CASE REPORT

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A rare cause of hydronephrosis: Retrocaval ureter

Lamiaie Bouimetarhan, Othman Ayouche, Ittimade Nassar

ABSTRACT

Introduction: Retrocaval ureter is a rare congenital anomaly resulting from an error in the embryogenic development of the IVC. It can be asymptomatic in the early phase of development and lately manifests as anhydronephrosis with variable degrees of severity, Ultrasonography as well as abdominal CT are required for the early diagnosis and prevention of renal failure. **Case Report:** We report the case of a 29-year-old patient presenting with paroxystic nephritic colic. An abdominal CT with contrast showed an urolithiasis of the proximal lumbar ureter with a type I retrocaval ureter. The patient underwent a surgery with an uneventful postoperative period. **Conclusion:** In conclusion our case suggest that rapid diagnosis followed by minimally invasive surgery can be determinant in the prevention of possible future renal failure. Especially in the pediatric population. Further investigation of the role of sonography for screening should be discussed. **Computed tomography with contrast has overtaken IVU and is considered the tool of choice for analysis and preoperative programming of the retrocaval ureter.**

Keywords: Computed tomography, Hydronephrosis, Lithiasis, Retrocaval ureter, Ultrasound

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INTRODUCTION

Retrocaval ureter is a rare congenital anomaly resulting from an error in the embryogenic development of the IVC. The inferior vena cava results from the evolution of three symmetrical systems with close anatomical relationships with the ureter during embryogenesis the posterior, subcardinal and supracardinal systems [1].

Retrocaval ureter can be considered secondary to the regression of supra cardinal right the vein in favor of the right sub cardinal vein [1]. This anomaly can be either unilateral on the right or bilateral but never on the left alone. Which can cause an obstructive syndrome [2]. Its incidence is approximately 1 in 1500 people with a 3:1 male preponderance [3, 4]. There are two types of retrocaval ureter with a surgical impact. type 1 in which the obstructive syndrome is related to an intrinsic abnormality of the retrocaval segment of the ureter and type 2 in which the obstruction is due to the extrinsic compression of a normal ureter [5, 6].

This classification is based on the radiological aspect and the point of narrowing of the ureter [1].

It can be asymptomatic in the early phase of development and lately manifests as a hydronephrosis with variable degrees of severity, Ultrasonography as well as abdominal CT are required for the early diagnosis and prevention of renal failure.

Lamiaie Bouimetarhan¹, Othman Ayouche¹, Ittimade Nassar¹

Affiliation: ¹Central Radiology Department, Ibn Sina Hospital, Mohamed V University, Rabat, Morocco.

Corresponding Author: Lamiae Bouimetarhan, Central Radiology Department, Ibn Sina Hospital, Mohamed V University, Rabat, Morocco; Email: lamiae.bmtrn@hotmail.com

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CASE REPORT

A 29-year-old patient with no history of chronic disease presented with paroxysmic nephritic colic with total macroscopic hematuria. There was no fever or other associated urinary signs.

The clinical examination found a normal patient, eupneic, pyretic with a low back pain. An ultrasonography of the kidney was performed and objectified a right kidney hydronephrosis without obvious obstacle.

An abdominal CT with contrast showed a obstructed segment of a right lumbar ureter compressed by an aberrant IVC at the level of the third lumbar vertebra (Figure 1), contiguous to an urolithiasis (5.7 mm) (Figure 2) with an upstream dilation of the proximal ureter (26.6 mm) and a « fish hook » appearance till the level of obstruction (Figure 3). There was also anhomolateralhydronephrosis (pelvic AP diameter of 45 x 30mm) and a delayed excretion of contrast. The diagnosis of a type Iretrocaval ureter was made.

The left kidney and bladder were without abnormality. The rest of the abdomen was without any noticeable abnormality. The patient underwent a surgery, which consisted of a section of the retro-caval portion and uretero-ureteral anastomosis and placement of a double J drainage. The patient had an uneventful postoperative period.

Figure 3: Coronal view Abdominal CT: C+ venous phase: Lumbar retrocaval ureter with an important hydronephrosis (Fish hook sign).

DISCUSSION

The persistence of the posterior sub cardinal vein causes the proximal IVC to remain ventral to the ureter; this condition is known as a circumcaval or retrocaval ureter first described by Hochsetter in 1893. It is an abnormal rapport between the ureter and the vena cava where the ureter describes an S trajectory behind the vena cava and circumscribes its medial edge pursuing its normal course [1, 7, 8].

The incidence is approximately 1 in 1500 people with a 3:1 male preponderance [3, 4]. There are two types of retrocaval ureter with a surgical impact. This classification is based on the radiological aspect and the point of narrowing of the ureter [8].

Type 1 is the more prevalent variety (90%) has a «S-shaped» or «fish hook» appearance till the level of obstruction which is usually a little farther from the lateral margin of the IVC at the level of third lumbar vertebra in this variety [8].

The type 2 represent only 10% of cases and has a smooth sickle shaped curve of the right ureter with the level of obstruction at the lateral margin of the third lumbar vertebra [8].

The pathology would rarely manifest itself in infancy. The average age of discovery is 40 years. The anomaly can be asymptomatic or accidentally discovered. Depending on the vessel amount of compression, a varying degree of hydronephrosis is noted in the collecting system. The majority of retrocaval ureters, 80%, manifest by various symptoms that consists of: flank pain, renal colic, recurrent infection of the upper apparatus urinary.

A microscopic or macroscopic hematuria can be seen in case of stasis lithiasis enclosed in the retrocaval part of the ureter. There may be other manifestations such as micturition disorders related to a cystitis, stenosis of the urethra, enuresis or a varicocele [9].

The first para-clinical examination prescribed in front of a painful symptomatology of the right flank, should be ultrasound because of its high sensitivity, specificity and the time taken for the study [9]. Although, it can only be

Figure 1: Axial view contrast enhanced abdominal CT: C+ venous phase: Retrocaval ureter with an important hydronephrosis.

Figure 2: Axial view non-enhanced abdominal CT: lithiasis of the lumbar ureter.

a screening exam which demonstrate hydronephrosis, its severity degree, the viability of renal parenchyma as well in the prevention of complications. IVU was the most common radiologic investigation method in retrocaval ureter diagnosis [10]. It manifested its limitation due to the examination time, which can strain aged patients, the necessity of accelerating the workflow in radiology centers but also due the difficulty to study the distal ureter to the level of stenosis [8].

Computed tomography with contrast has overtaken IVU as the investigative method of choice for the evaluation of hydronephrosis etiologies. It has a high sensitivity and specificity and can give the surgeon the topography of the abnormality by showing [8]:

- The obstructed segment of the ureter.
- The exact point of its medial swing.
- The level of compression of the right ureter by the aberrant vessel.
- The dilatation severity of the urinary tract proximal to the obstruction.
- Excretory function of the kidneys.

It can be also helpful in excluding differential diagnosis such as retroperitoneal fibrosis and retroperitoneal masses displacing the ureters from its normal course [11].

Even more recently, magnetic resonance urography can be interesting alternative without the radiation risk in case of pregnancy or counter indication of tomography.

Any radiologic method will show a right hydronephrosis above the circumscribed IVC segment.

The treatment depends on the clinical manifestation, degree of hydronephrosis and renal function. Asymptomatic patients are followed with periodic examinations

Two surgical types have been identified which have a surgical impact with the type requiring a surgical resection with the case for our patient, and the type 2 in which a plasty without resection can be attempted [12].

Surgical correction is usually indicated in case of obstructive symptoms or a worsening of kidney function, to preserve renal functioning and to provide long-term symptomatic relief [12].

Open surgery is the standard treatment for type 1 retrocaval ureter. It consists of a division of the dilated renal pelvis with transposition, resection of the retrocaval segment of the ureter and termino-terminal reanastomosis, with good long term results [12].

This technique was initially described and popularized by Harril as a form of treatment in patients with symptomatic hydronephrosis. Both flank and transabdominal approaches have been used for this procedure [12].

Transperitoneal and retroperitoneal laparoscopic repair of the retrocaval ureter are other repair approaches that offers advantages of a shorter hospital stay and early recovery [11, 13].

In children, abstention and a follow-up sonography can be attempted with a possibility of a nephrectomy

in case of irreversible renal damage, because of severe hydronephrosis and infection [2, 12].

In our case, the patient underwent open surgery with section of the retro-caval portion and uretero-ureteral anastomosis and placement of a double J drainage. The postoperative course was without complication [2, 12].

This technique can have the inconvenient of injuring the mural innervation of the ureter that result in ureteral hypotonia as well as urinary stasis in the long run [2, 12].

CONCLUSION

In conclusion, our case suggest that rapid diagnosis followed by minimally invasive surgery can be determinant in the prevention of possible future renal failure. Especially in the pediatric population. Further investigation of the role of sonography for screening should be discussed. Computed tomography with contrast has overtaken IVU and is considered the tool of choice for analysis and preoperative programming of the retrocaval ureter.

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Author Contributions

Bouimetran Lamiae – Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Ayouche Othman – Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Ittimad Nacer – Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor of Submission

The corresponding author is the guarantor of submission.

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Consent Statement

Written informed consent was obtained from the patient for publication of this case report.

Conflict of Interest

Authors declare no conflict of interest.

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