SINGLE SYSTEM UNILATERAL ECTOPIC URETER ASSOCIATED WITH CONTRA LATERAL RENAL AGENESIA. CASE REPORT

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Abstract

Unilateral single ectopic ureters associated with contra lateral renal agenesia and hypoplastic bladder are rare and difficult to treat. Urinary diversion is usually performed because of small bladder capacity. We report a case treated by staged operation without urinary diversion or bladder augmentation.

Keywords: ectopic ureter, incontinence, bladder hypoplasia.

Introduction

Single ectopic ureters are a rare malformation in children. Single-system ureteral ectopia differs from the more common double-system ectopia by a high incidence of associated malformations. Renal dysplasia is common, but does not bear a consistent relationship to the degree of ureteral ectopia. However, in case of bilateral single ectopic ureters or in unilateral ectopic ureter associated with contra lateral renal agenesia, subsequent malformation of the bladder trigone and bladder neck may result in additional voiding dysfunction. (1, 2)

The treatment of unilateral single system ectopia with a healthy contralateral kidney is simple; and it consists in unilateral uretero vesical reimplantation. Cases with bilateral single-system ectopia or unilateral single-system ectopia with contralateral renal agenesis present difficult management.

Methods

The authors report their experience with a girl, in whom unilateral single ectopic ureter with contra lateral renal agenesia was treated by ureteral reimplantation in early childhood and who did not gain adequate bladder control during following years.

Case report

We present the case of a female newborn how was presented in our department with ecographic diagnose of postnatal grade V left hydronefrosis, left lumbar palpable mass and no visualization of the right kidney. She also had biological signs of renal failure but with no urinary tract infection. The cistography showed a small bladder with no vesicoureteral reflux. Due to progressive deterioration of the renal function she underwent a CT scan that shows the absence of the right kidney, a hypoplasic bladder and a grade V left hydronephrosis. We decided to make an upper tract urinary diversion, but unfortunately because of the malfunction of the percutan pielostomy, we were forced to place a new pielostomy catheter via an open lobotomy, which was this time functional. At one month of age the descendent pielography shows a dilated left ureter and raise the suspicion of a left primary obstructive megaureter (fig 1). The child was dismissed at home with a left pielostomy and antibioprophilaxy.

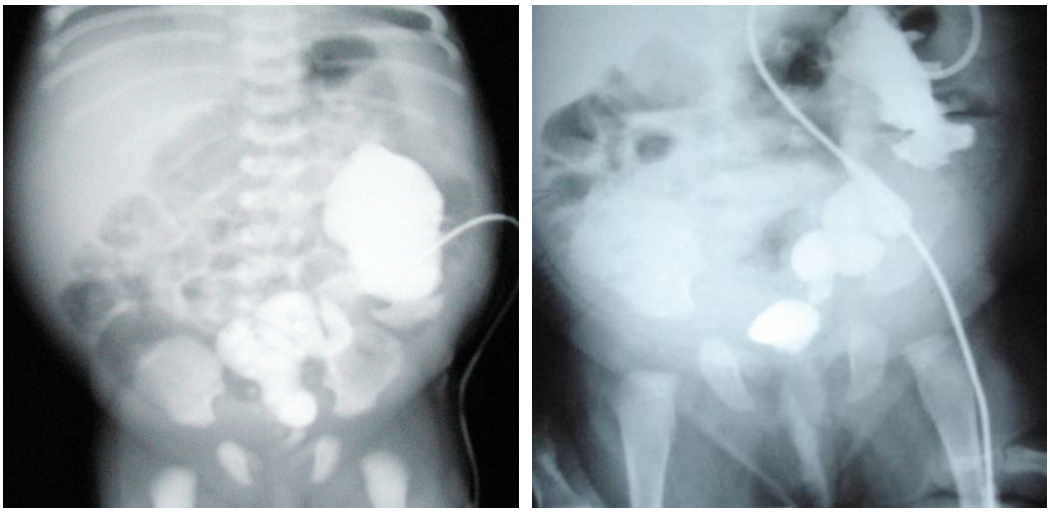

Figure 1 - Postnatal descendent pielography.

She came back 2 month later with normal renal parameters, and an acute urinary tract infection, which required intravenous antibiotheraphy. The cistoscophy revealed a hypoplasic bladder, with no right ureteral orifice and an ectopic left ureteral meatus, which was opened in the bladder neck. A repeated pielography showed no passage of the contrast through the ureteral meatus, so at the age of 4 months the ectopic ureter was reimplanted into the bladder by Politano-Leadbetter technique.

Postoperative follow-up revealed a bad functionality of the uretero-vesical junction with an increase of the hydronephrosis and progressive deterioration of the renal function. A new urinary diversion by pielostomy was necessary one month after reimplantation. The very small bladder show no improvement in volume, with a pour function on repeated attempts of pielostomy removal with short dry period at aproximally 3 years follow-up. (fig 2)

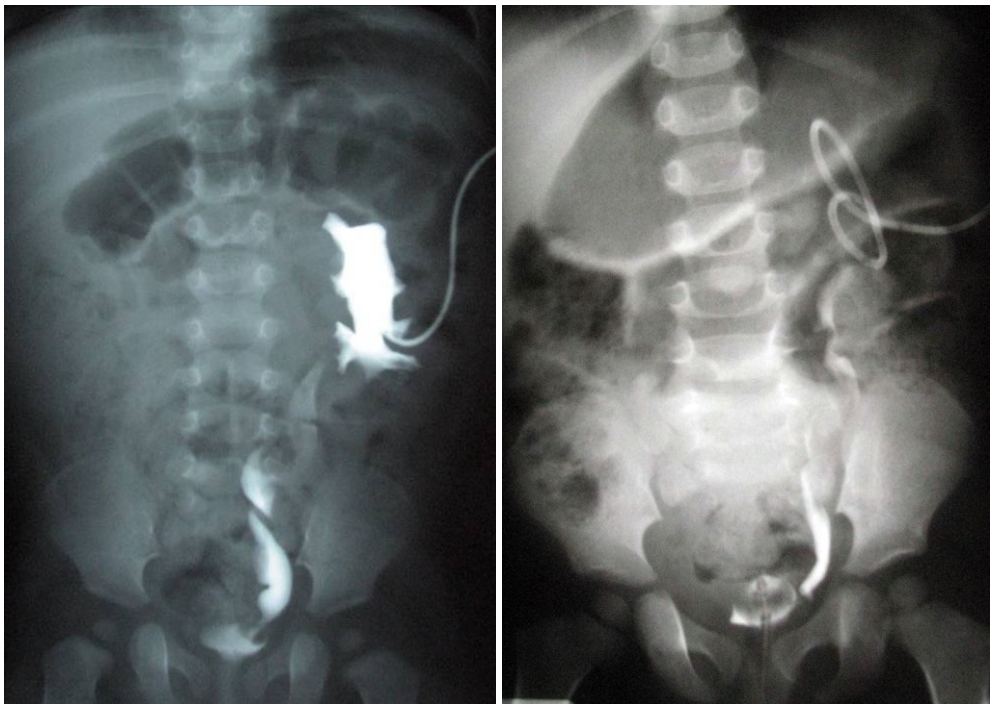

Figure 2. Postoperative pielography.

Discussions

Ureteral development begins in the 4th week of gestation, in close relationship with the renal embryology. At 4 weeks of gestation, a ureteric bud arises as an out pouching from the mesonephric duct and interacts with the metanephric blastema. The segment of the mesonephric duct distal to the ureteric bud becomes a common excretory duct and is eventually absorbed into the developing bladder to become part of the trigone. The point of origin of the ureteric bud is the future ureteric orifice. If the ureteric bud arises more proximally on the mesonephric duct than normal, the future ureteric orifice will be more medial and caudal because it has less time for its normal migration into the bladder, the ureteral orifice can ultimately be incorporated into one of the Mu¨llerian structures instead of into the bladder.(3)

In females, it is possible that the orifice may be distal to the urethral sphincter allowing continuous urinary incontinence, with the ureter inserted into the lower bladder, urethra, vestibule, or vagina. More rarely, it can empty into the uterus or a wolffian duct remnant such as Gartner duct or cyst.

In males, it empties into the lower bladder, posterior urethra, seminal vesicle, vas deferens, or ejaculatory duct. In very rare instances, it can empty into the rectum. The fundamental difference between ureteral ectopia in females and in males is that in females, ectopic ureters can terminate at a level distal to the continence mechanisms of the bladder neck and external sphincter and thus may be associated with incontinence. Approximately one-half of female patients with ectopic ureters present with a classic history of continuous dribbling incontinence despite what appears to be a normal voiding pattern.

An ectopic ureter can drain a single kidney, but about 70% are associated with complete ureteral duplication. In complete ureteral duplication with each segment having its own ureteral orifice in the bladder, the Weigert-Meyer rule applies. This rule states that the ureteral orifice of the upper pole moiety inserts into the bladder medial and inferior to both its normal location and the orifice of the ureter draining the lower renal segment. In these cases, the ureter draining the upper pole moiety frequently ends in an ureterocele, whereas reflux into the lower moiety typically occurs. The renal parenchyma abnormalities associated with duplex

systems are thought to be the result of abnormal origin of the ureteric bud. It is believed that the farther away the ureteral bud arises from its normal location, the more abnormal the resulting renal segment. The embryological defect in the group of children with single-system ectopia is clearly more than an anomalous ureteric bud (4, 5, and 6)

In female single ectopic ureters on unique renal unit cases, the development of trigone and bladder neck musculature does not occur because of the ectopic openings being outside the bladder, which therefore does not have the opportunity to distend with urine. Additionally, a poorly developed bladder neck and an improperly functioning urethral control mechanism prevent bladder growth. The surgical treatment of an ectopic ureter in a female depends on the associated renal function. Single-system ureteral ectopia to the genital system usually has poor function, and a nephroureterectomy is appropriate when the other kidney has a good functionality. With single-system ectopia to the bladder neck or urethra, the function may justify a reimplantation of the ureter into the bladder.

Treatment of unilateral ectopic ureter with contra lateral renal agenesia is challenging, similarly to incontinent epispadias. First of all, preservation of renal function should be the main consideration in these patients. Second, sufficient bladder capacity is necessary to rampant the dilated ureters. The most popular procedure to increase the size of the bladder is enterocystoplasty; however, there is a concern about future malignancy. A last resort is to abandon the native bladder to select urinary diversions such as ureterocutaneostomy, ileal conduit and continent urinary diversion. Incontinence, and bladder functionality remains the most troublesome problem in patients with unilateral single-system ectopic ureters. (7, 8)

In our case de reason to perform at an early age the neoureterovesicostomy was to improve bladder capacity and function. Unfortunately the damages on bladder neck and capacity were not ameliorated, so the patient had to maintain the pielostomy. On the future it will be necessary to perform a bladder augmentation in an attempt to ameliorate the bladder capacity and the continence. Long-term follow up with regard to bladder function is needed.

The diagnose and treatment of unilateral single system ureteral ectopia associated with contra lateral renal agenesia remains a challenge for the pediatric urologist. Despite of a well placed neoureterostomy these children will develop a bad bladder function, with short dry period and incontinence, which will require further surgical interventions.

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