Case Report

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Unilateral Incomplete Bifid Ureter Presenting with Calculus in Right Kidney with Hydronephrosis: A Rare Case Report

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Abstract

The anomalies associated to the duplications of ureter and kidneys result from an early division of ureteric diverticulum. The extent of duplication is decided by level of the division of ureteric diverticulum and metanephric blastema. In this case report, a bifid ureter was found on the right side of a male patient. Based on the available literature, the present case is very rare. Usually, it is detected at autopsy or as radiological finding if associated with a wide variety of clinical manifestations. The knowledge of this anomaly should be borne in mind, to deal with a case of repeated urinary infections, urinary reflux disorders, hydronephrosis or urinary calculi.

Key words: Bifid ureter, Calculus, Duplex ureter, Malformation

INTRODUCTION

Out of congenital anomalies of the abdominal masses found in neonates, congenital anomalies of the kidney and urinary tract (CAKUT) account for more than 50% and involve about 0.5% of all pregnancies.[1] Malformations of the urinary system are common and comprise about 3% of live births. These various malformations include ectopia, malrotation and other morphological variations such as number of kidneys, ureters, etc. Out of various malformations, Duplex ureter is reported 1 in 125 cases or 0.8% of a non-selected population. Duplex ureter is more common in females, with female to male ratio of 1.6:1 or 62% of females.[2] Duplication of the ureter might be complete or incomplete. If the duplication of a ureter is incomplete, then it has been known as the bifid ureter. Lowsley and Kirwori (1956), reported the incidence of an incomplete duplicate ureter to be 18 out of a series of 4215 autopsies studied. Amongst these 2 were the bilaterally incomplete duplicate, 7 were a unilaterally incomplete duplicate and 8 were the unilaterally complete duplicate.[3] According to recent studies by Russel *et al*., (2000), intravenous pyleograms showed ureteral duplication in an average 3%, on routine examination.[4] Presence of a bifid ureter is often associated with congenital hydronephrosis,[5] or sometimes with contralateral quadrafid ureter,[6] etc.

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CASE REPORT

Here, we present a case with a unilateral bifid ureter. This male patient aged 34 years, presented with pain in the abdomen and on sonography showed calculus in the right kidney with hydronephrosis. In the present case, intravenous pyelography showed dilated right system and normal left functioning system. The right system showed prompt excretion of contrast, double moiety; lower moiety was hydronephrotic with calculus in the ureter. During operation retrograde pyelography and DJ, stenting was performed on the right side. Later, for follow-up, magnetic resonance imaging (MRI) of the person, showed a clear bifid ureter on the right side, with no other associated

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congenital anomaly. For calculus, extracorporal shock wave lithotripsy was performed, and the calculus was totally fragmented. After confirming that we have achieved total clearance, DJ stent was removed.

In our case, the right-sided ureter had two limbs for most of its length. Later, the limbs joined at about a distance of 4 cm from the bladder wall as seen in MRI (Figure 1). The two limbs of this ureter showed their respective pelvis, coming out as separate entities from the hilum. The hilum was relatively longer than the hilum of left kidney. The pelvis of the upper limb had its exit at the upper end of the hilum and that of the lower limb at the lower limit of the hilum. The opening of the ureter into the bladder did not show any abnormality. During MRI examination, no other gross morphological abnormalities of thoracic, other structures of abdominal and pelvic viscera were revealed.

DISCUSSION

In the previous studies, bifid ureter had been detected in association with various congenital anomalies and defects. It has been associated with Goltz's syndrome[7] high cephalad kidney, duplication of pelvis,[8] and unilateral pulmonary hypoplasia.[9] Bifid ureter has also been reported in association with complete duplication of the contralateral ureter (Tundidor Bermúdez, 1999; Borrego *et al.*, 1995).[10]

Many of the investigators have reported this anomaly in association with other disease conditions. Isolated anomalies of the ureter are reported by only few authors. But in this present case, the unilateral incomplete bifid ureter of the right side was associated with no other abnormality.

Developmental Basis

At about 4th week of gestation, evaginations arise from the distal mesonephric duct. These evaginations are ureteric buds, which later interact with their surrounding mesenchyme called metanephric blastema. This interaction between ductal system and mesenchyme is very important and plays a major role in branching of the ureteric bud and subsequent development of the ureter, pelvis, and calyceal system up to collecting tubule of kidney.[11]

Molecular Basis Related to CAKUT

As with most organs, differentiation of the kidney involves epithelial mesenchymal interactions. Epithelium of the ureteric bud from the mesonephros interacts with mesenchyme of the metanephric blastema (Figure 2a).[12] The mesenchyme expresses WT1, a transcription factor that makes this tissue competent to respond to induction by the ureteric bud. Transcription factor WT1 is responsible for production of glial-derived neurotrophic factor (GDNF) and hepatocyte growth factor (HGF, or scatter factor) by the mesenchyme. Later, these GDNF and HGF stimulate branching and growth of the ureteric buds (Figure 2a). The epithelium of the ureteric buds, synthesizes tyrosine kinase receptors RET for GDNF and MET for HGF, by establishing signaling pathways between the two tissues.[13] In turn, the buds induce the mesenchyme via fibroblast growth factor 2 and bone morphogenetic protein 7 (Figure 2b). Both of these growth factors stimulate proliferation of the metanephric mesenchyme while maintaining production of WT1. Conversion of the mesenchyme to an epithelium for nephron formation is also mediated by the ureteric buds through expression of WNT9B and WNT6, which upregulate PAX2 and WNT4 in the metanephric mesenchyme. PAX2 promotes condensation of the mesenchyme preparatory to tubule formation, while WNT4 causes the condensed mesenchyme to epithelialize and form tubules (Figure 2b). Because of these interactions, modifications in the extracellular matrix also occur. These modifications are fibronectin, collagen I, and collagen III are replaced with laminin and Type IV collagen, characteristic of an epithelial basal lamina (Figure 2b). In addition, the cell adhesion molecules essential for condensation of the

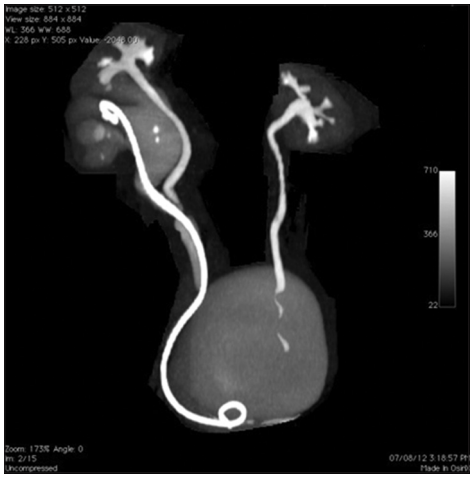

Figure 1: Magnetic resonance imaging

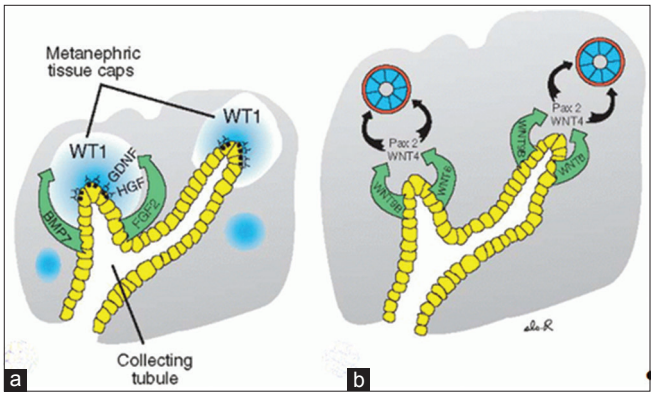

Figure 2: (a and b) Molecular basis of development of excretory system[12]

mesenchyme into an epithelium, syndecan, and E-cadherin, are synthesized.

The crucial event in kidney development is the first signaling process that induces the outgrowth of the ureter from the mesonephric duct.[14] Proteins like Forkhead box protein C1, slit homologue 2, and its receptor round about homologue 2[15] have been reported to confine GDNF expression to the caudal part of the nephric cord in mice studies, and mutations in genes encoding these proteins lead to an expansion of GDNF expression to the rostral part, resulting in multiple ureters.[16]

CONCLUSION

The anomalies pertaining to the duplications of the ureter and kidneys result from an early division of ureteric diverticulum. The extent of duplication depends on level of the division of the ureteric diverticulum and metanephric blastema. The variation reported in our case could be due to incomplete division of right ureteric bud in the cranial part.[12] The two limbs of this bifid ureter are having their respective pelvis coming out as separate entities from the hilum. The hilum of right kidney was relatively longer than the left kidney. The pelvis of the upper limb had its exit at the upper end of the hilum and that of the lower limb at the lower limit of the hilum. As the lower limb of the bifid ureter is dependent part of the system, it is more prone to calculus formation and later infection.

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