Case Report

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Rare presentation of ureteral duplication: stone localization in the upper moiety

Sunayana Chatterjee*, Hrishikesh Deshmukh, Vaibhav Thorat

Department of Urology, Bharati Vidyapeeth Medical College and Research Centre, Pune, Maharashtra, India

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*Correspondence:

Dr. Sunayana Chatterjee,

E-mail: sunayana.october13@gmail.com

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ABSTRACT

This case report describes a rare exception to the typical presentation of complete ureteral duplication, a relatively uncommon condition in comparison to single ureter or partial duplication. The patient presented with right flank pain and imaging revealed a duplex collecting system with a stone located in the upper moiety. Upper moiety complications, such as ectopic ureteric insertion and multicystic dysplastic moiety, are usually observed in duplex kidneys. Contrastenhanced imaging and endoscopic treatment were utilized for accurate diagnosis and management. The case deviates from the expected pattern based on the Weigert-Meyer rule, emphasizing the importance of considering unique presentations in ureteral duplication cases.

Keywords: Duplex system, Upper moiety, Weigert-Meyer rule, Hydronephrosis

INTRODUCTION

Congenital abnormalities of the kidney and urinary tract (CAKUT) comprise a highly diverse group of diseases characterized by various phenotypes in duplex systems (Figure 1). The etiology of duplex kidneys can be traced back to the initial induction steps of the ureter, wherein an additional ureteric bud emerges in a rostral position relative to the normal outgrowth. This paradoxical phenomenon, known as the Weigert-Meyer rule, leads to the upper (abnormal) kidney pole draining into the bladder distal to the orifice of the lower kidney pole.

Remodelling at the future ureter-bladder junction during development plays a significant role in this occurrence, as the ureter inserts into the developing bladder and undergoes ascending movement. Mackie and Stephens proposed a model explaining this process. Proper positioning of the ureter within the bladder is essential for the formation of a normal trigone, which prevents ureteric

reflux caused by a malfunctioning valve or a too-short ureter tunnel.

A prevalence of duplex kidneys is between 0.2 and 2% in the general population, with females being affected twice as frequently as males. It has an incidence rate of 0.8% in the healthy adult population. Duplex system is seen in 2–4% of patients investigated for urinary tract symptoms. Complete duplication is seen in 40% of cases, whereas partial duplication is seen in 60% of the duplication. Generally, duplex renal systems are asymptomatic and diagnosed incidentally. It has an incidence of twice and the system in the system in the system in the system in the system is seen in 40% of the duplication.

Overall, understanding the complex etiology and manifestations of duplex kidneys is crucial for accurate diagnosis and appropriate management of associated complications such as vesico-ureteral reflux (VUR) and hydronephrosis. We have discussed a case of complete duplex system with hydronephrosis due to ureteric calculus.

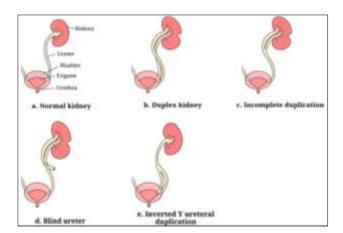


Figure 1: Classification of duplex kidney anatomy; (a) in comparison to a normal kidney, (b) complete duplication results in a duplex kidney characterized by two poles that drain into two ureters, (c) in cases of incomplete duplication, a Y-shaped ureter is formed, (d) blind ureters do not connect to the bladder, and (e) in the rare occurrence of inverted Y-ureteral duplication, two ureters merge before entering the kidney.

CASE REPORT

Our index case involves a 37-year-old female patient without any comorbidity who presented to the outpatient department (OPD) with a one-month history of right flank pain. The patient experienced fever spikes one month ago and multiple episodes of vomiting. Ultrasonography of the kidney, ureter, and bladder (USG KUB) revealed a normal-sized, shaped, and echo textured right kidney without hydronephrosis. However, a linear calcification measuring 21 mm with posterior acoustic shadowing was observed at the region of the right vesico-ureteric junction. Mucosal edema of the vesico-ureteric junction was also noted. Computed tomography of the kidney, ureter, and bladder (CT KUB) further confirmed the normal characteristics of the right kidney and the presence of a duplex collecting system (Figure 2a and b). Additionally, a 12.4×6 mm calculus (CT value: 1300-1400 HU) at the right vesico-ureteric junction resulted in a hydro ureter.

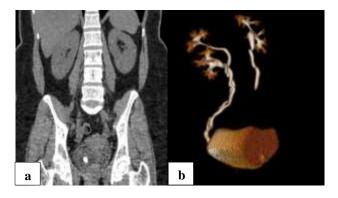


Figure 2: CT-KUB showed the following (a) stone in right lower ureteric orifice, and (b) 3D reconstruction showing complete duplication.

Subsequently, the patient underwent a right ureteroscopy lithotripsy (URSL) procedure, which revealed two ureteric orifices on the right side of the trigone. The upper moiety presented a calculus measuring approximately 2 cm. A guide wire was inserted through both orifices to demonstrate the anatomy (Figure 3a). Retrograde pyelography (RGP) confirmed the complete duplex system without any signs of hydroureter or obstruction (Figure 3b and c). The patient currently remains in a stable condition with no reported complaints.

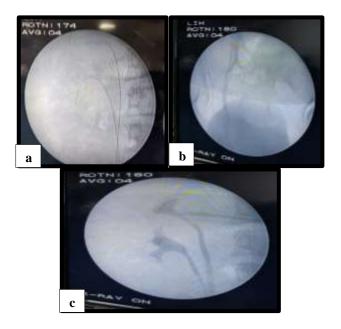


Figure 3: Retrograde pyelography findings (a) guide wire inserted through 2 orifices, (b) and (c) complete duplication with duplex moiety.

DISCUSSION

Complete ureteral duplication is a relatively rare condition compared to a single ureter or partial duplication. Patients with ureteral duplication often present with various complications specific to each moiety. Upper moiety complications include ectopic ureteric insertion, with or without an ureterocele, and multicystic dysplastic moiety. Lower moiety complications include vesico-ureteral reflux (VUR), renal scarring, and pelviureteric junction (PUJ) obstruction. VUR and renal scarring are the most common complications found in the lower moiety of duplex kidneys.³ Contrast-enhanced intravenous pyelogram (IVP) can effectively detect duplex ureteric systems and provide essential information about the collecting system, particularly in cases of renal stones.⁴ Contrast imaging is also beneficial for endoscopic treatment of ureteric and renal calculi, potentially saving time and preventing misinterpretations.4 In most cases, the orifice draining the lower moiety of the renal duplex is situated superiorly and laterally to the orifice draining the upper moiety, consistent with our patient's presentation. The lower moiety tends to be more frequently affected than the upper pole in duplex kidneys with double ureters, primarily due to the higher prevalence of complications associated with

the lower moiety.⁵ However, our case deviates from the typical pattern as the stone was found in the upper moiety, contrary to the expected outcome based on the Weigert-Meyer rule. This unique scenario highlights a rare exception to the rule.

CONCLUSION

This case report highlights the complexities of ureteral duplication and its associated complications. The importance of accurate diagnosis through contrast-enhanced imaging and the potential benefits of endoscopic treatment are emphasized. Understanding the complex etiology and manifestations of duplex kidneys is essential for appropriate patient care.

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