

## GIANT HYDRONEPHROSIS OF BILATERAL DUPLEX SYSTEMS ASSOCIATED WITH URETERAL ECTOPIA: A CASE REPORT

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Giant hydronephrosis is an uncommon clinical entity. Even more uncommon is the association of giant hydronephrosis with a double collecting system and ectopic ureter. Here, we report a case of giant hydronephrosis of the bilateral duplex systems associated with ureteral ectopia. The patient underwent upper pole nephrectomy and upper ureterectomy. To our knowledge, only four similar cases have been reported previously.

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**Key words:** Giant hydronephrosis, Duplex system, Ureteral ectopia

### INTRODUCTION

Giant hydronephrosis is defined as a hydro-nephrotic kidney containing more than one liter of fluid<sup>1)</sup> The condition is currently rare due to improved diagnostic methods and medical care, although it is relatively asymptomatic. The majority of cases are caused by obstruction at the ureteropelvic junction<sup>2)</sup>

An ectopic ureter is more common in females than in males and is often associated with a double collecting system. In contrast to male patients in whom urological symptoms such as dribbling incontinence and enuresis is rare, female patients with ureteral ectopia are seldom overlooked or misdiagnosed.

Recently, we encountered a 33-year-old woman with giant hydronephrosis of the right upper kidney caused by obstruction at the ureteropelvic junction associated with bilateral duplex systems and right ureteral ectopia who had a history of occasional dribbling incontinence in her childhood.

### CASE REPORT

A 33-year-old woman was hospitalized in June 1992 for examination of abdominal distension and persistent dull pain in the flank which had become slowly but unremittingly progressive after lumbar contusion in 1986. Past history included occasional dribbling incontinence in her childhood, although she had also maintained a normal voiding pattern in childhood. Physical examination showed a well-developed Japanese woman with a slightly distended abdomen which was soft and nontender to palpation. Laboratory data including complete blood count, blood chemistry studies and urinalysis were normal. Abdominal ultrasonography (US), drip infusion pyelography (DIP) plus an antegrade pyelography (AP) of the ectasized right upper renal pelvis (Fig. 1),

computed tomography (CT), and magnetic resonance imaging (MRI) revealed giant hydronephrosis of the right upper kidney associated with bilateral duplicated collecting systems. Cystoscopy demonstrated two left ureteral orifices and a right ureteral orifice in the trigone.

Eight days after admission, the patient under-



Fig. 1. DIP plus AP of the ectasized right upper renal pelvis shows giant hydronephrosis in the right upper kidney associated with bilateral duplicated collecting systems.



Fig. 2. CT revealed a non-enhancing giant hydronephrotic sac of right upper kidney origin.

went exploration through a right lumbar incision. Adjacent to a slightly compressed right lower kidney with its own normal ureter, there was a thin-walled hydronephrotic sac of right upper kidney origin. The sac showed obvious obstruction at the ureteropelvic junction and an almost normal-looking ureter was

recognized. The ureter of the hydronephrotic sac was dissected and it ended at the side of the vaginal wall. At the distal end near the vagina, the ureter showed slight cystic distension. These structures were extirpated en bloc. The sac contained 2,640 ml of fluid. Convalescence was uneventful. The patient was discharged from hospital 15 days after surgery. DIP on one-year follow-up revealed good function of lower pole moiety in the right kidney and in the left duplex system.

### DISCUSSION

Giant hydronephrosis is defined as a gigantic tumor in which the hydronephrotic sac contains more than one liter of fluid<sup>1)</sup>. Most cases are of congenital origin<sup>3)</sup> and are mainly caused by obstruction at the ureteropelvic junction or occasionally by duplication with or without ureteral ectopia<sup>2)</sup>. When refined imaging techniques such as CT or MRI were not available, correct preoperative diagnosis was made in less than 50% of the cases. The others were often misdiagnosed as ascites<sup>2,3)</sup>.

Of all ureteral ectopia, 80% are associated with double collecting systems and of these, 75% occur in

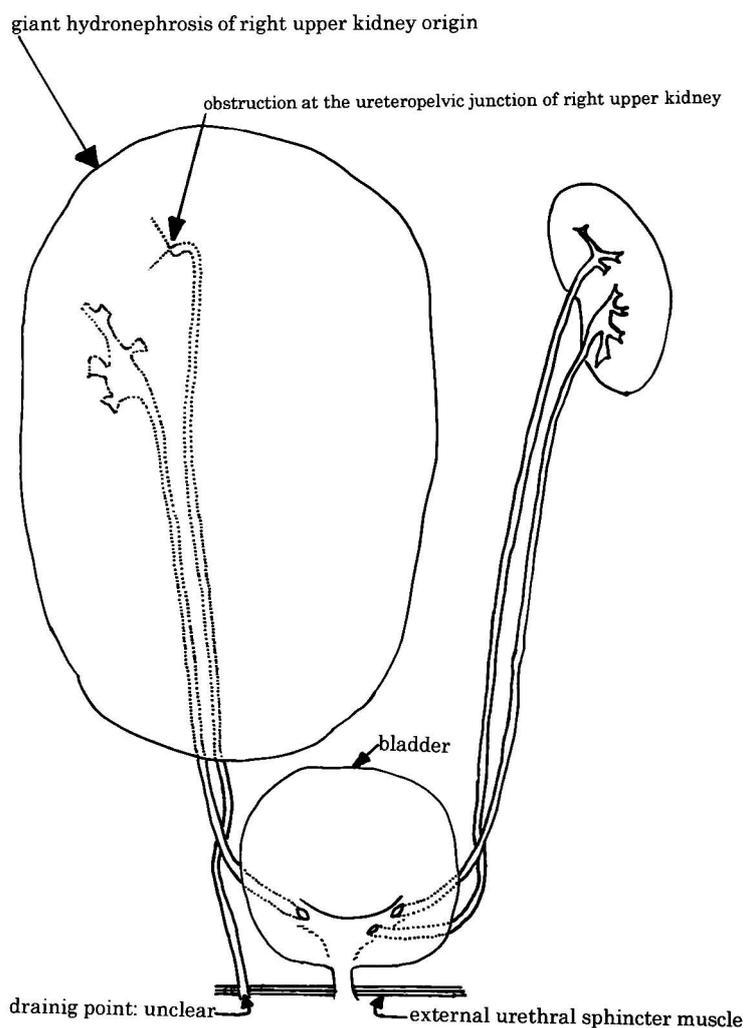


Fig. 3. Schematic presentation of preoperative anatomy.

Table 1. Giant hydronephrosis accompanied by duplex system(s) associated with ureteral ectopia.

Age/sex	Giant hydronephrosis		Duplex system	Ectopic ureter		Diagnostic procedure	Treatment	Follow-up	Reference
	side	fluid amount (ml)	side	side	drain to				
20/F	L, upper L, lower	>1,000 >2,700	L	L, lower	vagina	DIP, RP, AG, cystoscopy	L, upper and lower NUx	—	6
46/M	L, upper	2,160	L	L, upper	prosterior urethra	—	—	—	7
23/F	R, upper	5,300	Bil	R, upper	vagina	DIP, US, CT, AP, cystoscopy	R, upper pole NUx	22D, well	8
39/M	L, upper	10,000	L	L, upper	prostatic urethra	IVP, CT, RP, AG, cystoscopy	L, upper pole Nx	2Y, well	2
33/F	R, upper	2,640	Bil	R, upper	unclear	DIP, AP, US, CT, MRI, cystoscopy	R, upper pole NUx	1Y, well	present case

L, left; R, right; Bil, bilateral; DIP, drip infusion pyelography; RP, retrograde pyelography; AG, angiography; AP, antegrade pyelography; IVP, intravenous pyelography; US, ultrasonography; CT, computed tomography; MRI, magnetic resonance imaging; NUx, nephroureterectomy; Nx, nephrectomy; D, day; Y, year

women<sup>4)</sup>. In order of decreasing frequency, extra-vesical ectopic ureter may drain into the urethra, vestibule, vagina or uterus. Female patients with ectopic ureter may present with complaints of dribbling incontinence<sup>5)</sup>

In this case, the giant hydronephrosis of right upper kidney origin was probably caused by obstruction at the ureteropelvic junction because apparent obstruction was recognized on surgery. The ectopic ureter may have been passing in or by the external urethral sphincter muscle, and therefore the continence mechanism was preserved with a few incontinence episodes. Although the draining point of the ectopic ureter was unclear, at least it would have been distal to the external urethral sphincter muscle.

To our knowledge, only four similar cases of giant hydronephrosis accompanied by duplex system(s) associated with ureteral ectopia have been reported previously<sup>2,6-8)</sup> as shown in Table 1. Patients with distended abdomen may also require urological consultation.

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## 和文抄録

## 重複腎盂尿管と尿管異所開口を伴った巨大水腎症の1例

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症例は33歳女性。腹部膨隆を主訴として当科初診。腹部超音波検査, DIP と順行性腎盂造影, CT, MRI, 膀胱鏡検査等にて, 両側重複腎盂尿管の右上半腎より発生した巨大水腎症と術前診断された。腰部斜切開にて後腹膜腔に入ると右上半腎所属の尿管は, 明らかな腎盂尿管移行部狭窄を伴っており, これにより巨大水腎症が発生したと思われた。また幼少時に時折自覚していた dribbling incontinence の失禁量が多くないことを考え合わせると, 右上半腎所属の異所性

尿管は外尿道括約筋群の中かあるいは近傍を走行し, 失禁防止機構が働いていたことが推測された。尿管異所開口の部位は断定できないが, すくなくとも外尿道括約筋群よりも遠位であると考えられた。巨大水腎とその所属尿管は一塊として摘出され, 巨大水腎の容量は, 2,640 ml であった。術後1年目のDIPにて良好な経過を取っている。

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