CASE REPORT

Unilateral complete duplication of ureter and duplex kidney: A case report

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Abstract

Duplication of ureters is an uncommon congenital anomaly of the excretory system. Previous necropsy studies have showed its incidence ranging from 0.8% to 5%. Duplicated ureter (partial or complete) associated with double pelvicalyceal system draining the single renal parenchyma is known as duplex kidney. Here we report a rare case of complete duplication of the right ureter with two renal pelvises during routine cadaveric dissection of 1st MBBS, in an approximately 65 years old male cadaver. The ureters showed separate openings in the urinary bladder. Complete duplication of ureter with duplex kidney is a rare congenital anomaly. It is associated with various diseases like vesicoureteral reflux, urinary tract infection, nephrolithiasis, ureterocele, etc. The diagnosis of this congenital malformation is difficult due to the mild nature of the disease and hence may remain unrecognised. Clinicians must be aware of this variation for successful diagnosis and management of diseases.

Keywords: Duplication of Ureter, Duplex Kidney, Ureter, Weigert-Meyer law

Introduction

Ureter is a long tubular structure with thick muscular wall and a narrow lumen, extending from renal pelvis to urinary bladder [1]. Embryologically, duplex kidney with complete duplication of ureter develops when two separate ureteral buds arise from a single mesonephric duct. Whereas, if a single ureteral bud proximal to the ampulla bifurcates, it will form a duplex kidney with a bifid pelvis or ureter [2]. The incidence of unilateral duplication is six times higher than bilateral duplication; partial duplication is slightly more prevalent than complete duplication. It is more common in females than males [3]. Thus, our case is unique being unilateral complete duplication of ureter with duplex kidney in a male cadaver. Hence, recognizing and reporting of such rare cadaveric variations will significantly help in diagnosis and management of recurring urinary tract diseases with a great success.

Case Report

During the routine dissection of cadavers in the 1st MBBS class at our institute, we observed this case in an approximately 65-year-old male cadaver. During dissection of the abdomen and pelvic region along the posterior abdominal wall, a complete duplication of the right ureter was noted. One of the ureters was arising from the upper pole, and the other from the lower pole of the corresponding kidney. This was reconfirmed by taking the coronal section of kidney (Figure1). The ureters were running parallel to each other till they passed the pelvic brim. On entering the lesser pelvis, they crossed each other before opening into the urinary bladder. The interior of urinary bladder was exposed to note ureteral orifices. Both these ureters were draining separately into the urinary bladder. Normal tap water was injected with 20cc syringe to note the ureteral drainage. It was found that the orifice of the ureter draining the

upper pole was located inferior and medial to the orifice of the ureter draining the lower pole. The finding was normal with left ureter. No other significant anomalies were found (Figure 2).

Figure 1: Coronal section of right and left kidney (RK-Right kidney, LK-Left kidney, 1-opening of upper pole ureter, 2-opening of lower pole ureter, 3-opening of left ureter)

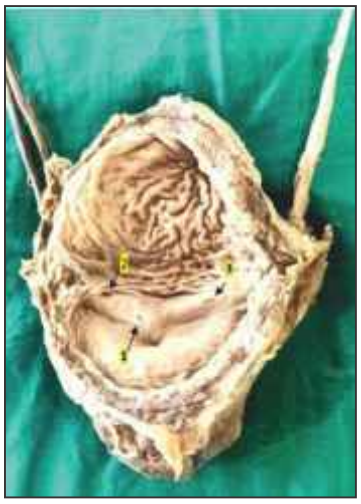

Figure 2: Interior of urinary bladder (1-opening of upper pole ureter, 2-opening of lower pole ureter, 3-opening of left ureter)

Discussion

Embryologically, the genitourinary system begins with the formation of pronephros and mesonephros. The two functional components of the adult kidney develop as the collecting part (ureter, pelvis, major and minor calyces) from ureteric bud and the excretory part (glomerulus, capsule and nephron tubules) from metanephric blastema at around 5th week of intrauterine life [4]. The expression of glial cell line-derived neurotrophic factor ligands of metanephric mesenchyme interacting with Ret/GFRa1 receptor complex situated on Wolffian duct epithelium through a restricted signalling pathway mediates the differentiation of ureteric bud from distal portion of the Wolffian duct. Multiple ureteric buds formation resulting into formation of multiple collecting systems occurs due to unrestricted signalling. Further successive division of the ureteric bud results in the formation of the ureter, renal pelvis, and major and minor calyces. Early splitting of ureteric bud results in duplication of ureters, and this may be complete or incomplete [5]. In case of double ureters, the lower ureter opens in the bladder at normal site, whereas upper ureter migrates more caudally due to caudal shift of the terminal part of the mesonephric duct and opens in the ectopic position. This is due to the fact that the terminal part of mesonephric duct undergoes loop formation in the posterior wall of the urinary bladder [6]. The ectopic ureter can open at various sites in a male or female [7]. Prakash *et al*. observed that in the majority of subjects with complete double ureters, the orifices of the ureters draining the upper pole open inferior and medial to the orifice draining the lower pole of

the kidneys stated by Weigert-Meyer law [8]. In our case the ureters perfectly followed the Weigert-Meyer law. Similar finding was also reported by Naga in her cadaveric case report [5]. While Matsuyama N, reported a case of bilateral duplication of ureter violating the Weigert-Meyer law, where two ureters merged before ending at ureterovesical junction [8]. The exception to Weigert- Meyer law was also noted by Slaughenhoupt *et al*., and Avni, Matos, Rypens *et al*. [7] as an accidental finding of ectopic ureter opening in a male and female patient respectively.

Dahnert reported sixty times more occurrence of complete duplication of ureter in first degree relatives of a patient with this anomaly. Bruno *et al*. stated that ureteropelvic obstruction is more common when a duplex kidney exists and can be inherited as an autosomal dominant pattern [2]. Hascalik *et al*. mentioned as the ureteral duplication may be genetically determined by autosomal dominant trait with incomplete penetrance. The largest known series of duplex ureters was published by Lowsly and Kirwori, reporting 18 cases of duplex ureters [5].

Various studies have shown that patients with an incomplete duplication of ureter are more prone to develop the ureteroureteric reflux while a complete duplication of ureter is associated with vesicoureteric reflux. The duplication of ureters is also associated with other congenital anomalies like Goltz syndrome, high cephalad kidney, external ear anomalies and hemivertebra [5]. In a duplex kidney drained by double ureter, lower moiety is more frequently affected in pelvic-ureteric junction obstruction as compared to the upper moiety due to the dominance of the lower pole system in majority of the cases [2]. Also, Sannathimmappa *et al*. said in his study that congenital malformations are one of the important risk factor for the development of urinary tract infections in children [9]. This case report is prepared according to CARE Guidelines [10].

Conclusion

The clinicians must have detailed knowledge about the normal anatomy and variations of kidney, ureter and urinary bladder for diagnosis and management of the recurrent cases of urinary tract diseases. It is of immense importance to avoid injuries to ureter during any of the surgical or radiological interventions.

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How to cite this article:
Nomulwar SG, Shinde VD. Unilateral complete duplication of ureter and duplex kidney: A case report. *J Krishna Inst Med Sci Univ* 2025; 14(2):160-163

Submitted: *21-Jan-2025* ***Accepted:*** *21-March-2025* ***Published:*** *01-April-2025*