Ureteral duplication with intrasinusal ureteral junction and epididymal ureteric ectopia

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ABSTRACT

We report a case of 47-year-old adult male who presented with episodic unilateral scrotal swelling associated with dragging pain, with a history of spontaneous resolution and associated non-colicky right flank pain. On evaluation, he was diagnosed to have ureteral duplication with intrasinusal ureteral junction and epididymal ureteral ectopia. Treatment included laparoscopic right lower pole nephrectomy for the thinned out draining segment with ectopic ureterectomy. In literature, very few cases of ureteral duplication in an inverted Y have been reported. In such cases, epididymal ureteral ectopia has been found to be extremely rare. Here, in this case, uncharacteristic symptoms mimicking chronic epididymo-orchitis, diagnostic difficulties and findings are described.

Key words: *Chronic epididymo-orchitis, Epididymal ureteric ectopia, Ureteral inverted Y duplication*

Aberrations in the normal development of Wolffian and Mullerian ducts, ureteral bud, urogenital sinus and bladder lead to ipsilateral genital and urinary tract anomalies including ureteral ectopia. The specific mechanisms responsible for ectopic ureters still remain undefined. Ectopic ureter in a single or duplex system in males always enters the urogenital system above the external sphincter and usually into Wolffian structures, including ejaculatory duct, seminal vesicles or vas deferens while in females it can enter anywhere from the bladder neck to the perineum and into the vagina, uterus, and even rectum [1-3]. Partial ureteral duplication is defined by the presence of a ureter that duplicates at a variable level before reaching the bladder. The incidence of ureteral ectopia was approximately 1 of 2000 people in one autopsy series [2], with slightly higher incidence seen in females [4]. More than 80% of females with ectopic ureters have duplex systems while majorities (75%) in males are singlets [5]. Caudal ureteral bifidity, first described in 1913 by Juvara [6], has been referred to as ureter fissus caudalis or inverted Y duplication. Here, of the two ureters, one opens in the bladder at orthotopic position while the other usually opens into the bladder in an ectopic position or outside the bladder, into ureterocele, seminal vesicle, epididymis, vas deferens, or ejaculatory duct. We hereby report a case with the clinical presentation of chronic epididymo-orchitis which on evaluation was diagnosed to be inverted Y ureteral duplication with intrasinusal ureteral junction and ureteral ectopia to epididymis. It is an extremely rare entity with hardly countable cases reported in the literature.

CASE REPORT

A 47-year-old male patient presented with complaints of episodic recurrent right unilateral scrotal swelling. Scrotal swelling appeared on exertion and was associated with dragging pain and occasional fever. The patient also had right non-colicky, non-radiating flank pain of 4 years' duration. On examination, right inguinal bubonocele was noted with thickening of right epididymis, which was non-tender. The clinical diagnosis of chronic epididymo-orchitis was made. His renal function tests were normal. Ultrasonography of the abdomen and scrotum revealed bilateral renal calculi with right mild hydronephrosis with double, dilated ureters with the ectopic ureter extending into the scrotum possibly draining into the vas deferens. Due to diagnostic dilemma, MR urogram was taken which revealed dilated right pelvicalyceal system and ureter with ectopic ureter seen coursing through the right inguinal canal and ending in the right epididymis (Fig. 1). Isotope scan to quantify the split renal function revealed non-functional right lower polar segment. Hence, the patient was planned for laparoscopic right lower pole nephrectomy with ectopic ureterectomy. Pre-operative retrograde ureteropyelography revealed right pelvicalyceal system communicating with right ectopic ureter which was ending near scrotum (Fig. 2). Intraoperatively ectopic ureter was seen lying in the lateral pelvic wall, well away from the common ureteral sheath. Rim of non-functioning renal parenchyma around the ectopic ureter was removed, and the ureter was mobilized up to the inguinal canal. Nephroureterectomy specimen was retrieved with small scrotal incision and ureter communication to right epididymis was ligated and divided (Fig. 3). Histopathology report turned out to be dysplastic renal segment with ureter having normal transitional urothelium. There were no post-operative complications, and for the past 2 years, the patient is on regular follow-up with no complaints.

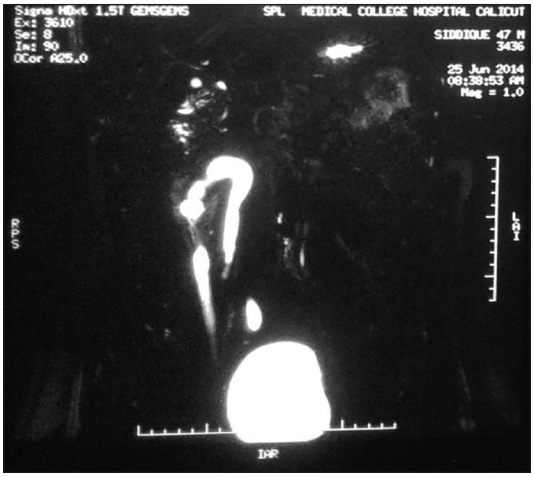

Figure 1: MR urogram showing ureteral duplication with intrasinusal ureteral junction and epididymal ureteral ectopia.

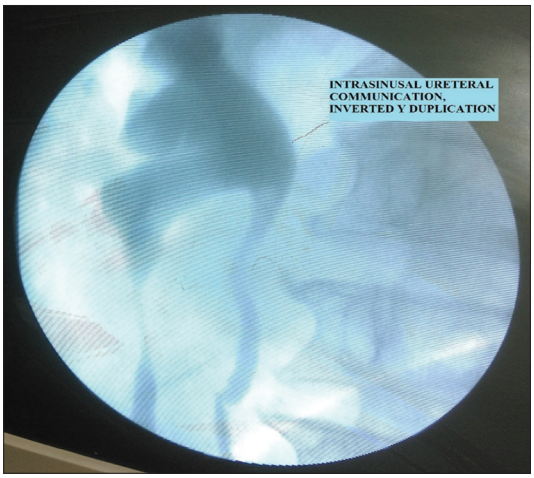

Figure 2: Retrograde ureteropyelography showing Right Pelvicalyceal system communicating with right ectopic ureter.

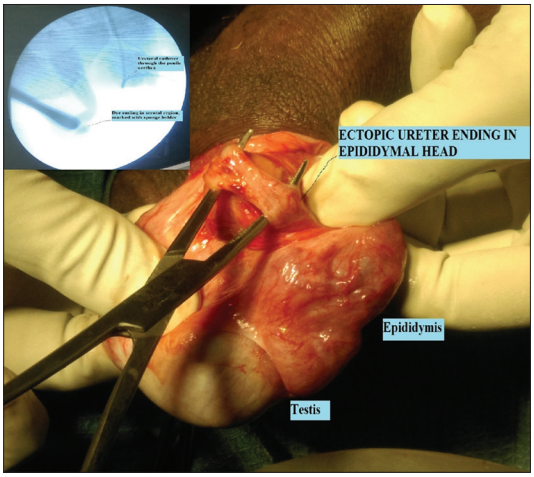

Figure 3: Intra-op picture showing ectopic ureter ending in epididymal head with inset showing the same finding on fluoroscopy.

DISCUSSION

Understanding the potential relationships of the abnormal ureter associated with ectopia which occurs because of aberrations in normal development of Wolffian and Mullerian ducts, ureteral bud, urogenital sinus, and bladder can facilitate clinical interpretation of these conditions. Ureteral duplication occurs when a single ureteral bud branches prematurely during its ascent or when two distinct ureteral buds arise from the Wolffian duct. When the buds are widely separated, one or both will be ectopic. If the superior bud projects from an abnormally high position along the mesonephric duct, caudal ectopia is the outcome. Ureteral communication in a duplex system is usually intravesical, but it may be extravesical too. Extravesical ectopia has been reported more commonly in female patients with urethra and vestibule being the most common drainage sites in 69% with associated complaints of urinary incontinence [7]. Extravesical drainage sites for males include seminal vesicles, epididymis, and ejaculatory duct [8]. However, inverted Y duplication with extravesical ectopia has been reported in only two patients till now. The ectopia was to the seminal vesicle in one and to the epididymis in the other [9]. Associated stone disease in the atretic segment of ureteral ectopia was reported by Suzuki *et al*. [10]. In another case, inverse Y ureter was reported with ectopic ureter opening into the ureterocele [11]. In this context, knowledge of the sites of ectopic insertion may be useful in planning imaging and surgical correction. Regarding non-functioning lower pole moiety, the ultimate maturation of renal blastema is dictated by its interaction with ureteral bud, and when it is altered, expected transformation of blastema to normal nephron does not occur [12]. That is why, this patient had an associated non-functioning lower polar segment. Symptoms of an ectopic ureter that opens into the genital tract are epididymo-orchitis, pelvic pain, frequency and urgency, painful ejaculation, and epididymitis, but they may not appear until puberty. Flank pain, fever and abdominal mass are common presentations of ectopic ureters left undetected until a later age [13]. Hence, detection of this pathology remains difficult because of its uncharacteristic symptoms and high risk of improper interpretation of the diagnostic images. Management includes nephrectomy of the associated dysplastic renal segment along with ectopic ureterectomy. Long-term prognosis is good as the patient becomes totally asymptomatic after proper surgical management.

CONCLUSION

The present case is therefore exceptional from this point of view; in that epididymal ureteral ectopia in partial ureteral duplication, in the fifth decade of life, is detected on unusual presentation masquerading as chronic epididymo-orchitis and thus is being reported. Awareness of the possible coexistence of ureteral and Mullerian abnormalities is important as well as the recognition of the possible effects of abnormal ureteral development on bladder and urethral development which would be essential in determining surgical strategies.

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