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Anatomical-Clinical Aspects of the Retrocaval Ureter: Report of 4 Cases and Review of the Literature

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Abstract: *Introduction:* Retrocaval ureter (RCU) is a birth defect, characterized by a spiral path of the ureter around the inferior vena cava. It is an abnormal in the development of the inferior vena cava, which can lead to ureteral obstruction. The aim of this study was to report 4 cases of RCU with a literature review. *Patients and method:* This was a retrospective, descriptive study in two department of urology including patients followed up and undergone a surgery for a RCU, from January 2016 to December 2021 in two tertiary hospitals of Dakar. We used radiological classification to describe the anatomical and clinical features. *Results:* Four adult patients suffering from RCU were diagnosed and treated over a 6-year period of time. Their mean age was 39 years (36 and 44 years). There were as many men as women. Low back pain was the most common sign. The Uro-CT scan showed a type I of RCU in 3 patients. For all of them, we performed an open surgical procedure to unhook the ureter followed by an uretero-ureteral anastomosis on a JJ stend. The portion of the ureter in retro cave atresic position was resected. The postoperative follow-up was simple. *Conclusion:* Retrocaval ureter is a very rare birth defect for which the right incidence is not known because of the clinical latency. Although classical surgery leads to positive outcomes, the minimally invasive approach is very promising.

Keywords: Ureter, Inferior Vena Cava, Hydronephrosis, Birth Defect, Pyeloplasty

1. Introduction

Retrocaval ureter (RCU) is a birth defect with a spiral path of the ureter around the inferior vena cava. It is an abnormal development of the inferior vena cava, which can lead to ureteral obstruction.

This rare abnormality was first described in 1893 by Hochstetter by autopsy [2] and first clinically diagnosed in 1940 by Harrill [3]. Although it's global prevalence is unknown, the occurrence of this abnormality is rare with an incidence between 0.06% to 0.17% worldwide corresponding to 200 cases reported [4]. Very few cases have been recorded in sub-Saharan Africa [5-7] and no patient suffering from RCU has been described in Senegal to date.

We report 4 patients presenting RCU diagnosed in two tertiary hospital at Dakar with a literature review.

2. Patients and Method

It was a retrospective descriptive bicentric study including patients that has undergone a surgery for RCU from January 2016 to December 2021. The study was conducted in the urology departments of the Principal Hospital of Dakar and the Aristide Le Dantec hospital. Kanawi and Williams's classification was used to describe anatomical and clinical features of RCU [8].

Type I is the most common with location of the retro-cavity segment at the height of L3, producing the classic inverse J image.

Type II, in which ureter crosses the inferior vena cava higher up, at the level of the renal hilum with a horizontal path of the ureter in its initial segment is rare.

2.1. Case 1

This was a 44-year-old female patient, without any particular medical or surgical history, presenting a right lumbar pain that had been evolving intermittently for about 1 year, with occasional episodes of renal colic. This symptomatology was associated with lower urinary tract symptoms. Physical examination was normal. Serum creatinine was normal and the Urine Cyto-Bacteriological Examination (UCBE) was sterile. Urinary tract ultrasound showed a stage II right uretero-hydronephrosis without evidence of obstruction. The Uro-CT scan showed a right ureter crossing the IVC at L4, leading to a stage 2 uretero-pyelocalical dilatation (figure 1).

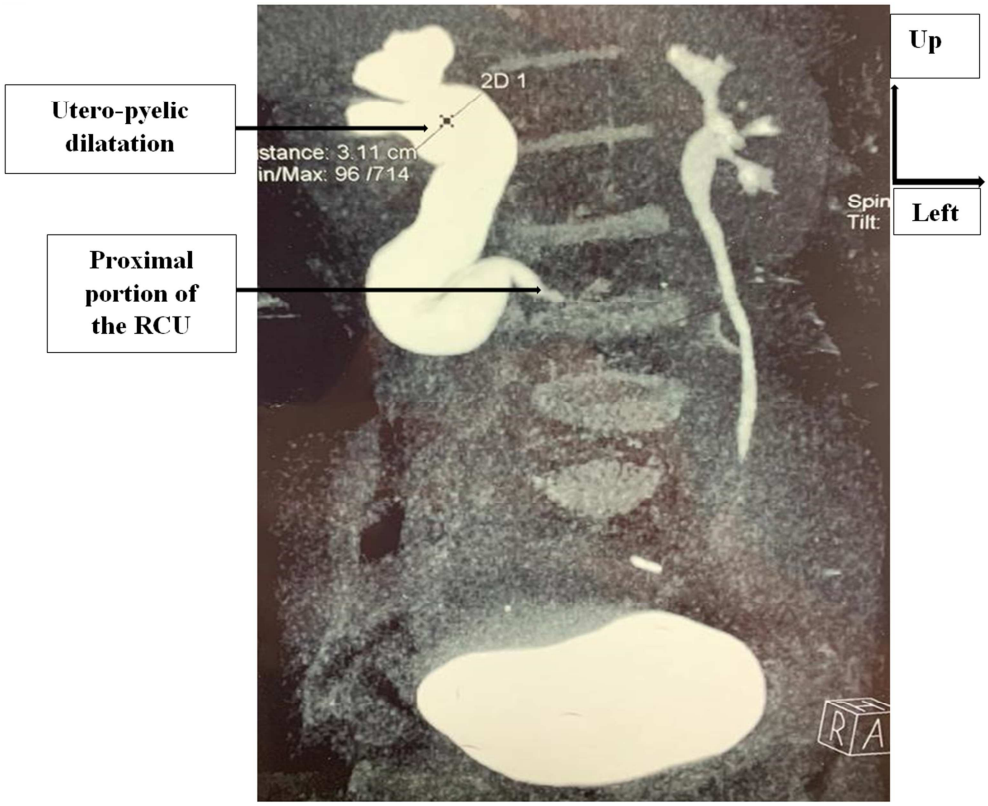

Figure 1. *Scannographic Aspect of typ1 of RCU.*

The diagnosis of a type I RCU complicated by an uretero-hydronephrosis was retained. Surgical treatment by right lumbotomy was indicated with after informed consent of the patient. The procedure consisted of sectioning the retrocaval segment, performing a ureteral uncrossing before proceeding to an end-to-end ureteral anastomosis anterior to the vena cava, with 4/0 Vicryl on a double J stend. The post-operative abdominal X-ray showed a JJ catheter in normal position. The post operative follow-up were simple. The removal of the double J stend was performed one month after surgery. Anatomopathological examination of the surgical specimen showed an organoid structure with a chronic non-specific inflammation.

2.2. Case 2

It was a 36-year-old active military patient referred by his doctor for the management for a right renal cavity dilatation discovered fortuitously during an abdominal ultrasound performed as part of a routine visit. The clinical examination was normal. The UCBE was sterile and the renal function normal. The uro-CT showed a right RCU type I complicated by a homolateral uretero-hydronephrosis. After the patient's agreement and favorable preoperative check-up, ureteral uncrossing and uretero-ureteral anastomosis by right lumbotomy was indicated. In peroperative view, the RCU portion was lumbar, with a small caliber (figure 2).

The procedure consisted of sectioning the retrocaval segment of the ureter, before an end-to-end ureteral anastomosis with 4/0 Vicryl on a double J stend (figure 3).

The postoperative abdominal X-ray showed a correct positioned JJ stend. The post operative follow-up were simple. The removal of the double J stend under local anaesthesia was done 3 months after the surgery.

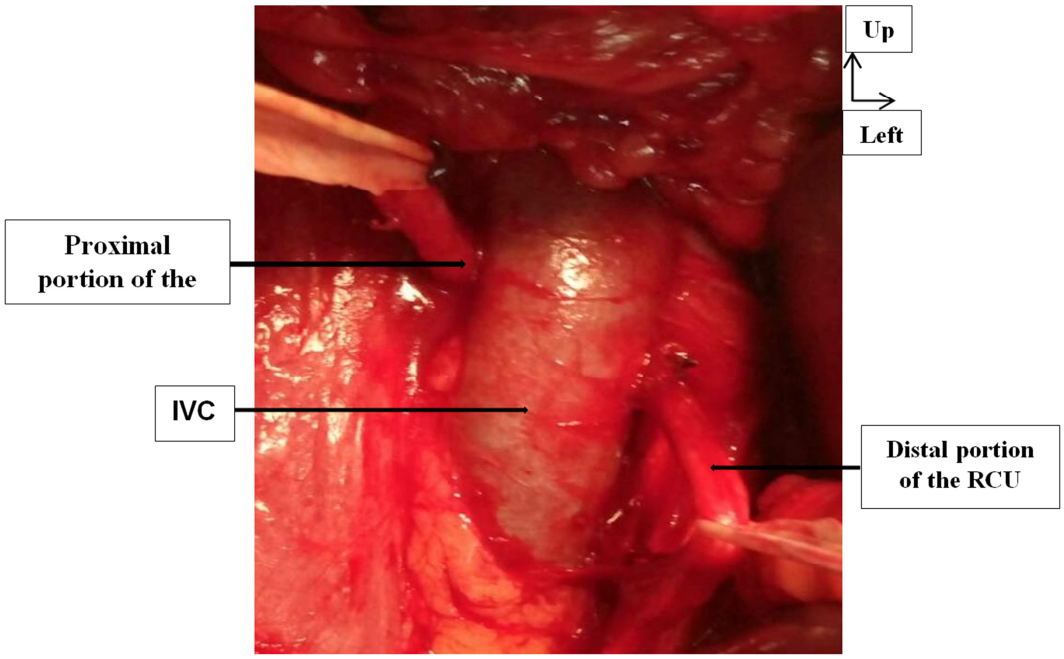

Figure 2. *Intra-operative view of RCU.*

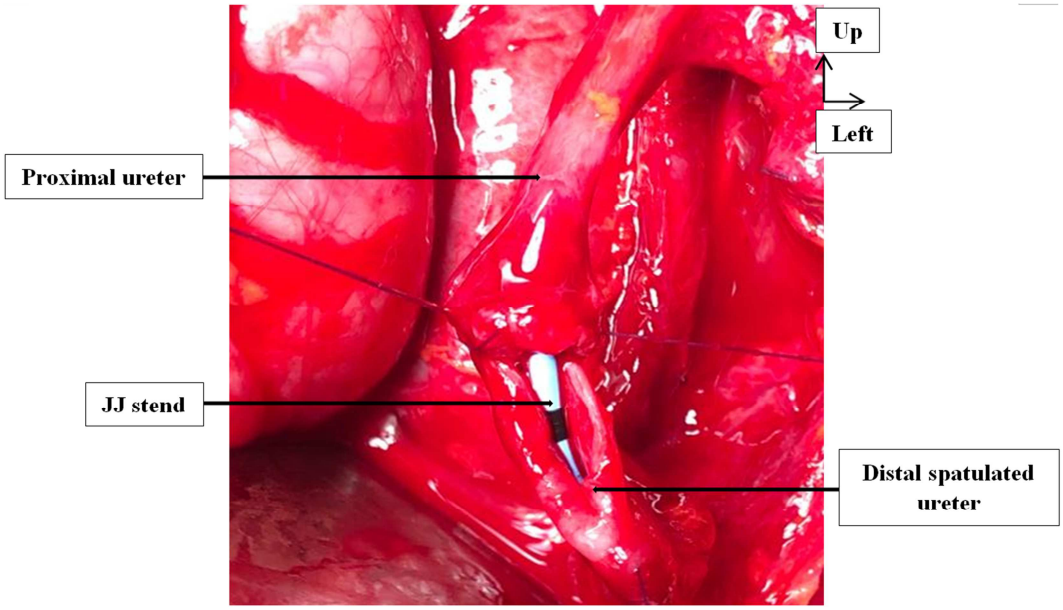

Figure 3. *Uretero-ureteral anastomosis on JJ stend.*

2.3. Case 3

Mr. I. D., 36 years old, was consultated for right lumbar pain and terminal, capricious hematuria evolving for nearly 2 months. The patient had no previous medical history. The physical examination was normal. The UCBE had isolated Escherichia Coli which was sensitive to ciprofloxacin and correctely treated. Serum creatinine was at 16.0 mg/l. Uro-CT showed a pyeloureteral junction syndrome and thinning of the renal parenchyma (figure 4).

Pyeloplasty was indicated with a lumbotomy approach. Exploration showed a RCU with a ureter crossing the IVC just under the renal hilum. The diagnosis of type II RCU was retained in per-surgery The procedure consisted of sectioning the retrocaval segment, performing a ureteral uncrossing before proceeding to an end-to-end ureteral anastomosis anterior to the vena cava, with 4/0 Vicryl on a double J stend (figure 5).

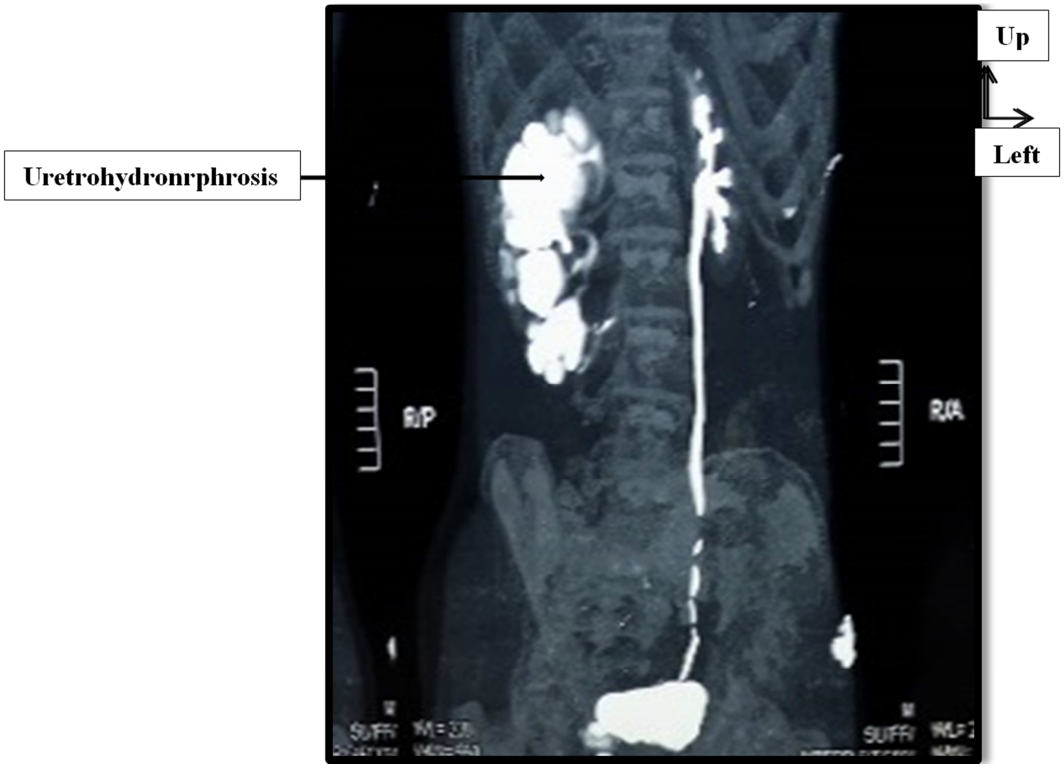

Figure 4. *RCU type II with the appearance of pyeloureteral junction syndrome.*

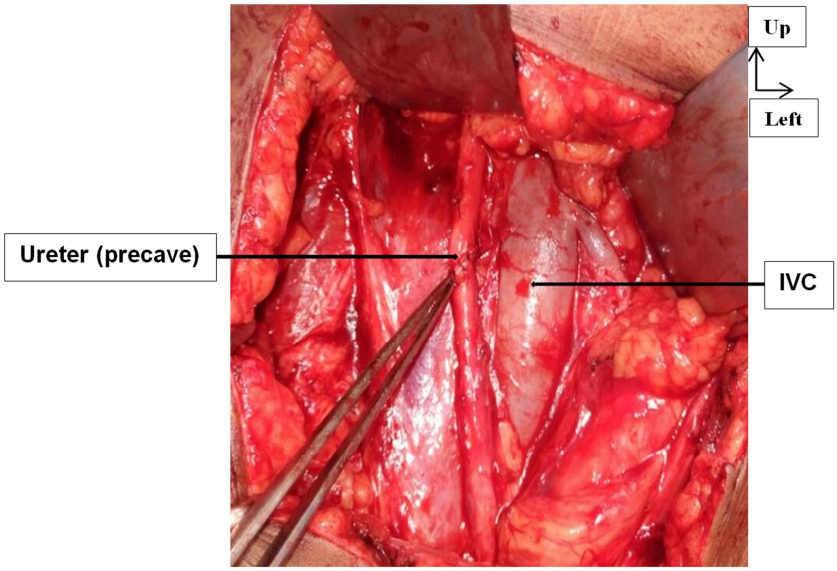

Figure 5. *Final aspect after uncrossing and ureteral anastomosis.*

Post-operative follow-up were simple. After one month of surgery the removal of the double J stend has been performed.

2.4. Case 4

It was a 41 years old female who presented with intermittent right lumbar pain evolving for 3 months. The physical examination showed a pain of the right upper and middle ureteral points. Renal function was normal and there was no urinary tract infection in the CBUE. The Uro-CT showed a URC type I, with a stage III pyelocalic dilatation. A ureteral uncrossing and uretero-ureteral anastomosis by right lumbotomy was indicated under general anesthesia. The procedure consisted of sectioning the retrocaval ureteral segment, before proceeding with and end-to-end uretero-ureteral anastomosis with 4/0 Vicryl around a double J stend. The patient was seen two months later and had no complaints and we proceeded to the removal of the JJ stend.

The clinical and therapeutic characteristics are summarized in Table 1.

Table 1. *Clinical and therapeutic characteristics of patients.*

Observation	1	2	3	4
Age (years old)	44	36	36	41
Gender	Female	Male	Male	Female
revealing symptoms	Right lumbar pain	Incidental	Right lumbar pain + hematuria	Intermittent Right lumbar pain
Anatomical type	Type I	Type I	Type II	Type II
Procedures	Uretero-ureteral anastomosis	Uretero-ureteral anastomosis	Uretero-pyeloplasty	Uretero-ureteral anastomosis
Post-operative follow-up	Simple	Simple	Simple	Simple
removal of double J stent	2 months after surgery	3 Months after surgery	1 months after surgery	2 Month after surgery

3. Discussion

RCU is a rare birth defect in which the ureter passes behind the inferior vena cava (IVC) [9]. This anomaly occurs between the 4th and 8th week of intrauterine development and is due to the abnormal formation of the infra renal IVC from the anteriorly located subcardinal vein instead of the posteriorly located supra cardinal vein [10]. Infra renal IVC is normally from the dorsally located supra cardinal vein, but when it develops from the ventrally located subcardinal vein, the ureter is trapped posteriorly, leading to the pre-ureteral vena cava [10]. This abnormality is rare with a worldwide incidence ranging from 0.06% to 0.17% [4]. Only over 200 cases have been reported in the world since the first one published by Hochstetter in 1893 [2]. This frequency is believed to be increasing due to improved radiological diagnostic tools (intravenous urography (IVU), computed tomography urography (CTU), particularly in asymptomatic cases. In sub-Saharan Africa, clinical cases or short series have been reported [5-7, 11, 12]. There is a high predominance of type 1 that is about 96% [9, 13]. In our short series, three cases were type I and the diagnosis was obvious on urocomputed tomography. However, in our third case the initial diagnosis was SJPU and it was the surgical exploration that led to a correct diagnosis. This was the case in one of the observations reported by Tembely et al [7]. In the series by Liu E et al [14], all nine cases were type I. URC is a health condition of the young adult with a mean age of 40 years [14], the long clinical latency could explain this delay in diagnosis. The average age in our series was 39 years with extremes of 36 and 44 years. Nevertheless, cases were described in children [5, 15]. This condition seems to occur more often in men [2]. Indeed, CRU is three times more frequent in men than in women (sex ratio 3 men/1 woman) [8]. Tengue [12] in Togo reported a sex ratio of 3 men/1 woman. However, there is no explanation for this male predominance [12, 15]. In our series, there were as many men as women. The main symptoms of RCU are right body side pain, recurrent urinary tract infection and hematuria. Pain is the clinical reflection of ureteral compression, which manifests radiologically as ureterohydronephrosis of varying grade. RCU can be complicated by ureteral calculi [16] or pyelonephritis [7]. However, it should be noted that some cases remain asymptomatic and are only discovered incidentally during evaluation of unrelated diseases.

Sometimes, RCU is asymptomatic and is discovered incidentally during a routine check-up [14], as was the case in our second observation. Spiral urotomography is considered the best investigation method, as it allows observation of the ureter and IVC [17]. Magnetic resonance imaging (MRI) may be better than CT because it can delineate the ureter from the IVC without radiation exposure [4]. In our study, uro-CT had led to the diagnosis of CRU in three patients. The principle of retrocave ureter surgery is to restore normal anatomy by uncrossing the ureter and the inferior vena cava. This procedure can be performed by sectioning and anastomosing the ureter or sectioning and anastomosing the inferior vena cava [1] (technique that is currently abandoned). This surgery can be performed open, laparoscopic transperitoneal, laparoscopic retroperitoneal, or robotic [1]. In sub-Saharan Africa, where laparoscopy has been slow to take off in urology, the open approach remains the main route for the treatment of CRU [5-7]. The absence of adequate technical facilities and above all of human resources qualified in coeliosurgery in our centers means that open surgery remains the keystone of the surgical management of this disease. Minimally invasive surgery has the advantage of reducing hospital stay and especially post-surgery morbidity in patients, particularly in this often young and professionally active population [13, 16].

4. Conclusion

CRU is a very rare congenital defect, it's right incidence is not known due to clinical latency. With the rise and accessibility of radiological explorations, isolated cases are increasingly reported in sub-Saharan Africa. Treatment is based on Anderson-Kuss pyeloplasty, the minimally invasive approach of which is the most promising.

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