Other full case

An H-shaped complete ureteric duplication: embryology or erosion

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Summary

The authors report the case of a duplex urinary collecting system that forms a single chamber about 2 cm proximal to the bladder with distal bifurcation and drainage into the bladder via distinct ureteric orifices. This anatomical variant has not been described before.

BACKGROUND

Duplex systems are the most commonly encountered congenital abnormality of the renal tract, with a reported incidence of 0.8%.¹ The ureters may remain separate throughout their course, entering the bladder by two distinct ureteric orifices. This is referred to as complete duplication. Fusion of the ureters along their length is referred to as either bifid systems or bifid ureters, depending on whether the ureters fuse proximally or distally.

We report an anatomical variant of a complete duplex urinary system that has not been described in the literature previously.

CASE PRESENTATION

A 41-year-old lady was referred with repeated urinary tract infections, intermittent right loin pain and low-grade pyrexia. She was initially investigated with a plain x-ray, intravenous urogram (IVU) and CT of the kidney, ureter and bladder. This demonstrated a normal left renal tract, with a complete duplication on the right (figure 1). A large calculus was noted on the right, in close proximity to the vesico-ureteric junction (VUJ).

At cystoscopy, two ureteric orifices were noted on the right. The ureteroscope was inserted via the medial ureteric orifice and the stone was seen in the distal ureter. The stone was totally fragmented by laser and towards the end of the procedure, to our surprise, the lateral ureter was noted to join the medial ureter to form a common chamber only to bifurcate again proximally. When guidewires were inserted in both ureteric orifices it was clearly identified ureteroscopically that there was an H-shaped complete ureteric duplication (figures 2 and 3).

OUTCOME AND FOLLOW-UP

The patient has been followed up for 2 years postprocedure. Four months following her initial procedure, she had an x-ray IVU for right-sided loin pain, which disclosed hydronephrosis and a proximal hydroureter. This was followed up with a ureteroscopy. Two narrowings were found, which impeded passage of the ureteroscope

and a ureteric stent was passed. This was removed after 1 month, and by a further 2 months the patient was almost symptom free. She no longer suffers from urinary tract infections. Apart from some mild left-sided loin pain, the patient is symptom free.

Figure 1 X-ray intravenous urogram. Complete ureteric duplication noted on right, with a large calculus close to the vesico-ureteric junction.

Figure 2 Retrograde urogram. Contrast noted in both ureters with contrast pooling in common chamber. Guidewire is seen in the medial ureter.

DISCUSSION

The anatomical variant of complete ureteric duplication where the two ureters join to form a single ureter for a short segment proximal to the VUJ has not been described previously. In contrast, the embryology of complete ureteric duplication, bifid systems and ureters, is well described.

Genitourinary system development begins with the formation of the pronephros and mesonephros within intermediate mesoderm. A series of mesenchymal epithelial interactions leads to the development of the Wolffian duct, which elongates caudally to fuse with the cloaca (the precursor of the bladder and urethra).

The distal part of the Wolffian duct within the hindlimb region receives signals from the adjacent metanephric mesenchyme to induce ureteric bud formation. The principle signalling pathway, involves glial cell line-derived neurotrophic factor (GDNF) ligand expression from adjacent metanephric mesenchyme, interacting with Ret/GFR α 1 receptor complex located on the Wolffian duct epithelium.² Clearly to induce ureteric bud formation at the appropriate region, GDNF expression within the metanephric mesenchyme must be restricted. In animal models, loss of function of transcription factors which repress GDNF expression in adjacent mesenchyme, lead to duplex/multiplex urinary collecting systems.² Unrestricted signalling to the Wolffian

duct may elicit formation of multiple ureteric buds, leading to multiplex/duplex collecting systems. However, the molecular signalling defects underlying duplex collecting systems, is not completely understood.

Once the ureteric bud penetrates the metanephric mesenchyme, it branches and continues to branch within the substance of the metanephric mesenchyme to form the collecting system of the metanephric kidney. The remaining ureteric bud between the Wolffian duct and the metanephric mesenchyme forms the ureter. Premature branching prior to entering the substance metanephric mesenchyme is thought to account for the formation of bifid ureters.

We speculate that embryologically the two ureteric buds may have fused close to their origin on the Wolffian duct, and then bifurcated before reaching the metanephric mesenchyme, to account for this variant.

Another explanation could be that the urinary calculus may have eroded through the wall of the two ureters, thus forming a single chamber. Erosion of impacted urinary calculi through the wall of the ureter has been described previously,³ though no such cases eroding into an adjacent duplex ureter have been reported.

Whatever the causal explanation, the possibility of such an anatomical variant must be borne in mind, while managing patients with bifid urinary collecting systems.

Figure 3 Ureteroscopic view inside single chamber. Guidewires seen entering single chamber.

Learning points

- ▶ Duplex urinary systems are the most common congenital abnormality of the renal tract.
- ▶ These can be either complete duplications or bifid ureters/systems.
- ▶ In patients with complete duplications, one should be wary of possible fusion along the course of the ureters.

Competing interests None.

Patient consent Obtained.

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