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Case Report

Duplicated Collecting System with Lower Pole Ureteropelvic Junction Obstruction

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Two examples of the rare case of complete duplicated collecting system with lower pole ureteropelvic junction obstruction are described. Ureteropelvic junction obstruction (UPJO) and duplicated collecting systems seldom occur in combination. Complete duplication of the ureter may be asymptomatic or recognized when complications develop as a result of reflux into the lower pole ureter or obstruction of the upper pole with an ectopic ureterocele. It is difficult to choose an optimal therapy due to the high variability in function, degree of obstruction, damage and potential for regeneration in growing kidneys. The diagnosis and management of UPJO of the lower pole in complete duplicated collecting systems are discussed.

Key Words: Duplicated collecting system, ureteropelvic junction obstruction

INTRODUCTION

Although ureteropelvic junction obstruction (UPJO) and duplicated collecting systems are common abnormalities in pediatric urology, they rarely occur in combination. Particularly with lower pole UPJO, not only have very few cases been reported but its exact rate of occurrence is unknown.[1,2] UPJO can occur with partial or complete duplicated collecting systems and in conjunction with other abnormalities such as ectopic ureter, ureterocele and vesicoureteral reflux. The diagnosis and optimal management of UPJO combined with duplicated collecting systems are difficult due to high anatomical variability, the degree of obstruction and chronic damage.[3] We report our experience with the management of two cases of UPJO of the lower pole in complete duplicated collecting systems.

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CASE REPORT

Case 1

The ultrasonogram of a 1-month old girl who had been prenatally diagnosed with right hydronephrosis at 24 weeks gestation showed lower pole hydronephrosis in the right kidney (Society of Fetal Urology Grade; SFUG IV) (Fig. 1A), but no evidence of dilatation of the right ureters. The physical examination and laboratory findings were normal. Voiding cystourethrography (VCUG) did not show reflux. A ^{99m}Tc-DMSA renal scan showed radioisotope uptake was normal in the upper third portion of the right kidney but markedly decreased in the lower portion (Fig. 2A). A duplicated collecting system and ureteral obstruction were suspected. Cystoscopy showed two oval shaped ureteral orifices in the right side, one in the normal site and the other in the infero-medial site. The oval shaped left ureteral orifice was in the normal site. To find the site and length of the ureteral obstruction, retrograde pyelography (RGP) was performed. In RGP, the contrast medium did not pass through the right lower UPJ but was revealed in the right upper pole (Fig. 3). RGP demonstrated a completely duplicated system combined with UPJO in the lower pole of the right kidney. The patient underwent right-lower-pole pyeloureterostomy to

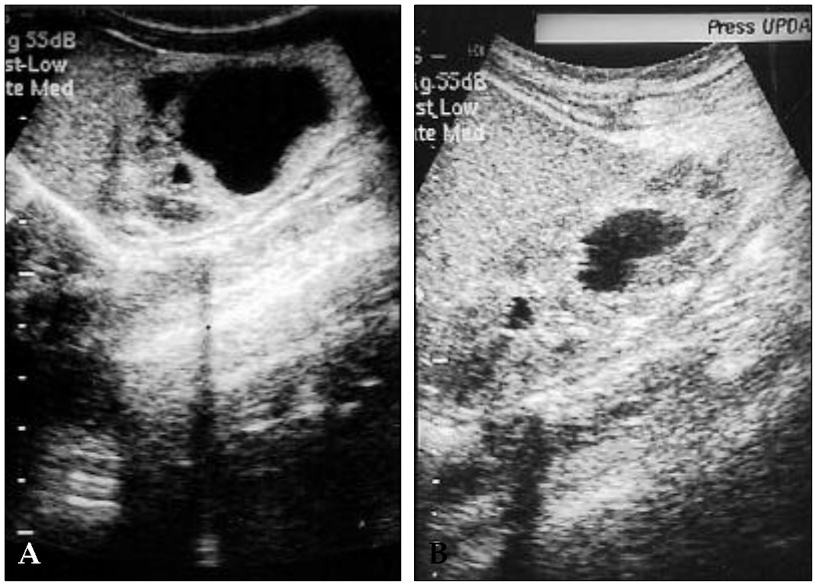

Fig. 1. (A) Preoperative ultrasonogram shows normal renal parenchyma in the upper pole and severe hydronephrosis of the lower pole. (B) Ultrasonogram performed seven months after surgery shows reduced hydronephrosis in the lower pole of the right kidney.

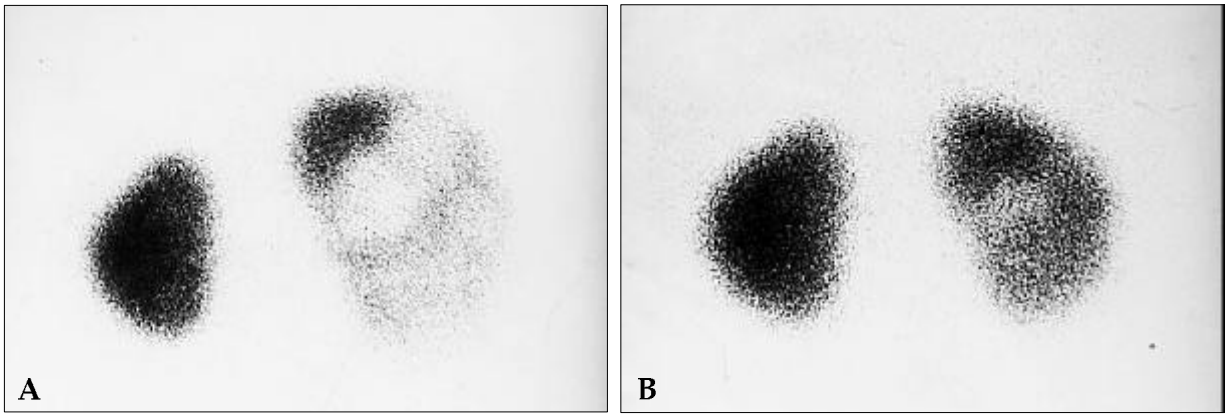

Fig. 2. (A) Preoperative ^{99m}Tc-DMSA renal scan shows decreased radioisotope uptake in the lower pole of the right kidney. (B) ^{99m}Tc-DMSA renal scan performed at postoperative seven months shows increased radioisotope uptake in the lower pole of the right kidney.

the upper pole ureter, and in surgery a narrowing portion about 2 cm long was found in the right lower ureteropelvic junction. Since the renal parenchyma was well preserved grossly and the upper ureter was normal, pyeloureterostomy was performed to connect the lower pole pelvis to the upper pole ureter. Pathological examination revealed fibrosis existing in the submucosa and muscle layer of the lower ureter.

Postoperative ultrasonography and a ^{99m}Tc-DMSA renal scan, performed at 7 months after surgery, showed significant improvement in hydronephrosis with reduced SFUG to I (Fig. 1B) and the renal scan revealed increased uptake of radioisotope to the right lower pole (Fig. 2B).

Case 2

A 40-day-old boy had been diagnosed with hydronephrosis on a prenatal ultrasonogram. Postnatal ultrasonography showed SFUG IV with normal right upper pole (Fig. 4). A ^{99m}Tc-DMSA renal scan showed decreased uptake of radioisotope in the right lower pole. VCUG showed grade III reflux into the right lower pole with poor drainage from the right lower pole. A right duplicated system with UPJO was suspected. The right upper ureter with ureterocele was not dilated. Cystoscopy with RGP showed one right ureteral orifice and a right ureterocele. RGP showed contrast medium in the upper renal pelvis

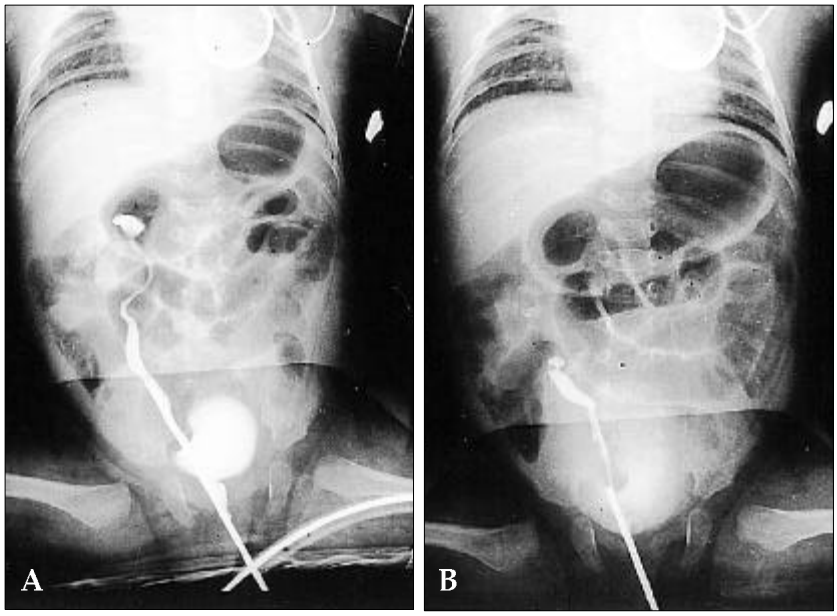

Fig. 3. Retrograde pyelogram (one month after birth): (A) Contrast material is present in the upper pole of the renal pelvis. (B) Contrast material failed to pass the ureteropelvic junction in the

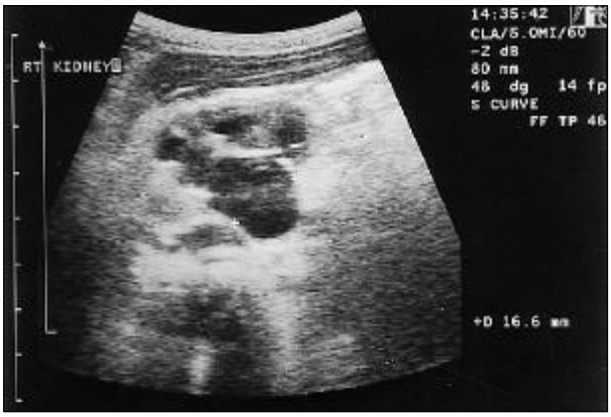

Fig. 4. Preoperative ultrasonogram shows normal renal parenchyma in the upper pole and severe hydronephrosis in the lower pole of the right kidney.

through the right ureterocele which was punctured with Nd-YAG LASER and also showed the lower UPJO through the right ureteral orifice. Antibiotic prophylaxis was administered and serial follow-up examinations were performed. VUR resolved spontaneously after three months. But a diuretic ^{99m}Tc-MAG3 renal scan demonstrated persistent right UPJO (Fig. 5A). The patient underwent right lower pole pyeloureterostomy to the upper pole ureter. During the operation, a narrowing portion was found in the right lower ureteropelvic junction. Since the upper pole renal parenchyma and ureter were well preserved grossly and the lower pole pelvis was severely dilated, dismembered pyeloplasty was performed in conjunction with pyeloureterostomy connecting the lower pelvis to the upper ureter.

A postoperative ^{99m}Tc-DMSA renal scan performed at six months after surgery demonstrated significantly increased uptake to the right lower pole and a diuretic ^{99m}Tc-MAG3 renal scan at 14 months after surgery showed resolution of UPJO (Fig. 5B).

DISCUSSION

Duplicated collecting system is the most common congenital abnormality of the upper urinary tract. In many cases, it is incidentally detected with prenatal sonogram.[4,5] The occurrence rate of duplication of the urinary tract is 0.7% and for complete duplication, it is 0.2%.[6] Although it occurs twice as commonly in females, complete duplication with UPJO of the lower pole occurs more commonly in males.[7] Other abnormalities that occur in conjunction with duplicated collecting system are renal dysplasia, ureteral obstruction, ectopic ureter and ureterocele connected with the upper pole. VUR appears in the lower pole ureter because the submucosal segment of the bladder is shortened as a result of

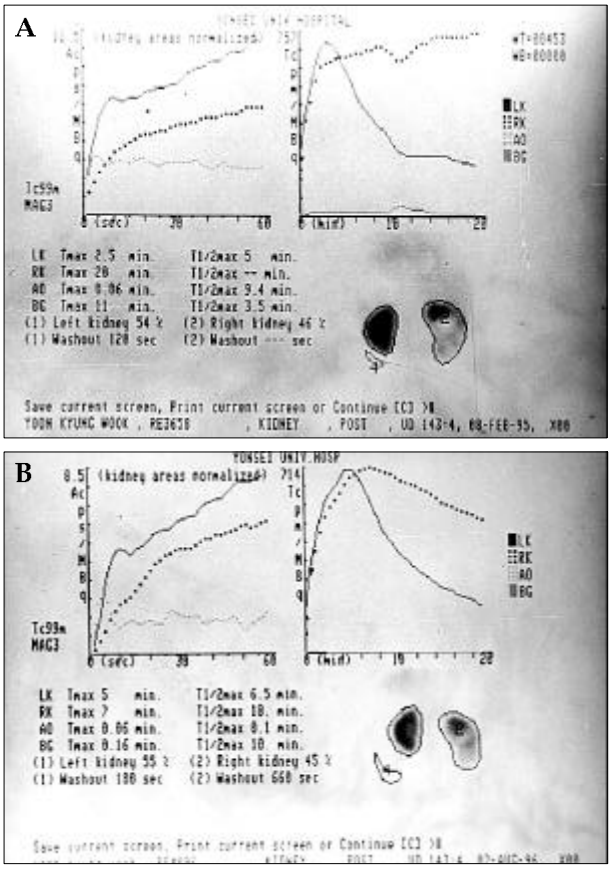

Fig. 5. (A) Preoperative ^{99m}Tc-MAG3 diuretic renogram shows obstructive pattern in the right kidney. (B) ^{99m}Tc-MAG3 diuretic renogram performed at postoperative fourteen months shows nonobstructive pattern in the right kidney.

lateral displacement of the right ureteral orifice.[2,8] The upper pole ureter does not have true UPJ but it can prevent reflux with its long distal ureter in the submucosal segment of the bladder.

Thus, even though it has been reported that hydronephrosis in the lower pole is primarily due to UPJO or VUR,[2,9] its occurrence in the upper pole kidney may be secondarily produced by the ureteral obstruction. This reflux of the lower pole ureter may play a causal role in the evolution of some cases of UPJO in a duplicated collecting system. It is postulated that ureteral dilatation of the collecting system owing to reflux may lead to elongation, tortuosity, kinking, and fibrosis of the ureter. This may be further complicated by fixation of the UPJ from periureteral inflammation leading to persistent UPJO.[2,9] If duplicated collecting system with hydronephrosis is suspected, cystoscopy and RGP are advisable for confirmation of ureteral obstruction. In addition, evaluation of renal function with a ^{99m}Tc-DMSA renal scan is essential for predicting the recovery of the postoperative renal function.[10]

In the case of simple ureter with UPJO, the indications for surgical correction at age younger than three months are calyceal clubbing associated with parenchymal thinning (parenchymal thickness less than 50% of that of ureteropelvic junction obstruction) on ultrasound and/or recurrent urinary tract infection despite chemoprophylaxis. In patients older than 3 months, surgery is performed when symptoms are presented with or without recurrent urinary tract infection, when ultrasound reveals clubbed calices with definite obstruction on diuretic renogram or when differential renal function is less than 35% in unilateral ureteropelvic junction obstruction.[3] Since no definite indications are available in the case of complete duplicated system with UPJO, we considered the above indications in our study.

The treatment of the obstruction in duplicated collecting system depends on the site of obstruction and the degree of renal function in the affected segment. In the case of complete duplicated collecting system with UPJO, the available surgical methods are pyeloplasty, pyeloureterostomy, calyceoureterostomy, end to side ureteroureterostomy and hemi-nephrectomy. In end to side ureteroureterostomy, when functional failure occurs due to ureteral peristalisis difference between the upper and lower poles, the leaked urine is not re-absorbed so infection is likely and narrowing of the suture site may occur.[5] In normal upper pole kidney, pyeloureterostomy that connects the lower renal pelvis with the upper ureter is the optimal therapeutic choice. In pyeloureterostomy, the drainage of urine is effective, the problems associated with the reflux that may occur in end to side ureteroureterostomy are minimized, and the ureteral stump does not become the source of infection.[9] In addition, the surgical outcome of grossly normal upper pole ureter without dilation represents a good prognosis. When the lower pole kidney has reduced renal function, dismembered pyeloplasty may be considered, while when one pole in a complete

duplicated collecting system does not properly function, hemi-nephrectomy may be the most appropriate option.

In the two cases presented here, preoperative tests demonstrated an intact upper pole and a defective lower pole. Having predicted the presence of renal parenchyma in the lower pole kidney, we performed pyeloureterostomy to connect the lower pole renal pelvis with UPJO to the upper ureter. The abdominal ultrasonogram and ^{99m}TC- DMSA renal scan at post operative six months revealed significantly improved renal function of the lower pole with no recognized complications such as fever or urinary tract infection.

We believe that careful preoperative evaluation of the duplicated collecting system with UPJO is crucial. ^{99m}Tc-DMSA renal scan, VCUG, abdominal ultrasonogram, and RGP all aid in the proper diagnosis and treatment, allowing the best surgical approach to be planned. We also believe that in the case where the upper pole functions properly and is combined with UPJO in the lower pole, pyeloureterostomy connecting the lower pole renal pelvis to the upper pole ureter is the one of the optimal surgical modalities in treating complete duplication with UPJO of the lower pole ureter.

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