Ectopic Ureteric orifices with Bilateral Duplicated Collecting System

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Abstract

Ectopic ureter occurs three times more commonly in females than in males. More than 80% of female ectopic ureters involve duplicated systems, while the majority of male ectopic ureters are of a single system , this rare case of ectopic ureteric orifice was diagnosed in 10 year old female child who presented with continuous dribbling of urine day and night ,after clinical examination together with Intravenousurography and cystoscopic examination the final diagnosis of bilateral complete duplicated system with unusual ectopic four ureteric orifices was settled .

Key wards: **Ureteric orifice, congenital abnormalities, urinary incontinence**.

فتحات حالب هاجرة فى كليتين متعددة القنوات

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الخلاصة

فتحة الحالب الهاجرة شائع اكثر لدى الاناث مما هو عليه عند الذكور بثلاثة اضعاف وان اكثر من ثمانون بالمئة من فتحات الحالب الهاجرة لدى الاناث تحدث فى الكلى المتعددة القنوات بينما تحدث هذه الحالة عند الذكور فى كلى احادية القناة الجامعة للادرار . حدثت هذه الحالة النادرة جدا عند طفلة ذات العشر سنوات من العمر التى كانت تعانى من تبول لاارادى ليلا ونهرا وعلى شكل قطرات مستمرة منذ الولادة وعند اجراء الكشف السريرى واجراء فحوصات السونار والاشعة الملونة تبين بان سبب حالة المريضة هو تشوه خلقى فى المسالك البولية حيث تبين انها تمتلك كليتين متعددى القنوات الجامعة لادرار اى انها تمتلك اربع حوالب بواقع اثنان لكل كلية ومتكاملة الى ما بعد المثانةوان فتحات هذه الحوالب الاربعة جميعها هاجره خارج المثانة وخارج صمام الاحليل حيث ان مواقعها كانت على جانبى الفتحة التناسلية وبواقع فتحتين من كل جانب وان هذه الحالة قد تحدث فى كلية واحدة وقناة جامعة واحدة وفى جهة واحدة ولكن حدوثها كما فى حالة مريضتنا هذه فى كليتين ذواتى قنوات متعددة متكاملة اي ان لها اربع حوالب وفتحات جميعها هاجره تعتبر من الحالات النادرة جدا وقد تكون الاولى من نوعها.

الكلمات الدالة:
التشوهات الخلقية البولية– فتحات حوالب هاجرة–السلس البولى.

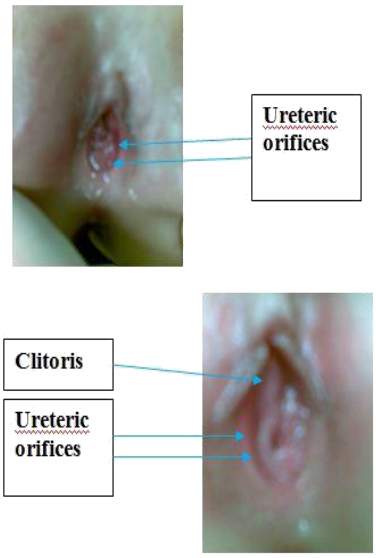
Introduction

Ectopic ureter occurs three times more commonly in females than in males. More than 80% of female ectopic ureters involve duplicated systems, while the majority of male ectopic ureters are of a single system [1, 10]. Complete ureteral duplication occurs when the mesonephric duct gives off a second ureteral bud. The ureteral bud closest to the urogenital sinus (UGS becomes the lower pole ureter and the bud further away becomes the upper pole ureter [2].

As the common excretory duct is absorbed, the lower pole ureteral orifice migrates cephalad and laterally; however, the upper pole ureteral orifice migrates caudally and medially to obey the Weigert-Meyer law [3]. Because the lower pole ureteral bud is absorbed more rapidly, the detrusor submucosal tunnel becomes short , this short submucosal tunnel predisposes the lower pole ureter to reflux, In contrast the upper pole ureteral bud is absorbed slowly resulting in a long submucosal tunnel. In males, the upper pole ureteral orifice may insert into the posterior urethra, prostatic utricle, seminal vesical, ejaculatory duct, or vas deferens, in females the upper pole ureter may insert into the urethra, vestibule, vagina, cervix, uterus, Gartner's duct, or a urethral diverticulum [4]. Approximately half of all affected females experience persistent urinary incontinence because the ectopic ureter drains distal to the external sphincter, males may have symptoms of urinary obstruction or infection. When the ectopic ureter drains into the posterior urethra, males may experience urinary frequency and urgency but not incontinence, If the seminal tract is involved, symptoms may not appear until onset of sexual activity, and include prostatitis, epididymitis or hematospermia [5].

Case Report

10 year old female child presented by her family complaining enuresis , she was treated so (enuresis) for long periods by pediatrician with no response after detail history taking the complain was not only at night but continuous day – night dribbling of urine , physical examination revealed no abnormal findings, abdominal Ultrasonography showed only bilateral minimal dilation of pelvicalyceal systems, IVU revealed bilateral complete duplication , bilateral hydro uretero nephrosis with no contrast seen in the urinary bladder and presence of contrast material on the child’s underwear and here upper thigh was, examination under anaesthesia before erforming Cystoscopy and after gentle separation of the labia majora four orifices were seen 2 on each side of the vaginal introitus all draining urine (Picture.1 A-B) .



Picture.1 A-B Dribbling of urine from orifices on the either sides of the vaginal introitus

Cystoscopy showed small capacity , spastic bladder , with no ureteric orifices identified and the diagnosis of bilateral complete duplication with ectopic of all (four) uretric orifices became clear .

Discussion

There are two distinct forms of ectopic ureter, one draining a duplex kidney and the other connected to a single kidney, the latter being termed single system ectopic ureter [6]. In 80–90% of females an ectopic ureter is associated with duplication of the collecting systems, at least in the western world [7, 13] whereas in Asia the opposite seems to hold true in girls. The ratio of duplex kidneys with ectopic ureter to single dystopic kidneys with ectopic ureter in Caucasians is 4:1 whereas in Asians it is reported to be 1:20 [8], although our case is an Asian one , it is a complete duplex system with ectopic ureters .

[8] reported 44 patients with single system ectopic ureter encountered over a 30-year period, while only two cases of ectopic ureter associated with a duplex system were seen during that period. Overall, female patients are affected twice as commonly as males, although single system ectopic ureter is reported to be more common in males [9]. The diagnosis and localization of ectopic ureter can be a challenging task. In symptomatic patients, as in our case , a high index of clinical awareness is necessary to clinch the diagnosis of ectopic ureter. Imaging modalities including Ultrasonography, Intravenous urography (IVU), and Micturating Cysto Urethrogram (MCU) are useful and should be applied judiciously [10].

Regarding our case we reached the diagnosis from the results of Ultrsounography , Intravenous Urography together with EUA (examination under anesthesia) and Cystoscopy

An ectopic ureteral orifice, i. e., the location of one or more ureteral openings in a position outside the bladder, while infrequent is no longer regarded as a medical curiosity or as a rare finding , Furthermore the poor hygiene and subsequent mental anguish over the "wetness" caused by this anomaly can usually be remedied if the correct diagnosis is made and competent treatment carried out [11]. On the other hand, if the malformation is overlooked, incompletely evaluated, or improperly treated, poor or discouraging results will be obtained. For these reasons, familiarity with the varied clinical pictures observed in patients with this anomaly and with the basic embryological deviations responsible for it is of importance to both the pediatrician and the urologist in order to enable either of them to suspect the malformation promptly, to interpret the various clinical findings correctly, to outline a thorough clinical evaluation, and to select the best treatment option[12] . Several factors were responsible for delayed diagnosis , including parental neglect in bringing the child to early medical attention, physicians not recognizing the significance of urinary dribbling, physical examinations not meticulously performed, omission and/or ineffective use of imaging studies, and incorrect interpretation of radiologic tests.

Conclusion

Although ectopic ureteral orifice is no longer regarded rare and cases of ectopic ureter in unilateral duplex system been reported, but the presence of bilateral complete duplication with presence of four ectopic ureteric orifices (our case) is a rare phenomenon and may be the first reported case ,and every child when present with enuresis should undergoes full and careful evaluation before treatment and continuous urinary incontinence in females with a normal voiding pattern should prompt an evaluation for ureteric ectopia .

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