CASE REPORT

Upper pole multicystic dysplasia and ureteropelvic junction obstruction associated with obstructive-refluxing megaureter in a neonate with a single kidney

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A 20-day-old male neonate presented with fever and hydronephrosis. Evaluation revealed that the patient had a single left kidney and a rare combination of multiple congenital malformations: upper pole segmental multicystic dysplasia, ureteropelvic junction obstruction, and an obstructive and refluxing megaureter (ureterovesical junction obstruction). We performed percutaneous drainage of the infected and obstructed upper collecting system and then used a sequential approach to manage the patient's anomalies. First, we performed an upper pole partial nephrectomy and pyeloplasty with a modified Y ureterostomy. Second, when the child was older, we performed ureterovesical reimplantation with ureteral tailoring. Currently, after 5 years of follow up, the patient has stable renal function.

Key Words: multicystic dysplasia, megaureter, ureteropelvic junction obstruction, ureterostomy

Introduction

The segmental form of multicystic dysplastic kidney is rare, and to date, only 25 cases have been reported in the literature.[1] To the best of our knowledge, only two reports of cases of ureteropelvic junction obstruction with hydronephrosis and segmental multicystic dysplasia in a kidney with a single collecting system have been published. This condition may be more common in clinical practice, but it might be underreported.

We present what we believe is the first published report of a neonate with a single kidney who had a segmental upper pole cystic dysplasia associated with obstructive-refluxing megaureter together with ureteropelvic junction obstruction. We performed an upper pole partial nephrectomy and pyeloplasty with modified Sober Y ureterostomy followed by ureteral tailoring with reimplantation. We believed this sequential approach would be more appropriate than a simultaneous approach to treat the multiple related anomalies.

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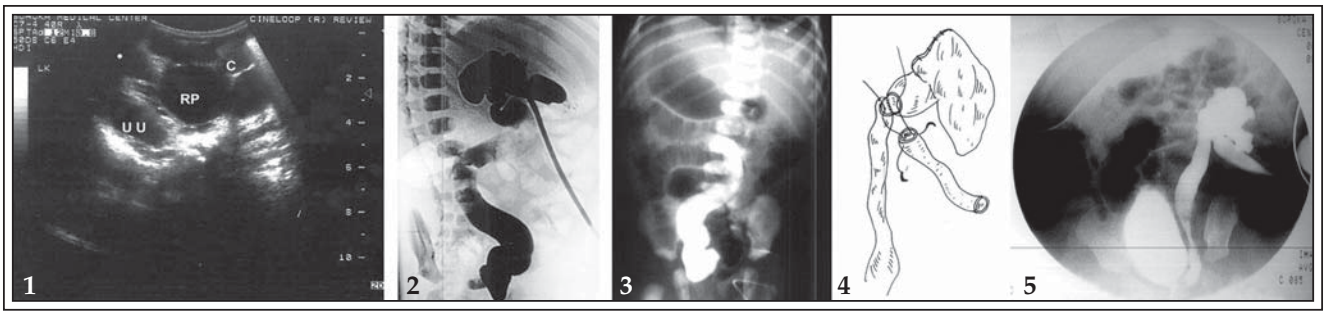
Case report

A 20-day-old male neonate presented with fever, left flank mass, and an elevated serum creatinine (3 mg/dL). Ultrasound revealed that the patient had a solitary left kidney with a dilated upper ureter, severe hydronephrosis (distension of the renal pelvis and calyces), and hydroureter. The ultrasound also revealed multiple cystic formations in the upper pole of the kidney and a dilated upper urinary tract. The thin parenchyma appeared hyperechoic, suggesting dysplasia, Figure 1.

Since the patient had oliguria, elevated serum creatinine, and fever, we performed an emergency percutaneous nephrostomy. The antegrade nephrostogram revealed what appeared to be an ureteropelvic junction obstruction as well as an obstructive megaureter (ureterovesical junction obstruction), as shown in Figure 2. The collecting system appeared to be caudally displaced. The ultrasound image also revealed that upper pole of the kidney was replaced by a multicystic mass.

The patient's clinical status improved, and a subsequent voiding cystourethrogram revealed massive reflux in a dilated ureter and a normal urethra. The upper collecting system did not fill with contrast agent during the cystography, which again suggested that



there was an obstruction at the ureteropelvic junction. In addition, there was a prolonged retention of contrast agent in the refluxing ureter, which suggested the presence of an obstruction at the ureterovesical junction, Figure 3.

Three weeks after the percutaneous nephrostomy, the patient's serum creatinine was 0.9 mg/dL. The patient had been afebrile for more than a week.

We performed an upper pole partial nephrectomy, a dismembered pyeloplasty, and a modified Y Sober ureteropyelostomy, Figure 4, due to the unresolved obstruction at the ureterovesical junction. Pathology findings from the resected ureteral specimen (obtained during pyeloplasty) were consistent with an ureteropelvic junction obstruction with inflammation, muscular hyperthrophy, and surrounding fibrosis. The resected upper pole sample had embryonic mesenchyme, immature glomeruli, and primitive ducts.

Two months later, we performed an exploration of the lower ureter as well as ureterovesical reimplantation. We obtained good functional results, Figure 5. Take down of the modified Y Sober ureterostomy limb was performed later during follow up.

Today, after almost 5 years of follow up, the patient has serum creatinine values that range from 3.5 mg/dL to 4 mg/dL. He is being followed in the pediatric nephrology and urology outpatient clinic. His bladder has normal capacity and compliance, and there has been no progression in the hydronephrosis. The patient voids with a good stream and without a postvoiding residue. Consequently, his lower urinary tract is ready for an eventual future renal transplant.

Discussion

Multicystic dysplastic kidney has a multifactorial etiology, which includes ureteral and infundibular atresia, deficient development of the blood supply, inadequate branching of the ureteral bud, and inhibition of meta nephrogenesis. Mutations of *PAX2*, *WNT-4*, growth factor genes, or prolonged obstruction may inhibit mesenchymal to epithelial cell conversion and result in persistent mesenchymal elements, i.e.

Figure 1. The ultrasound of the patient's left kidney shows a dilated upper ureter (UU), pelvis, renal cysts and rims of hyperechoic parenchyma suggesting dysplasia.

Figure 2. Left percutaneous nephrostomy. The image is consistent with ureteropelvic junction obstruction and obstructive megaureter (ureterovesical junction obstruction) with a poorly filling bladder (b). The upper collecting system is displaced caudally by multiple cysts.

Figure 3. Postvoid phase of the micturating cytography shows reflux in a severely dilated ureter. The contrast agent does not fill the renal pelvis, which suggests obstruction at the level of the ureterovesical junction (prone position).

Figure 4. The technical principle of the modified "Y" Sober temporary ureterostomy. Due to concomitant pyeloplasty, we were compelled to use a free graft from the upper ureter as one of the arms of the "Y" ureterostomy.

Figure 5. Nephrostography performed via the ureterostomy stump shows a good functional result of the pyeloplasty and the ureterovesical reimplantation.

dysplasia. Early obstruction due to atresia can also lead to dedifferentiation, a process in which epithelial cells revert into pre-epithelial mesenchymal cells. Dysplasia can also result from ureteral bud formation occurring at an ectopic site on the Wolffian duct. As a result, the bud penetrates the peripheral area of the metanephric blastema and induces a marginally dysplastic kidney.[2,3]

Congenital segmental upper pole multicystic dysplasia of the kidney is rare, which is most probably due to these events in the upper moiety of an originally duplex metanephric kidney.[4] Most probably multiple events result in ureteric and collecting system atresia, with subsequent cystic dysplasia of the remainder of the collecting system. Due to involvement of the urteric bud in this complex process, associated duplication and other Wolffian duct anomalies such as reflux and/or megaureter also occur.[5,6]

Ureteropelvic junction obstruction is the most common cause of hydronephrosis in the newborn, and

multicystic dysplastic kidney is the most common cystic abnormality seen in infants. Segmental multicystic dysplastic kidney is much less common, and the combination of both is rare.[1,6] This pathology may be more common clinically, but it may be under reported.

The association of ureteropelvic junction and ureterovesicular junction obstruction in the same kidney has been previously described.[7] Preoperative diagnosis is often difficult because the distal obstruction is masked by the proximal ureteropelvic junction obstruction. Only after the ureteropelvic junction obstruction is repaired does the ureterovesicular junction obstruction become apparent clinically or with radiography. This was not the situation in our case. The diagnoses were made simultaneously. In addition, the rare association of vesicoureteral reflux with ureterovesicular junction obstruction was also diagnosed during cystography.[8]

The association of all three of these congenital abnormalities--four, if we consider vesicoureteral reflux coexistent with ureterovesical junction obstruction--appears to be very rare, since no other case report has been published in the English literature, to the best of our knowledge.[9,10]

The embryological events that may have caused the associated complex malformations most probably occurred at two distinct points in time. Multicystic dysplasia results from an early insult, while hydronephrosis secondary to ureteropelvic junction obstruction presumably occurs later.[2] We can only assume that initially in the early embryonic stage, a duplex kidney was about to develop but the early malformative genetic event that caused the upper pole segmental dysplasia affected the corresponding future ureter as well, resulting in atresia of the ureter associated with the upper cystic dysplastic moiety—as ureteral atresia is frequently associated with multicystic dysplasic kidneys. The complex embryological insults affected the ureteral bud and metanephros, which are two developmentally interrelated embryonic structures. The metanephric blastema develops into the metanephric kidney by the penetration of the ureteral bud, with the cooptation of several and only partially elucidated gene products.[2]

The association of the multiple malformations in this patient is rare and dealing with them in a congenital renal unit was challenging. We felt that a sequentially staged approach was appropriate, since it would allow repeated reassessment of the neonate after each step, given the already present renal failure in the single kidney.[11]

The modification of the classic Y Sober ureterostomy as shown in Figure 4 was necessary due to the concomitant pyeloplasty and the unresolved obstruction at the ureterovesical junction.[12] We choose not rely only on a percutaneous nephrostomy, because of the frequently associated bacterial and fungal colonization and the displacement frequently associated with prolonged indwelling nephrostomies in neonates. To perform a technically appropriate ureterovesical reimplantation with tailoring, an appropriate waiting period was necessary between the partial nephrectomy with pyeloplasty and the planned ureterovesical reimplatation.

We were not able to perform a pyelostomy because of the reduction in size of the renal pelvis after the dismembered pyeloplasty. To preserve as much length of ureter as possible for reimplantation and tailoring, we could not perform loop ureterostomy.

Preserving as much vascularized periureteral tissue as possible, with a tension free and watertight anastomosis and a longer-than-usual ureteral limb stenting (10 days), assured a good "take" of the free ureterostomy limb.

We do not advocate regular use of this technique. However, for this patient, we felt that we had no other option, and the physiological conditions that warranted this technique were present. □

References

1. Carmack AJ, Castellan M, Perez-Brayfield M, Gosalbez R. Segmental multicystic dysplasia and ureteropelvic junction obstruction in a nonduplicated kidney. *J Pediatr Surg* 2006;41(4): E1-E3.
2. Glassberg KI. Normal and abnormal development of the kidney: a clinician's interpretation of the current knowledge. *J Urol* 2002;167(6):2339-2351.
3. Pope JC 4th, Brock JW 3rd, Adams MC, Stephens FD, Ichikama I. How they begin and how they end: classic and new theories for the development and deterioration of congenital anomalies of the kidney and urinary tract, CAKUT. *J Am Soc Nephrol* 1999; 10(9):2018-2028.
4. Diard F, Le Dosseur P, Cadier L, Calabet A, Bondonny JM. Multicystic dysplasia in the upper component of the complete duplex kidney. *Pediatr Radiol* 1984;14(5):310-313.
5. Robson WL, Rogers RC, Leung AK. Renal agenesis, multicystic dysplasia and ureteropelvic junction obstruction - a common pathogenesis? *Am J Med Genet* 1994;53(3):302.
6. Corrales JG, Elder JS. Segmental multicystic kidney and ipsilateral duplication anomalies. J Urol Vol 1996;155(4):1398-1401.
7. McGrath MA, Estroff J, Lebowitz RL. The coexistence of obstruction at the ureteropelvic and ureterovesical junctions. *AJR Am J Roentgenol* 1987;149(2):403-406.
8. Blickman JG, Lebowitz RL. The coexistence of primary megaureter and reflux. *AJR Am J Roentgenol* 1984;143(5):1053-1057.
9. Walker D, Fennell R, Garin E, Richard G. Spectrum of multicystic renal dysplasia: diagnosis and management. *Urology* 1978;11(5): 433-436
10. Jeon A, Cramer BC, Walsh E, Pushpanatha C. A spectrum of segmental multicystic renal dysplasia. *Pediatr Radiol* 1999; 29(5):309-315.
11. Bomalaski MD, Hirschl RB, Bloom DA. Vesicoureteral reflux and ureteropelvic junction obstruction: association, treatment options and outcome. *J Urol* 1997;157(3):969-974.
12. Kaneti J, Sober I. Pelvi-uretero-cutaneostomy en-Y as a temporary diversion in child. Soroka experience. *Int Urol Nephrol* 1988;20(5):471-474.