كل المقلـ وب لحـ رفـJ بالإضافة إلى توسع الجزء القريب للحالب، والموه في الكلية اليمنى. بناء على نتائج الفحوصات تم إجـ راء عمليـ ة ناجحـ ة للمريض، تم فيها نقل، ومفاغره (توصيل) طرفي الحالب أمام الوريد الأجوف السفلي.

الكلمات المفتاحية: شذوذ خلقي، الحالب المحيط بالأجوف السفلي، موه الكلية والحالب، مفاغره (توصيل) طرفي الحالب.

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