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Imaging based case report of ectopic urethral ureter in an infant: A case report and literature review

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ABSTRACT

We present a case of ectopic urethral ureteral insertion in a 2-week old female diagnosed on a voiding cystourethrogram after abnormal prenatal and postnatal renal ultrasounds. The case is remarkable due to initial opacification of only the left upper pole moiety without bladder opacification on the voiding cystourethrogram due to direct canalization of the left ureter. Prompt diagnosis is important to prevent frequent urinary tract infections and urinary incontinence with lower ectopic urethral ureter insertions. Voiding cystourethrogram is an important tool in the workup of renal anomalies and is typically used to evaluate for reflux, however, keen attention can reveal other abnormalities such as ureteral ectopia in rare instances.

Key Words: Ectopic ureter, duplicated ureter, Weigert-Meyer, urethral ureter, duplex moiety.

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Introduction

Urogenital tract anomalies are among the most commonly diagnosed anomalies in the fetus and neonate with the advancement of screening sonography. Ectopic ureter is one of the few conditions often diagnosed in neonates and children, though occasionally it can be diagnosed in adults. Ectopic ureter is described as any ureter that does not insert in the trigonal region of the urinary bladder [1]. Ureteral ectopia is a congenital anomaly caused due to abnormal caudal migration of the ureteral bud during embryogenesis. Incidence within neonates is around 1 in 2000 with the rate in the general population ranging from 1 in 2000 to 4000 [2]. Overall, there is a significant female predominance, with females being affected approximately 6 times more than males, though this may be falsely elevated as males are often asymptomatic [3]. In females, the insertion sites include the bladder, urethra, vestibule, vagina or rarely, the uterus [4]. For males, ectopic insertion sites include the bladder, posterior urethra, seminal vesicle, vas deferens, or ejaculatory duct [5].

Ectopic ureteral insertions are ubiquitously associated with duplicated renal collecting systems, especially in the western world with association rates around 80-85% [2]. However, in Asia, a majority of cases of ureteral ectopia are not linked to duplicated collecting systems [6]. The discrepancy of association with a duplicated collecting system between the

western world and Asia is not well elucidated. Renal anomalies, including horseshoe kidney and cross-fused renal ectopia, are not typically associated with ureteral ectopia. Duplicated renal collecting systems are comprised of upper and lower pole moieties, each of which is predisposed to its own pathology and insertion pattern. Insertion of the moieties follows the classical Weigert-Meyer rule, with the upper pole moiety inserting inferomedial while the lower pole moiety will insert superolateral [2]. The upper pole moiety tends to obstruct, while the lower pole moiety tends to reflux.

Ureteroceles are associated with ectopic insertion of the upper pole moiety in a duplicated collecting system, but are not always seen [2]. Additionally, the upper pole moiety classically obstructs, which predisposes the it to secondary pathologies such as calculi. On the other hand, the lower pole moiety often refluxes, increasing risk for recurrent urinary tract infections.

Ureteral ectopia in non-bladder locations also can present with urinary incontinence, especially in females. Within the literature, several cases of ectopic ureteral insertion are described by Senel in female patients with one case of insertion upon the posterior urethra. However, no case report that we are aware of demonstrates initial direct visualization of the ectopic ureter with voiding cystourethrogram.

Case report

A 2-week old female born via normal spontaneous vaginal delivery at 39 weeks 1 day presented to outpatient radiology for a voiding cystourethrogram for further workup due to abnormal findings initially noted on a prenatal sonographic survey and confirmed with postnatal ultrasound. Prenatal sonographic survey was not available for review as it was performed at an outside hospital and not archived in a digital format. Records indicated that prenatal sonographic survey demonstrated an abnormal left kidney concerning for a duplicated collecting system or upper pole cyst.

Postnatal renal ultrasound is seen in Figures 1A and 1B which demonstrates dilation of the upper pole collecting system and distal left ureter consistent with left hydroureteronephrosis.

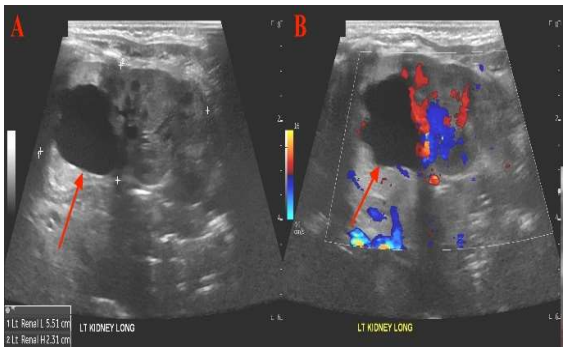

Fig. 1. 2-week old female with abnormal prenatal ultrasound for renal evaluation. **(A)** Grayscale longitudinal ultrasound of the left kidney demonstrating severe dilation of the renal pelvis (red arrow). **(B)** Color Doppler longitudinal ultrasound of the left kidney which does not demonstrate flow in the dilated upper pole pelvicalyceal system.

These findings were suspicious for a duplicated left collecting system and as a result, voiding cystourethrogram was recommended. Voiding cystourethrogram was performed with canalization of the urethra using a 5 French Foley catheter. A dilated left ureter was seen inserting near the location of the urinary bladder though no evident insertion site was demonstrated on ultrasound in Fig. 2.

Initial fluoroscopic image following contrast instillation is seen in Fig. 3A which demonstrates opacification of the left ureter and collecting system. There was severe

dilation of the left renal pelvis and blunting of the calyces with severe hydroureter.

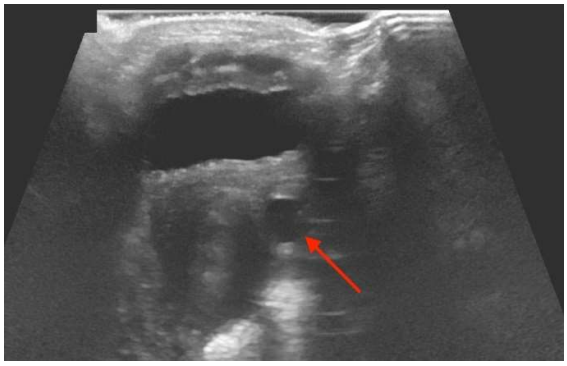

Fig. 2. 2-week old female with abnormal prenatal ultrasound for renal evaluation. Grayscale ultrasound of the urinary bladder which demonstrates a dilated left ureter posteriorly (red arrow) without evidence of direct connection to the bladder.

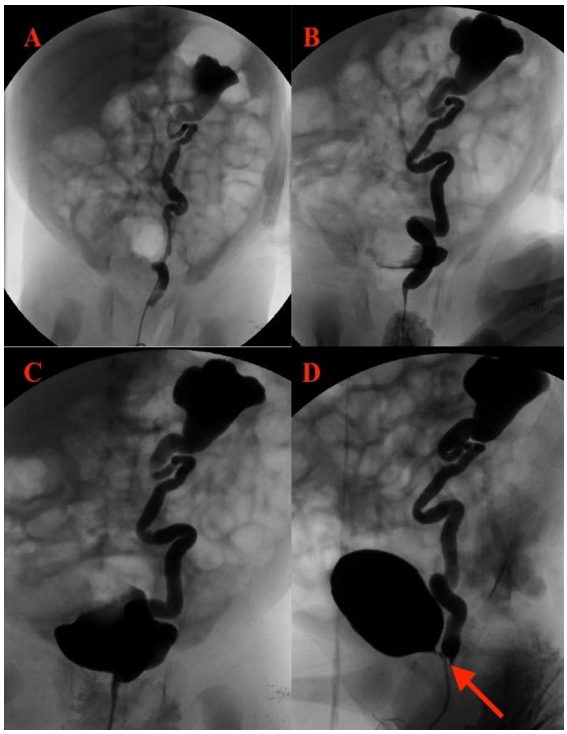

Fig. 3. 2-week old female with hydronephrosis and suspected duplicated collecting system on renal ultrasound. Voiding cystourethrogram images obtained after contrast instillation pre-void. **(A)** Initial anteroposterior projection image obtained from voiding cystourethrogram (VCUG) that demonstrates Foley catheter tip within the distal left ureter. Subsequent contrast instillation opacifies the left ureter and left upper pole renal pelvis and calyces. There is severe hydroureteronephrosis. **(B)** Subsequent anteroposterior projection image obtained from voiding cystourethrogram that was obtained with contrast instillation while the Foley catheter was being withdrawn which demonstrates early contrast opacification of the urinary bladder. **(C)** Voiding cystourethrogram anteroposterior projection image obtained with continued contrast instillation demonstrates eventual filling of the urinary bladder. Opacification of the left upper pole collecting system and ureter remains. **(D)** Lateral projection image from voiding cystourethrogram after contrast instillation demonstrates the distal aspect of the left ureter clearly identifying its insertion upon the membranous urethra (red arrow) with no connection to the urinary bladder.

The urinary bladder was not visualized in this image. Further review of this image indicated that the Foley catheter was within the left ureter. Subsequently, contrast was continually instilled while the catheter was withdrawn, eventually opacifying the urinary bladder as seen in Figures 3B and 3C. Lateral projection image was obtained once the urinary bladder was sufficiently distended, seen in Figures 3D with clear demonstration of the ectopic insertion of the left ureter upon the proximal membranous urethra. Imaging was diagnostic of a complete left ureteral duplication with ectopic insertion on the posterior urethra.

Voiding cystourethrogram image obtained post-void seen in Fig. 4 demonstrates minimal residual contrast within the urinary bladder. Left upper pole collecting system and ureter remain opacified with contrast. No evidence of right sided vesicoureteral reflux was demonstrated.

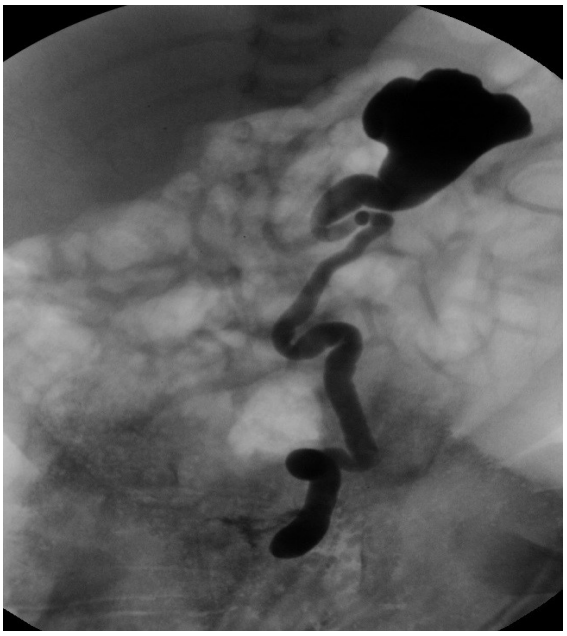

Fig. 4. 2-week old female with hydronephrosis and suspected duplicated collecting system on renal ultrasound. Anteroposterior projection image from voiding cystourethrogram obtained post-void demonstrates minimal residual contrast within the urinary bladder. Left upper pole collecting system and ureter remain opacified with contrast. No evidence of vesicoureteral reflux.

Discussion

Renal abnormalities should initially be worked up with sonography, which is able to directly characterize or visualize secondary signs of an underlying etiology. Renal ultrasound can identify obstruction or ureteroceles which are secondary signs commonly associated with ureteral ectopia [4]. Additionally, sonography can at least suggest the diagnosis of a duplicated collecting system, in which ureteral ectopia is likely to coexist [4]. Generally, voiding cystourethrogram (VCUG) is the follow-up exam recommended after an abnormal renal ultrasound to evaluate for vesicoureteral reflux. Intravenous pyelogram (IVP), also known as intravenous urogram (IVU), is generally less utilized as the upper pole moiety is often nonopacified due to renal failure [7]. With ectopic ureteral insertion outside of the urinary bladder, there should be no visualization of the ureter even with presence of reflux on a voiding cystourethrogram [3]. Therefore, in ureteral ectopia, the VCUG exam is expected to be normal. Intravenous pyelogram may opacify the duplicated collecting system well if it remains functional and can rarely demonstrate the ectopic insertion site. However, as noted above, IVP is generally less utilized due to decreased sensitivity with renal failure from obstructive uropathy leading to nonopacification of the obstructed moiety. VCUG was diagnostic in our case and especially remarkable was the direct catherization of the ureter through the ectopic urethral insertion, allowing for direct visualization of the ectopic ureter, a finding not commonly expected even with ureteral ectopic on VCUG.

Treatment options depend on the age of the patient and symptomatology. Additionally, the function of the kidney which is drained by the ectopic ureter should be assessed. Generally, in pediatric populations, surgery is the treatment of choice. On the other hand, conservative therapy is preferred in adults. For surgical treatment, if renal function is preserved in the affection portion, ureteroneocystostomy is recommended, otherwise, partial nephroureterectomy is the alternative in nonfunctional renal systems [2]. Considerations of surgical approach such as laparoscopic versus open are up to the experience and discretion of the treating surgeon.

Differential diagnostic considerations when working up renal anomalies in a neonate include duplication of the collecting system, vesicoureteral reflux, urinary bladder diverticulum along with ureteral ectopia.

Duplication of the collecting system is a common congenital renal tract anomaly that may have an incomplete or complete collecting system duplication ranging from mere renal pelvis duplication to completely duplicated ureters. Imaging features are variable depending on the level of duplication, however, similar to the other differential diagnostic considerations, imaging workup is the same, including renal sonography, VCUG and possibly IVP. Complete duplication will always be concurrent with ureteral ectopia. However, in incomplete duplication, renal sonography may demonstrate ureteroceles and hydroureteronephrosis if present. Vesicoureteral reflux is reflux from the urinary bladder to the upper urinary tract more commonly seen in children and predisposing them to urinary tract infections. The primary modality to evaluate for vesicoureteral reflux is VCUG, which will demonstrate micturitional contrast reflux into the ureter with varying severity. A possible confounding factor in this diagnosis is that reflux can be seen in the lower pole moiety of duplicated collecting systems which can coexist with ectopia in complete duplication. Urinary bladder diverticulum are outpouchings of the wall with mucosal herniation and often are incidental findings that can be seen on computed tomography, magnetic resonance, ultrasound and IVP imaging. In contrast to ureteral ectopia, urinary bladder diverticula end in a blind pouch with no connection to the collecting system. Ureteral ectopia can present with urinary incontinence, recurrent urinary tract infections or can be seen with abnormal fetal ultrasound. Renal sonography can demonstrate hydroureteronephrosis, ureteroceles or duplicated collecting system in association with ureteral ectopia. With ureteral ectopia, VCUG can be unremarkable, demonstrate vesicoureteral reflux or hydroureteronephrosis. As previously discussed, ectopia will nearly always demonstrate a duplicated collecting system, whether it be seen on sonography, VCUG or IVP.

Conclusions

Ectopic ureteral insertion is a congenital anomaly commonly associated with a duplicated collecting system with a female predominance. Renal ultrasound can demonstrate hydroureteronephrosis or ureterocele while voiding cystourethrogram can demonstrate obstruction or reflux that is often associated with ectopic ureters. No case report that we are aware of demonstrates initial direct visualization of the ectopic ureter with voiding cystourethrogram.

Compliance with ethical statements

Conflicts of Interest: None.

Financial disclosure: None.

Consent: Patient confidentiality has been maintained and written consent has been obtained from the patient's parents for the publication of patient information and clinical pictures and can be provided as required.

Open Access Statement

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