Single system ectopic ureter in females: A single center study

Department of Pediatric Surgery, IMS, BHU, Varanasi, India

Correspondence: Vijay D. Upadhyaya, Department of Pediatric Surgery, IMS, BHU, Varanasi - 221 005, UP, India. E-mail: upadhyayavj@rediffmail.com

ABSTRACT

The purpose of this study was to inquire into the clinical features and methods for the diagnosis and management of single-system ectopic ureters associated with renal dysplasia. Materials and Methods: A total of 13 female patients were studied. Mainstay of diagnosis was ultrasonography of KUB region and intravenous urography and renal scan was used to confirm the diagnosis. Histopathological evaluation was done in all cases for documentation of renal dysplasia. Result: In eight cases ectopic ureter with dysplastic kidney was seen on left side and in five it was on right side. All the patients were treated with nephroureterectomy of the affected side because of poor functioning of ipsilateral dysplastic kidney. Conclusion: Continuous urinary incontinence in females with a normal voiding pattern should prompt an evaluation for ureteric ectopia and when initial evaluation yields diagnosis of solitary kidney the clinician should be aware of the possibility of a hypoplastic and/or dysplastic on one side and normally functioning kidney on opposite side. Nephroureterectomy is the treatment of choice for unilateral single system ectopic ureter with renal dysplasia of affected side.

KEY WORDS: Dysplastic kidney, females, nephroureterectomy, single system ectopic ureter

INTRODUCTION

An ectopic ureter is defined as an ectopic ureteric orifice outside the posterolateral extremity of the bladder trigone.[1] An ectopic ureter that inserts either into the urethra distal to the sphincter or into the vagina in a girl typically presents with continuous wetting despite an otherwise normal micturition pattern. In most cases, the ectopic ureter arises from the upper moiety of a duplex kidney and detecting such an ectopic system is relatively straightforward. However, in a small proportion of patients the ectopic ureter may arise from a small, dysplastic and poorly functioning kidney. In general, 80% of ectopic ureters arise from the upper pole of a completely duplicated system. Ectopic ureters draining single systems are rare[2,3] occurring only in 20% cases.[4] Successful removal of the dysplastic kidney with its draining ureter results in immediate and complete cure of the incontinence problem. However, the small nonfunctioning kidney, which can often be ectopic, may defy detection even with repeated examinations using conventional imaging modalities and cystourethrovaginoscopy under anaesthesia, thus leading to a significant delay in the correct diagnosis and appropriate treatment. Though this moiety is more common in male, but contrary to the literature we have encountered all 13 cases in female patients during this period. We retrospectively analyzed the clinical features and methods for the diagnosis and management of single-system ectopic ureters associated with renal dysplasia.

MATERIALS AND METHODS

The study was carried out at the university hospital BHU, Varanasi. A total of 13 females of single system ectopic ureter associated with dysplastic kidney were admitted from 1995 to 2005. In a retrospective review, the case notes, including presentations, findings on examination, imaging studies, operative details, pathology findings and postoperative outcomes, were studied. All patients were subjected to intravenous urography (IVU), ultrasound and renal scan. Prior to exploration, cystourethrovaginoscopy was done in all patients after filling the bladder with methylene blue dye either via per urethral route or by suprapubic approach under general anesthesia and exploration was performed in all 13 patients [Table 1] and renal dysplasia was further confirmed by pathologic examination.
RESULTS

All the patients were girls, with a median age of 2.5 years (range four months to three years). Eight cases occurred on the left and five on the right side. The patients had developed and grown normally, presented with wetting as well as normal voiding. They had the same volume of dribbling during the daytime as well as in night, without any relation to position. Two patients had history of abdominal pain. None of them presented with features of urinary tract infection. Methylene blue test was positive in 10 cases.

All the patients were examined by ultrasonography, intravenous urography and renal scan [Table 1]. Intravenous urography showed very poor function of affected kidney in nine cases where as in four cases it was non conclusive however hypertrophy of contralateral kidney was observed in 30.7% of cases only. Renal scan was done in all cases for confirmation as well as for decision making. In renal scan, affected kidney was visualized in 12 cases (differential function was ranging form 1-7% in affected kidney) and in one case affected kidney was not visualized. Perineal examination including vaginocystoscopy revealed ectopic opening in vagina in five cases, in four cases the ectopic opening was present in vestibule whereas in one case, the opening was present in urethra and was not visualized in three cases. However methylene blue test showed clear fluid in perineum which indirectly confirms that ectopic opening is outside the sphincter.

All patients underwent nephroureterectomy of the affected side. In five cases the affected kidney was present at its normal position and in eight cases it was present in pelvis. The size of affected kidney in all cases was very small ranging from $3 \times 2 \times 1 \text{ cm}$ to $4.2 \times 2.6 \times 1.5 \text{ cm}$ [Table 1]. During operation we were not able to identify renal vein and artery separately and mass ligature was done in nine cases where as in four cases both of them were ligated individually.

Dribbling and wetting disappeared right after operation. All patients recovered well and were discharged on the fifth to seventh postoperative day. At follow-up after one to seven years, they had developed normally. The pathology examination reported renal dysplasia.

DISCUSSION

Urinary incontinence is a common symptom in children; under most circumstances it is caused by functional disturbances of the detrusor-sphincter complex, the treatment outcome of which is often unpredictable. Wetting from organic causes, e.g. infrasphincteric ureteric ectopia, is much less common but is particularly important because of the potential for immediate cure by surgical intervention. A typical history of continuous or intermittent dribbling of urine despite an otherwise normal pattern of voiding, often starting in early childhood, should prompt a clinical suspicion of an ectopic ureter. The diagnosis can be further affirmed by a simple wet-pad test for dribbling of clear urine after the introduction of methylene blue into the bladder.

As per the literature single-system ectopic ureter are more common in male. We are presenting a series of 13 cases in females and contrary to literature we have not encountered any male patient during this period. Such ectopic ureters are frequently found in association with VACTERL syndrome though we observed this association in only 15.45% of cases. A small, dysplastic, poorly-functioning single kidney associated with an ectopic ureter may be difficult to localize, leading to delay in diagnosis or even to misdiagnosis as a solitary kidney. In the present series most of affected kidneys were localized by renal scan which also help in decision making of the management.

Table 1: Clinical and pathologic characters of single-system ureters associated with renal dysplasia

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age</th>
<th>Kidney localized by</th>
<th>Site of kidney</th>
<th>Urt. Opn.</th>
<th>Pathology</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1.5 yr</td>
<td>US, RS</td>
<td>E</td>
<td>V</td>
<td>MG</td>
</tr>
<tr>
<td>2</td>
<td>2 yr</td>
<td>US, IVU, RS</td>
<td>E</td>
<td>VS</td>
<td>IMG</td>
</tr>
<tr>
<td>3</td>
<td>3 yr</td>
<td>IVU, RS</td>
<td>N</td>
<td>VS</td>
<td>IMG</td>
</tr>
<tr>
<td>4</td>
<td>3 yr</td>
<td>IVU, RS</td>
<td>E</td>
<td>U</td>
<td>IMG</td>
</tr>
<tr>
<td>5</td>
<td>3 yr</td>
<td>IVU, RS</td>
<td>N</td>
<td>NL</td>
<td>IMG</td>
</tr>
<tr>
<td>6</td>
<td>6 month</td>
<td>US, RS</td>
<td>E</td>
<td>NL</td>
<td>MG</td>
</tr>
<tr>
<td>7</td>
<td>9 month</td>
<td>US, RS</td>
<td>E</td>
<td>V</td>
<td>MG</td>
</tr>
<tr>
<td>8</td>
<td>2 yr</td>
<td>IVU, RS</td>
<td>N</td>
<td>VS</td>
<td>IMG</td>
</tr>
<tr>
<td>9</td>
<td>3yr</td>
<td>IVU, RS</td>
<td>E</td>
<td>V</td>
<td>IMG</td>
</tr>
<tr>
<td>10</td>
<td>4 month</td>
<td>INTRA-OP</td>
<td>E</td>
<td>NL</td>
<td>MG</td>
</tr>
<tr>
<td>11</td>
<td>1yr</td>
<td>US, RS</td>
<td>N</td>
<td>VS</td>
<td>IMG</td>
</tr>
<tr>
<td>12</td>
<td>2 yr</td>
<td>IVU, RS</td>
<td>E</td>
<td>V</td>
<td>IMG</td>
</tr>
<tr>
<td>13</td>
<td>1.5 yr</td>
<td>US, RS</td>
<td>N</td>
<td>V</td>
<td>MG</td>
</tr>
</tbody>
</table>

US - Ultra sound, IVU - Intra venous urography, RS - Renal scan, E - Ectopic position, N - Normal position, V - Vagina, VS - Vestibule, U - Urethra, NL - Not localized, MG - Mature glomeruli and tubules, reduced in quantity, IMG - Immature glomeruli and tubules surrounded by connective tissue
The etiology of renal dysplasia is not yet quite clear. The presumed embryologic mechanism is that the ureteric bud arises more cranially than usual from the mesonephric duct, so that it is taken into the bladder wall at a lower level during the seventh week of gestation or does not join the bladder wall at all. Maizels and Stephens postulated that abnormal growth of the spine precludes normal renal ascent and may even be responsible for renal agenesis and hypoplasia. The ostium localizes along a defined pathway from the bladder neck to the seminal vesicles in males where as it opens into the mesonephric duct remnants in females. This abnormal location of the ureteric bud probably leads to abnormal interaction with nephrogenic blastema and renal dysplasia. Therefore, an ectopic ureter is frequently associated with ectopia of the corresponding kidney and a variable degree of renal dysplasia. It is also postulated that failure of vascular supply during embryologic development prevents the kidney from developing normally and causes formation of a small primary organ that contains embryonal tissues. Some researchers believe that it is the obstruction of the ureter during an early phase of the embryo that stops development of the kidney. All 13 cases had renal dysplasia, in five cases kidney was located at normal site whereas in eight cases it was at lower location, ureter was narrow, non fibrosed and non obstructive, except in one case where the ureter was dilated and tortuous.

Depending on the location of the ostium, ectopic ureters may produce a highly variable clinical character which delays the diagnosis and therapeutic result. An ectopic opening into the bladder neck may produce obstructive uropathy, vesicoureteral reflux, recurrent infections or enuresis ureterica; that is, urinary dribbling associated with an otherwise normal voiding pattern results from ectopia distal to the bladder sphincter in female patients.

Highlights of clinical and pathologic features in our series are: A) Our series consists of all girls, similar to reports by Chowdhary et al. and Li et al. though others had reported that single-system ureteral ectopia is more commonly in males. B) All the patients were under the age of three years of which two were infants. C) In 38% of cases, kidney was located at normal position. Was not dilated, distorted or fibrosed in 12 cases. D) Strong association with ipsilateral renal dysplasia noted by others was confirmed in our series. E) The ureter orifice was ectopic but the ureter was not dilated or distorted, not fibrosed in 12 cases, which is different from the dilated and distorted ectopic ureter seen with duplex system. F) We used IVU primarily for locating the kidney as well as for contralateral functional hypertrophy and renal scan to confirm the diagnosis which also helps in decision making of the management, where as other have used computed tomography for the same. G) Except for mild wetting and dribbling, these patients had no other symptoms and the severity of wetting and dribbling was far milder than that of patients with duplicated kidney (indirect evidence of dysplastic kidney) and an ectopic ureter.

Intravenous urography and ultrasonography are the most common examinations for diagnosis of ectopic ureter. In the present series in six cases, affected kidney was localized by ultrasonography whereas Borer et al. were not able to detect single ectopic kidney in their series. Though Li et al. were able to locate the kidney in seven out of eight of the cases. In present series in 61% cases the affected kidney was located by IVU which is contrary to the report of Li et al. Kawamura N et al. used renal scan where IVU was not able to detect the ectopic kidney in the cases of poor functioning single ectopic ureter but in our series all of these patients were examined by renal scan which also helped us in decision making. Recently laparoscopy is used to localize the position of ectopic kidney and was able to detect the position of ectopic kidney in all cases.

Treatment can be in form of ureteral reimplantation when relative renal function is 10% or more or nephroureterectomy when relative renal function is less than 10%. In patients with bilateral anomalies or marginal renal function, reimplantation is of choice for salvaging renal mass. In our series, excision was a simple and valid treatment. Before ureteronephrectomy, it is better to fill the bladder with methylene blue saline either via a thin needle or perurethrally and observe whether blue liquid comes from the perineum so as to confirm the diagnosis. Because the dysplastic kidney is usually located abnormally in the retroperitoneum below the normal kidney location, care is needed not to misdiagnose it as a solitary kidney by exploring only the normal kidney location. Its merits include minor trauma, less time spent, rapid recovery, short hospitalization and low cost and it may be especially suitable when the precise location of the dysplastic kidney cannot be predicted preoperatively.

Continuous urinary incontinence in females with a normal voiding pattern should prompt an evaluation for ureteric ectopia. When the initial evaluation yields the diagnosis of a solitary kidney, clinicians should be aware of the possibility of a hypoplastic and/or dysplastic, often ectopic, contralateral kidney with an
ectopically draining ureter. Nephroureterectomy is the treatment of choice for unilateral single system ectopic ureter with renal dysplasia of affected side.

REFERENCES


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