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(CASE REPORT)

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Bilateral complete ureteropielocalicial duplicity associated with ectopic ureteral vaginal insertion in childhood: Case report

Ítalo Grizolia Junqueira de Sousa [1,*], Gustavo Notari de Moraes [2], Gabriela Carvalho Maciel [3] and Luiz Carlos Maciel [4]

[1] *Medical student at the University of Taubaté, City of Taubaté, State of São Paulo, Brazil.*
[2] *Assistant Professor I of the Discipline of Urology at the University of Taubaté, City of Taubaté, State of São Paulo, Brazil.*
[3] *Medical student at the University of Taubaté, Caraguatatuba Campus, City of Caraguatatuba, State of São Paulo, Brazil.*
[4] *Assistant Professor III of the Discipline of Urology at the University of Taubaté, City of Taubaté, State of São Paulo, Brazil.*

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Abstract

Ureteral duplication associated with ectopic ureteral insertion is a rare congenital anomaly clinically more common in females. Early urinary incontinence is the main symptom of ureteral ectopy in the first childhood of these patients. Contrast computed tomography and magnetic resonance imaging are the most effective imaging methods to rule out or confirm the diagnosis of ectopy associated with ureteral duplication. In cases where significant surgical functions are performed, the reconstructive surgical approach with reimplantation of the ureteral unit is the technique of choice.

We present the case of a 6-year-old female patient who was diagnosed with bilateral complete ureteropielocalicial duplicity confirmed by intravenous pyelography. Confirmation of ureteral ectopy only occurred intraoperatively. The surgical correction consisted in reimplantation of the ectopic ureteral unit in the patient's bladder, since preserved renal functions evidenced in renal scintigraphy with DMSA. After performed the procedure, the patient remained asymptomatic and with normal urinary patterns.

For the accomplishment of the present study, we used as data source the patient's medical records, useful for the realization of the clinical history from the diagnosis to the definitive surgical treatment that concluded the case.

Ureteral duplication associated with ectopic ureteral insertion should be perfectly understood and had with a possible diagnosis for cases of urinary incontinence and recurrent urinary infections. Investigation through detailed physical examination and the use of appropriate imaging tests may help in the early detection of this rare anomaly of the genitourinary tract.

Keywords: Ureteropielocalicial duplicity; Ureteral ectopy; Urinary incontinence; Congenital anomaly

1. Introduction

Ectopic ureter is single or duplicate, which has no insertion around the bladder trigone [1]. The occurrence of ureteral duplication associated with anomalous implantation of ureter is a rare clinical entity, with a prevalence among newborns of 1/2,000 and 1/2,000 – 4,000 among the general population [2]. Clinically, ureteral ectopy is 2 – 12 times more common in females [3,4].

* Corresponding author: Ítalo Grizolia Junqueira de Sousa

It is estimated that 7.5% - 17% of ureteral ectopy cases are bilateral [3]. Among women, 80% of ectopic ureters are related to drainage of duplicate collector systems [4]. In the male population, unique collection systems are more prevalent [4]. Depending on the site of ectopic ureteral insertion, the clinical presentation of ureteral ectopy symptoms may vary between men and women [2]. Usually, males do not present urinary incontinence. In these cases, the ureteral anomalous implantation site is above the external urinary sphincter [2]. The diagnosis of ectopic ureter among female patients is made early in childhood, presenting as a constant drip of urine through the external genitalia [5,1]. It is important to highlight that, in most cases, the voiding patterns of these women are normal, with preserved voluntary micturition [2].

Imaging tests are mandatory to confirm the diagnosis of duplicity associated with ureteral ectopy [2]. Although ultrasonography remains the test used for an initial evaluation [6], contrast computed tomography and magnetic resonance imaging are the most effective methods to rule out or confirm the presence of ectopic ureter [7]. Despite confirmation by tomography, differential diagnoses such as overactive bladder should be eliminated [2]. The treatment of symptomatic patients with ectopic ureter insertion is surgical [8]. The analysis of renal function status is the main parameter for the study of the best therapeutic approach [4].

Because it treats a rare clinical picture within medical practice and, sometimes, an underestimated diagnosis, which has direct impacts on the patient's quality of life, the case addressed in this study aims to highlight the relevance of ureteral duplication associated with ectopic ureter implantation, and thus assist in its early identification and treatment.

2. Material and methods

For the present case report, a detailed analysis and review of the patient's entire medical records were performed. Thus, we had access to all medical evolution, laboratory tests, urodynamic and imaging tests performed by the patient between years 2014 and 2020, useful for the establishment of the diagnosis of ureteral duplicity associated with ectopic ureteral insertion, as well as its subsequent definitive surgical treatment.

Due to the patient's age, the study included the application of the Free and Informed Consent Form, in addition to the Free and Informed Consent Term, which were read and signed by the legal guardians and the patient, respectively. The beginning and follow-up of the research occurred only as a median signature of the terms cited.

To better deepen on the theme of this original article, we used as bibliography previous studies on ureteral duplicity with ectopic ureteral insertion. The analyzed studies were published in the lastten (10) years, in the English and Portuguese, published in the PubMed database.

3. Case report

We present the case of a 6-year-old female patient who, 3 months after birth, had recurrent febrile urinary infections.

The beginning case investigation, at 7 months of age, led to the diagnosis of grade IV vesicoureteral reflux for both units on the left side expressed in voiding urethrocystography (Figure 1), in addition to bilateral complete pielic duplicity on intravenous pyelography examination (Figure 2), and prophylactic antibiotic therapy was instituted. After the study, the patient was reevaluated, during which time the previous symptoms were maintained, which began to be associated with urinary incontinence - evidenced by continuous loss of urine, which kept the child's underwear constantly wet, requiring changing it several times a day.

The second diagnosis established was that of an overactive bladder, although urodynamic study did not show such a condition. Anticholinergic therapy with oxybutynin hydrochloride was introduced at this stage. Treatment was maintained until the patient was 5 years of age, without significant improvement of urinary symptoms in the period.

At 6 years of age, the child was reevaluated by Urologists from a new Hospital Group. At this stage, the detailed physical examination of patient's genitourinary system was observed for the first time, continuous loss of urine was observed from the child's vagina. A new retrograde voiding urethrocystography evidenced total remission of reflux. The investigation followed with computed tomography and magnetic resonance of abdomen and pelvis, which did not evidence new information about the case.

Based on detailed physical examination and clinical history, the patient was referred for surgical correction. Anomalous implantation of the right ureter in the vaginal region was confirmed intraoperatively. A cystoscopy was performed,

which identified two left ureteral meatus and a right ureteral meatus which was catheterized. A right hemi-pfannenstiel incision was carried out. Two right ureters were identified, one previously catheterized and the other not. The non-catheterized ureter was opened, and a guide wire was inserted inside, which progressed distally to the peri-urethral region, confirming ureteral ectopy.

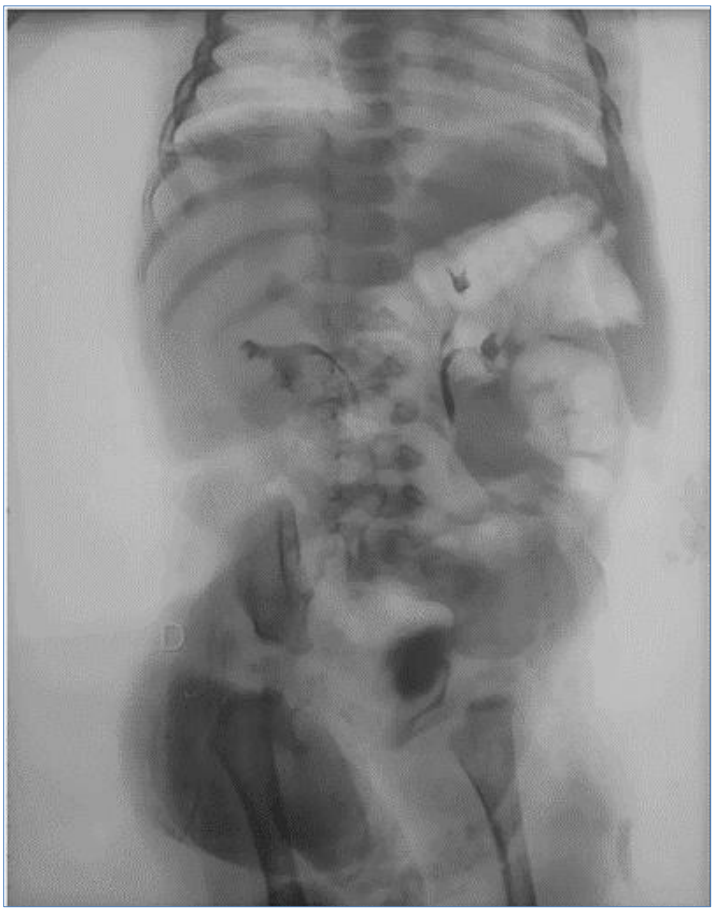

Figure 1 Voiding cystourethrography. Vesicoureteral reflux is observed for both units on the left side

For the present case, a reconstructive surgery was chosen with block reimplantation of ectopic right ureter upper unit. The technique was justified by preserved renal functions evidenced in renal scintigraphy with DMSA.

Since surgical correction in September 2020, the child has remained without urinary and dry infections.

4. Discussion

Ureteral duplication constitutes a urological congenital anomaly; in the population, its estimated incidence is 0.7% [6]. Presented as a rare clinical entity, the association of ureteral duplicity with its ectopic implantation has an incidence among newborns of 1/2,000 and 1/2,000 – 4,000 among the general population [2].

Embryologically, the duplicate ureter arises after the altered division of the ureteral bud, and the upper bud is associated with the upper renal pole and the lower bud with the lower renal pole [8]. In general, this duplicity obeys the Law of Weigert – Meyer [8]. Duplication may be partial or complete, in which two pelvic systems and two ureters occur. Partially in complete duplication corresponds to 70% of cases of duplicity. In this, both ureters join before insertion in the bladder trigone [6].

The ureter that drains the upper pole is more likely to come anomalously [8]. As in the present case of our patient, the ureter responsible for draining the upper pole was in the genital region. The intravenous pyelography performed shows right upper renal pole and its ureter not filled by contrast (Figure 2). The site of ectopic insertion of the ureter determines the clinical forms of symptomatic presentation between men and women [2]. In males, the posterior urethra

is the most common site of ectopic ureteral implantation in 47% of cases [4]. In these men, the presence of urinary incontinence is rare [2].

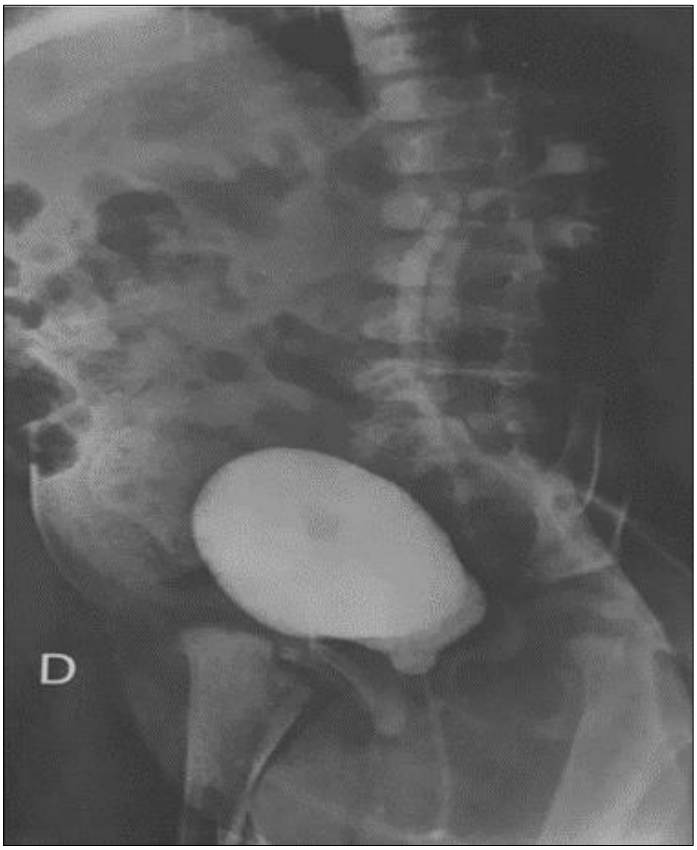

Figure 2 Excretory urography. Complete pielic duplicity is noted

As demonstrated in this study, recurrent urinary tract infections associated with urinary incontinence represent frequent symptoms of ectopia ureteral since early childhood of these women [5,1]. In the present case, recurrent urinary infections, and incessant dripping of urine through the external genitalia were persistent complaints since the patient's three months of age.

In females, the bladder neck and upper urethra followed by the vaginal vestibule, between vagina and urethra, represent the most prevalent sites of ectopic ureteral insertion in 33% of patients; the vagina is a site in 25% of cases, and the cervix or uterus are found as the focus of ureteral implantation in <5% of women with ectopic ureter [2].

Imaging tests are mandatory to confirm the diagnosis of ureteral duplicity associated with ectopic ureteral insertion [2]. Ultrasound remains as the first choice due to low cost and its ability to detect major abnormalities such as ureterocele and hydronephrosis [6]. However, small anomalies may not be evidenced and a complete visualization of the ectopic ureteral insertion site may be challenging [6]. The same limitations apply to intravenous pyelography [2]. In this spectrum, contrast computed tomography and magnetic resonance imaging are today the most effective imaging methods to rule out or confirm the diagnosis of ureteral ectopy due to greater accuracy [7]. Magnetic resonance imaging provides additional information on the anatomy of the urinary tract, facilitating the recognition and characterization of adjacent genitourinary anomalies in children with renal hypofunctioning and ureteral ectopia [9]. For diagnostic investigation of our patient, both computed tomography and resonance of the abdomen and pelvis did not bring relevant information for the conclusion of the case. Ureteral ectopy was initially suspected from the physical examination of the patient's genitourinary system, which showed urine dripping from the child's vagina. Confirmation of the ureteral anomalous implant occurred only intraoperatively.

The symptomatologic similarity with other urological and non-urological diseases is a great delay factor in the diagnosis of ureteral duplicity associated with ectopic ureteral insertion. In the clinical history of the diagnostic investigation of our patient, six years passed until ureteral ectopy was confirmed. During this period, the child remained treated in a

mistaken way for an overactive bladder, a fact that contributed to moderate deterioration of renal function (Figure 3) over the years without the correct diagnosis and treatment.

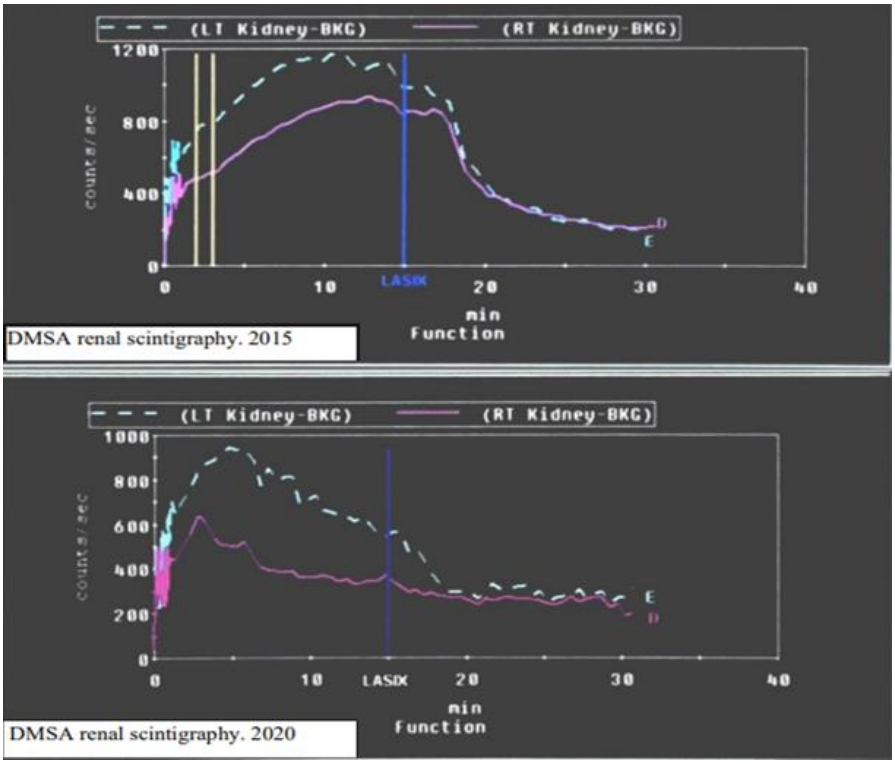

Figure 3 Comparison between renal scintigraphy with DMSA performed between 2015 and 2020. A slight degree of deterioration of renal function was observed during the period in which the patient remained without the diagnosis of ureteral ectopy

Possible diagnoses to be discarded include urinary tract infections, chronic kidney diseases, diabetes, overactive bladder and, in cases of previous pelvic surgeries, vesicovaginal fistula [2]. Other diseases such as congenital heart diseases and dysplasias may also be observed in patients with ectopic ureter [3]. The literature indicates that more than 50% of children with anorectal malformations have associated genitourinary problems [10]. The severity of urological anomalies found in these patients are proportional to the severity of anorectal malformations [10]. In our patient, no associated abnormalities were diagnosed.

The treatment of symptomatic patients with ectopic ureteral insertion is surgical [8]. For the study of the best therapeutic approach, the analysis of the status of renal function is the main parameter [4].

The surgical intervention seeks to solve urinary incontinence, prevent future complications, preserve renal function, and eliminate the recurrence of urinary tract infections [2]. Partial heminephrectomy is the most used surgical approach for correction of ectopic ureters related to dysplastic or hypofunctional upper renal pole [11]. In cases where significant renal function is detected from renal scintigraphy, the reconstructive surgical approach is the technique of choice. A static renal scintigraphy with DMSA showed preserved renal functions with depressed tubular function in mild degree in the right kidney of our patient.

As in the case presented, the reimplantation of the ectopic ureter in the patient's bladder is also an alternative [11]. When ureteral reimplantation is the chosen method, the study of ureteral vascularization becomes mandatory. When confirming that both ureters share the same blood supply, block reimplantation is performed [11].

Since the definitive surgical treatment with ureteral reimplantation, our patient has remained dry and without urinary infections.

Our case report is part of a spectrum of cases that study ureteral duplication associated with ectopic ureteral implantation and aim to highlight its relevance in medicine. A detailed evaluation through careful physical examination

and complementary imaging tests were fundamental for the early identification of this rare anomaly of the genitourinary tract.

5. Conclusion

Because it is a rare condition within medical practice, which has direct impacts on the patient's quality of life, ureteral duplication associated with ectopic ureteral implantation should always be a diagnosis to be considered within the possible etiologies of urinary incontinence in childhood.

Compliance with ethical standards

Acknowledgments

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Disclosure of conflict of interest

No conflict of interest was declared by the authors.

Statement of ethical approval

As it is a case report, the beginning of the present study was approved by the Research Ethics Committee of the University of Taubaté, under the Certificate of Presentation of Ethical Appreciation number 58235521.0.0000.5501.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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