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Case Report

Weigert-Meyer Law Violation with Lower Pole Obstruction and Ectopia

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ARTICLE INFO

Article history:

Received: 24 January, 2020

Accepted: 15 February, 2020

Published: 28 February, 2020

Keywords:

Weigert-Meyer law

ectopic ureter

hydronephrosis

duplicated collecting system

lower pole obstruction

ABSTRACT

Duplication of the collecting system is the most common congenital abnormality of the upper urinary tract (0.8-2%), with unilateral duplication more common than bilateral [1-4]. In complete duplication, ureters typically follow the Weigert-Meyer rule of the upper pole inserting caudomedial (may be obstructed) and the lower pole inserting cranio-lateral (may reflux) in the bladder. Very rarely is there deviation from this pattern. Few cases of uncrossed double ureters with lower pole dysplasia, ureterocele, or ectopia have been reported [5-8]. We report a breach in the law with a case of an asymptomatic 1-month old new-born male with lower pole ureteral ectopia and obstruction. We discuss the importance of thorough work up with emphasis on differential diagnoses and care team-based decisions in the evaluation of paediatric hydronephrosis.

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Background

Duplication of the collecting system is the most common congenital abnormality of the upper urinary tract, with incidence reported to be between 0.8-2% [1-4]. Unilateral duplication is more common than bilateral [4]. Surgical correction is sometimes indicated in patients in order to preserve renal function and prevent complications, like infection [1, 2, 9]. In complete duplication, the ureters typically follow the Weigert-Meyer general rule of the ureters crossing with the upper pole ureter ending up more caudal and medial and the lower pole ureter inserting more cranio-lateral in the bladder. A case of uncrossed double ureters with lower pole dysplasia have been reported in a symptomatic older child [5]. Ureterocele and ectopia to a vas deferens and epididymis have been previously described, though there was no evidence of hydronephrosis of the lower pole in either case [6-8]. We report a case of uncrossed double ureters with nonfunctioning lower pole and ureteral obstruction at the level of the ureterovesical junction (UVJ), breaching the law.

Case Presentation

A healthy new-born term male presented with left prenatal hydronephrosis. Pregnancy was complicated by gestational diabetes. He

has no family history of genitourinary anomalies. His renal ultrasound at one month of age demonstrated duplicated system with grade 2 left upper pole hydronephrosis and grade 4 left lower pole hydronephrosis (Figure 1).

Investigation

Two weeks later, he underwent repeat renal ultrasound (RUS), voiding cystourethrogram (VCUG), and nuclear renal scan with Lasix washout (NRS) to further characterize his hydronephrosis. His RUS was similar in appearance, and VCUG was negative for reflux. His NRS showed differential function of 55% on the left and 44% on the right. The left side was overestimated due to the hydronephrosis as well as delayed excretion and perfusion in the left lower pole.

To further delineate the unusual anatomy, magnetic resonance urogram (MRU) was obtained, which demonstrated known dilated left hydronephrosis of the lower pole with distal peak inserting laterally near the bladder neck (Figure 2a). The distal peak is an indication of an adynamic segment causing UVJ obstruction which has resulted in no salvageable renal parenchyma in left lower pole (Figure 2b & 2c).

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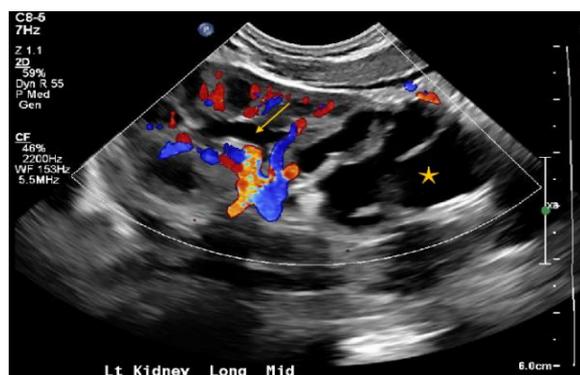


Figure 1: Renal ultrasound at one month of age demonstrating grade 2 left upper pole hydronephrosis (arrow) and grade 4 lower pole hydroureteronephrosis (star).

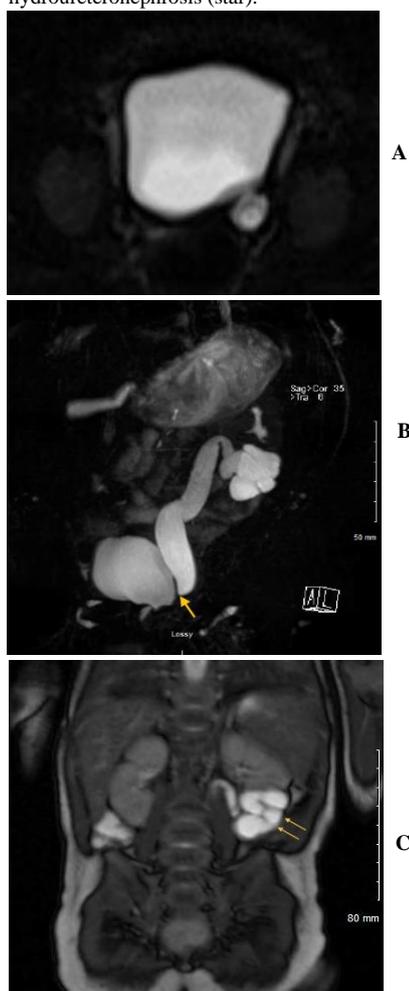


Figure 2: **A)** Axial magnetic resonance urography (MRU) showing insertion of the dilated left lower pole distal ureter inserting laterally near the bladder neck. **B)** MRU three-dimensional reconstruction demonstrating duplicated system with no evidence of functional renal parenchyma. The dilated ureter comes to a peak distally (arrow), likely an adynamic segment near the ureterovesical junction. **C)** MRU coronal image showing no salvageable renal parenchyma of the lower pole (double arrows).

Throughout the infant's course of over a year, he has had no urinary tract infections (UTI). He has been growing and developing normally. Eventually, his left upper pole hydronephrosis resolved. Since he has been asymptomatic and there is no evidence of salvageable lower pole renal parenchyma, the child's parents and the care team have elected for observation of the left lower pole hydroureteronephrosis. He will continue to be monitored with serial ultrasounds to ensure normal drainage and appearance of the left upper pole moiety. Intervention may be considered if the child develops recurrent UTI, kidney stones, or progressive upper pole dilation.

II Differential Diagnosis

Antenatal hydronephrosis has many root causes or differential diagnoses with similar appearances. Characteristics of the hydronephrosis and sequential imaging (RUS, VCUG, NRS, MRU) can help narrow down the differential diagnoses. Bilateral versus unilateral hydronephrosis can have separate implications. Both bilateral and unilateral hydronephrosis can be idiopathic and resolve spontaneously with time. Unilateral hydronephrosis should be evaluated with postnatal RUS in the first 3-8 weeks of life, and bilateral hydroureteronephrosis should be promptly evaluated with RUS and VCUG [10]. It is also important to remember that a postnatal RUS in the first 48 hours of life may underestimate the degree of hydronephrosis due to the physiologic oliguria of the newborn [10].

Vesicoureteral reflux (VUR) is a common cause of hydronephrosis and hydroureteronephrosis. In this case, urine is free to travel back up the ureter to the kidney, resulting in damage to the kidney over time with back pressure. VUR is diagnosed on VCUG, but the timing of obtaining a VCUG versus starting prophylactic antibiotics for mild to moderate hydronephrosis is unclear [10]. Neonates with severe hydronephrosis should be placed on microbial prophylaxis and evaluated with VCUG [10]. Risk of VUR increases with family history of VUR and can be unilateral or bilateral. If VUR is diagnosed, per American Urological Association guidelines, a child less than one year of age should be placed on continuous prophylactic antibiotics if they have had a febrile UTI or if they have severe VUR (Society of Fetal Urology Grades III-V) [11]. If they have mild VUR (SFU Grades I-II), children less than one year of age may be offered prophylactic antibiotics [11]. Circumcision may also be offered to boys less than one year of age to reduce risk of UTI in the first year [11]. In the case of our neonate, unilateral hydronephrosis was present on antenatal RUS and postnatal RUS with noted duplication of collecting system; his VCUG was negative, ruling out VUR as the cause of his hydroureteronephrosis.

Another cause of unilateral hydronephrosis can be secondary to an obstruction anywhere along the course of the ureter. Obstruction at the ureteropelvic junction (UPJ) is a common cause of hydronephrosis and may be caused by high insertion of the ureter on the renal pelvis or by a crossing vessel. Obstruction at this level may also be due to benign fibroepithelial polyps within the ureter. Obstruction at the level of the ureterovesical junction (UVJ), other than the aforementioned bladder wall thickening and ureteroceles, can be secondary to an aperistaltic segment of ureter resulting in an obstructed non-refluxing megaureter. When the obstruction is in the distal ureter, dilation of the ureter is observed. Obstruction of the ureter at any point along its course may be diagnosed with NRS after ruling out VUR with VCUG. Any of these

causes of unilateral hydronephrosis can also occur bilaterally [10]. NRS was obtained for this child at 6 weeks of age, which demonstrated duplicated left collecting system with poor perfusion of the lower pole; the left upper pole was hydronephrotic but appeared to drain appropriately.

Bilateral hydroureteronephrosis would raise suspicion for posterior urethral valves (PUV), especially if accompanied by oligohydramnios in utero in a male fetus. The obstruction is at the level of the bladder outlet; any time bladder outlet obstruction is suspected; the neonate should be promptly decompressed (with catheter) and started on antibiotic prophylaxis before radiographic intervention [10]. With outlet obstruction, the bladder becomes thickened and trabeculated, which can secondarily cause ureterovesical junction obstruction. Thickening of the bladder may also occur with neurogenic bladder, which may be due to spinal dysraphism (spina bifida, meningocele, myelomeningocele). Voiding in the first 24 hours after birth is crucial for neonates with these diagnoses; they may require a catheter to ablate or bypass the valves or drain the bladder if unable to void.

Another cause of bilateral hydroureteronephrosis could be a large unilateral ureterocele or bilateral ureteroceles (failure of Chwalla's membrane to apoptose in utero). Ureteroceles are often diagnosed on ultrasound of the bladder but can also be visualized on VCUG. Ureteroceles can be associated with duplicated collecting systems, either unilaterally or bilaterally. Tracking wet diapers of neonates with ureterocele is important because the ureterocele can occlude the bladder neck, obstructing the outlet. Obstructing ureteroceles should be treated promptly.

Close examination of the kidneys on RUS can show dilation of only half of the kidney's collecting system, indicating likelihood of duplication. Duplication of the collecting system can be partial, where the ureters join together before inserting into the bladder, or it can be complete, in which two separate ureters insert into the bladder or another structure (ectopic). Ectopic insertion can be a cause of obstruction or of VUR, resulting in hydroureteronephrosis in both cases. VCUG is required to rule out VUR. Ureteral ectopia can be further evaluated with MRU. In males, ureters have been observed to insert ectopically into the bladder or Wolffian structures [7, 8]. In females, ureters can insert into the vagina as well, but this does not typically present with hydronephrosis; rather, it presents with persistent diurnal incontinence. Multicystic dysplastic kidney (MCDK) can also appear like severe hydronephrosis. However, this is usually able to be differentiated from hydronephrosis if there are no connections apparent between cysts on RUS. Involution of MCDK is observed over time with RUS [10].

III Treatment

Standard treatment of severe lower pole hydroureteronephrosis during initial work up involves daily antibiotic prophylaxis until the child is proven to not have vesicoureteral reflux. Classically, the upper pole is obstructed, and the lower pole refluxes once the upper pole obstruction is corrected. The process can be relatively long to reach the point of no vesicoureteral reflux, so infants can be on daily antibiotic prophylaxis for months. With the differences in female and male anatomy, boys are less prone to UTI, especially if circumcised. Because the child was male,

circumcised, and had not yet developed a UTI, the care team elected to observe without prophylactic antibiotics.

The other consideration during work up is a cystoscopy under general anesthesia to fully understand the child's anatomy and definitively choose a course of action with the knowledge gained. However, with the accuracy of MR urography, general anesthesia and urethral manipulation could be avoided for this child, which was an important goal for the child's family. As such, observation was the best course of action for this child and his family.

IV Outcome and Follow-Up

Throughout the infant's course, he has had no UTI. He has been growing and developing normally. Eventually, his left upper pole hydronephrosis resolved. As such, since he has been asymptomatic and there is no evidence of salvageable lower pole renal parenchyma, the child's parents and the care team have elected for continued observation of the left lower pole hydroureteronephrosis. He will be monitored with serial ultrasounds to ensure normal drainage and appearance of the left upper pole moiety. Intervention may be considered if the child develops recurrent UTI, kidney stones, or progressive upper pole dilation.

Discussion

Obstruction and vesicoureteral reflux (VUR) are common presentations of a duplicated system [1, 2, 4, 9]. However, traditionally the upper pole obstructs whereas the lower pole refluxes [1-4]. This is due to the embryologic origin of this anomaly in which the upper pole ureter forms from a ureteric bud that has branched too high on the mesonephric duct, which can cause obstruction. The ureter draining the lower pole, however, is prone to reflux as it inserts more cranio-laterally into the bladder, with a shorter intramural tunnel at the ureterovesical junction [1, 2, 4, 9]. Hydronephrosis to the lower pole can occur for a myriad of reasons, including VUR, ureterovesical junction obstruction, upper pole ureterocele, ureteropelvic junction obstruction or from extrinsic compression by tumor or blood vessel [2, 3].

A case of uncrossed double ureters with lower pole dysplasia has been reported in a symptomatic older child [6]. Lower pole ureter with associated ureterocele and another case with lower pole ureteral ectopia to a vas deferens and epididymis have been previously described, though there was no evidence of associated hydronephrosis of the lower pole in either case [6-8]. Distal obstruction secondary to what appears to be an adynamic segment of the lower pole ureter with an ectopic course closer to the bladder neck is exceedingly rare, and no case series reviewed demonstrated this sequela of a duplicated system [1-9]. Interestingly, this ureter inserts laterally at the bladder neck, so not only are the ureters uncrossed, but the ectopia is unusually lateral. We report a case of uncrossed double ureters with nonfunctioning lower pole and ureteral obstruction at the level of the ureterovesical junction (UVJ), breaching the law.

Another consideration is the management approach to this child. A long-time practice of pediatric urologists is to place children with duplication and lower pole high grade hydronephrosis on prophylactic antibiotics due to the high likelihood of VUR. However, this child has had no UTIs in over a year of life and is a circumcised male, so the risk and side

effects of antibiotics are greater than the benefit for him. For the parents of the child and the care team, avoidance of daily antibiotic prophylaxis and general anesthesia for diagnostic cystoscopy was important, prompting the complete radiologic work up of the hydronephrosis.

Learning Points

- i. Prenatal hydronephrosis is a relatively common congenital anomaly that has several possible etiologies.
- ii. Complete work up of hydronephrosis is appropriate to provide the best care possible for the child.
- iii. Not all incidences of hydronephrosis require surgical intervention, and appropriate selection of imaging and clinical judgment can provide quality care while avoiding overtreatment or risks associated with anesthesia.
- iv. Completely duplicated collecting systems of the urinary tract usually follow the Weigert-Meyer rule of crossing ureters; however, all cases deserve a complete work up with the child's best interest in mind

Acknowledgements

The authors would like to acknowledge Dana Mazuru, MD, University of Missouri Department of Radiology, for her contributions in case imaging review.

Abbreviations

MCDK: Multicystic Dysplastic Kidney
MRU: Magnetic Resonance Urography
NRS: Nuclear Renal Scan
PUV: Posterior Urethral Valves
RUS: Renal Ultrasound
SFU: Society of Fetal Urology
UTI: Urinary Tract Infection
UPJ: Ureteropelvic Junction
UVJ: Ureterovesical Junction
VCUG: Voiding Cystourethrography
VUR: Vesicoureteral Reflux

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