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Extensive chondroid differentiation in a Wilms tumor following chemotherapy: Clinical relevance and implications

Kalpana Kumari1, Priyanka Naranje2, Tripti Nakra1, Seema Kaushal1, Diya Roy1, Saket Davera3, Sandeep Agarwala3, Ahitagni Biswas4, Venkateswaran K. Iyer1, Amit Kumar Dinda1

1Department of Pathology, All India Institute of Medical Sciences, New Delhi, India
2Department of Radiodiagnosis, All India Institute of Medical Sciences, New Delhi, India
3Department of Pediatric Surgery, All India Institute of Medical Sciences, New Delhi, India
4Department of Radiotherapy and Oncology, All India Institute of Medical Sciences, New Delhi, India

ABSTRACT

Wilms tumor (WT) is the most common childhood tumor of the kidney, which histologically mimics various stages of nephrogenesis. Spectrum of chemotherapy induced histopathological changes has been described in literature, and is one of the most important predictors of disease outcome and survival rates. Extensive chondroid differentiation and absence of necrosis in a post-chemotherapy nephrectomy specimen of a radiologically proven Wilms tumor is an unusual finding. Herein, we present the case of a 6-year-old girl from South Africa who received a 10-week course of chemotherapy upon radiological diagnosis of WT, and post-surgery nephrectomy specimen revealed extensive mature cartilage island formation on histology raising diagnostic dilemma. Reporting of such cases is extremely important to recognize tumor morphological heterogeneity, particularly post-chemotherapy, and developing consensus for selecting further treatment and clinical follow-up.

Key Words: Wilms tumor, chondroid, post-chemotherapy, stromal.

Introduction

Wilms tumor (WT) is the most common childhood tumor of the kidney, constituting 6 to 7% of childhood cancer in developed countries, [1] and accounts for approximately 12% of all childhood cancers in the underdeveloped countries [2]. WT mimics normal nephrogenesis, and histologically is composed of a variable admixture of epithelial, blastemal and stromal components [3]. Heterologous components i.e. striated and smooth muscle, cartilage, bone, adipose tissue, etc. are present in only 10% of WT [4]. Recently, studies have shown that two-hit inactivation of the WT1 gene is more prevalent in stromal-predominant WT [4]. Currently, pre-chemotherapy/ up-front surgery-based system developed by the National Wilms Tumor Study Group (NWTSG) and post-chemotherapy- based system developed by the
International Society of Pediatric Oncology (SIOP) are the treatment strategies selected based on individual patient risk of tumor spillage, rupture and recurrence [5]. Patients following SIOP protocol usually do not undergo a biopsy prior to starting therapy [5]. Preoperative chemotherapy is known to affect original tumor histology, sometimes drastically, resulting in reduced or enhanced individual tumor components, and often inducing maturation [6,7]. WT has good prognosis with survival rate of 90% in developed countries following either regime [1,2]. Tumor stage and histopathological features following neoadjuvant chemotherapy as well as after upfront surgery are the most important predictors of disease outcome and survival rates [5]. Herein, we present a case of 6-year-old girl from South Africa who received 10 weeks course of chemotherapy upon radiological diagnosis of WT, and post-surgery nephrectomy specimen revealed extensive mature cartilage island formation on histology raising diagnostic dilemma. Reporting of such cases is extremely important to recognize tumor morphological heterogeneity and assess response following chemotherapy. Indeed, requires developing consensus for selecting further treatment course for such cases.

**Case report**

A 6-year-old girl from South Africa presented to the pediatric surgery out-patient department with a complaint of swelling in the right flank. The swelling was first noticed by her parents 8 months ago, when the size was that of a lemon. However, they found a rapid increase in the size of the swelling. History of fever, pain abdomen, hematuria, and hypertension was absent. Past medical history was non-contributory. Contrast enhanced CT revealed a large heterogeneously enhancing mass measuring 17 X 16 X 11 cm with well-defined margins arising from lower pole of the right kidney. The mass extended from T11 to S1 vertebral body, crossing the midline. Right renal vein and inferior vena cava (IVC) were free of thrombus (Fig. 1; A, B & C). A radiological diagnosis of Wilms tumor was suggested. Pre-operative fine needle aspiration cytology (FNAC) was not done. With this diagnosis patient was started on chemotherapy. DD4A regimen (vincristine, dactinomycin, and doxorubicin) was administered for 10 weeks. Post-chemotherapy CECT revealed reduction in size of the tumor to 15X9 cm with change in the internal attenuation and enhancement pattern (Fig. 1; D, E & F). Right nephrectomy and hilar lymph node dissection was undertaken.

On histopathological examination, the right nephrectomy specimen measured 20X13X10 cm, weighing 1325 grams (Fig. 2). Attached ureter measured 10.5 cm in length and 0.6 cm diameter. External surface of the kidney was bosselated with a thick, adherent capsule. Serial slicing showed a large well encapsulated mass replacing almost the entire renal parenchyma, with a thin rim of normal renal tissue at the lower pole. Cut surface of the tumor was lobulated, firm, pearly white to myxoid, with intervening pin-point yellowish areas. Focal fleshy areas were also identified. Microscopic examination from different tumor areas predominantly showed islands of mature cartilage lacking nuclear atypia (approximately 80% of tumor area), surrounded by nondescript mesenchyme with focal myxoid stroma (approximately 15%). At places, skeletal muscle differentiation was noted (approximately 5%), along with areas of hyalinization. Epithelial, blastemal or malignant stromal component were not seen.
Necrosis was present in less than 1% of the total area sampled. MIB-1 labeling index was negligible in the chondroid islands (Fig. 3). Differential diagnoses considered were stromal-predominant WT with heterologous elements, ossifying renal tumor of infancy and intrarenal teratoma. Immunohistochemistry (Fig. 4a, b) for WTI showed cytoplasmic and nuclear staining in the skeletal muscle component and helped to confirm the diagnosis. Non-descript stroma showed negative staining for smooth muscle actin, desmin, myogenin and S100. Based on the post-chemotherapy induced histological changes in form of predominance of chondroid elements and WT1 staining in the skeletal muscle component, a final diagnosis of stromal-predominant WT with extensive chondroid differentiation was made. Thereafter, patient received radiotherapy to the flank 10.8 Gy/6 cycle for six days and adjuvant chemotherapy planned upto 24 weeks as per DD4A regimen.

**Discussion**

Histologically, WT mimics various stages of nephrogenesis [4]. Depending upon the components present, it can be mesenchymal predominant, epithelial or mixed. WT containing heterologous element are considered to arise from intralobar nephrogenic rests (ILNRs) rich in stroma [4]. Fetal rhabdomyomatous WT containing abundant skeletal muscle are chemoresistant and often shows poor volumetric response has been described in literature [7,8]. Presence of extensive cartilaginous differentiation along with foci of rhabdomyomatous differentiation, and the complete absence of necrosis, tubules, blastema and epithelial component in the index
Fig. 2. Gross photomicrograph: Nephrectomy specimen depicting large well circumscribed tumor with small portion of normal renal tissue at the upper pole. Cut section is lobulated, firm, pearly white to myxoid, with intervening pin-point yellowish areas.

Fig. 3. Photomicrograph depicting lobules of cartilage islands separated by well demarcated areas showing skeletal muscle differentiation (A, H&E, x100). Extensive chondroid islands with intervening stromal component. Necrosis absent (B, x100). Cartilage islands displaying benign appearing chondrocytes (C, x200). Non-descript stroma lacking atypia (D, x200).
case is an unusual type of tumor response to chemotherapy in a radiologically proven WT. In routine practice, the diagnosis of Wilms tumor relies on classical morphological features supplemented by nuclear staining for WT1 on immunohistochemistry [9]. WT1 shows strong nuclear expression in majority of cases, particularly in epithelial and blastemal components, while it may be extremely low or even absent in stromal components [4]. WT1 gene mutation is present in approximately 15 to 20% of WT [9]. However, recently, germline mutations have also been described in the majority of stromal-predominant WTs, leading to ectopic myogenesis [4]. In such a setting, the gene product of WT1 is aberrantly expressed in the cytoplasm of tumor cells, indicating arrest of mesenchymal to epithelial lineage transformation, and this can be detected by immunohistochemistry using an antibody against the N-terminus of WT1 [4]. Cytoplasmic WT1 expression in the skeletal muscle component in the present case thus supported the diagnosis of WT, although only focal nuclear staining was seen.

Studies have shown that chemotherapeutic agents used in treatment of WT induce necrosis of immature and actively proliferating cells i.e. the blastemal component, and cell maturation in other components [6,9,10]. The differentiated component appears histologically benign with negligible proliferating index [10]. However, rate at which the therapeutic agents induce differentiation process i.e. rhabdomyomatous and chondromatous is still unknown [7,10]. In the present case, extensive cartilaginous differentiation presumably represents a maturation response following chemotherapy, or survival of well differentiated component in the original tumor population. The lack of evidence of chondroid differentiation on preoperative imaging favors the former over the latter. Absence of necrosis after chemotherapy is also suggestive of a stromal predominant WT lacking blastemal or other rapidly proliferating components. Reduction in tumor volume after chemotherapy is a measure of clinical response for determining postoperative treatment. Stromal predominant WTs are known not to reveal volume reduction post-chemotherapy [6,10]. In the index case, there was only slight reduction in tumor volume, suggesting poor volumetric response and need for further therapeutic intervention, and further supporting our hypothesis that extensive cartilaginous differentiation in the present case occurred as a maturation of stromal component following chemotherapy. However, studies have supported that poor
volumetric response may not represent treatment failure or aggressive tumor behavior [10,11].

**Conclusion**

Extensive presence of heterologous elements may confound the histological diagnosis in absence of classical triphasic appearance of WT. Volumetric response is not the only measure of response to chemotherapy; maturation and terminal differentiation to benign stromal derivatives without volume reduction can also be considered a response to chemotherapy, limiting its metastatic potential.

**Compliance with ethical statements**

*Conflicts of Interest: None.*

*Financial disclosure: None.*

*Consent: All photos were taken with parental consent.*

**ORCID ID of the authors**

Kalpana Kumari /0000-0002-8396-6809
Priyanka Naranje /0000-0002-3147-8643
Tripti Nakra /0000-0001-9907-7531
Seema Kaushal /0000-0002-6190-5909
Diya Roy /0000-0002-0175-7641
Saket Davera /0000-0002-6211-2134
Sandeep Agarwal /0000-0002-8345-6678
Ahitagni Biswas /0000-0002-3453-5340
Venkateswaran K. Iyer /0000-0003-1933-7562
Amit Kumar Dinda /0000-0002-5840-1830

**References**

Loop ureterocystoplasty for multiple reimplantation failures of refluxing megaureter to atrophic bladder: A novel technique and its long term outcome

Shigeru Nakamura, Taiju Hyuga, Kazuya Tanabe, Taro Kubo, Satoru Inoguchi, Shina Kawai, Hideo Nakai
Department of Pediatric Urology, Jichi Medical University, Children’s Medical Center Tochigi, Japan

ABSTRACT

We encountered a 9-year-old boy with a small bladder who had previously undergone multiple ureteroneocystostomies for unilateral refluxing megaureter. He underwent excision of the affected non-functioning kidney and ureterocystoplasty used the dilated regional ureter, in which the loop shaped urinary bladder was reconstructed without detubularization of the dilated ureter. The long-term postoperative course has been satisfactory. There have been no reports of ureterocystoplasty used a dilated ureter after multiple ureteroneocystostomies and none describing ureterocystoplasty in which the ureter was looped. This case is presented herein.

Key Words: Augmentation cystoplasty, ureterocystoplasty, ureteroneocystostomy.
small bladder who had undergone 3 times ureteroneocystostomies for unilateral refluxing megaureter but in whom recurrent febrile urinary tract infection (UTI) could not be controlled. The patient underwent excision of the affected non-functioning kidney and ureterocystoplasty used the dilated regional ureter, in which the loop shaped urinary bladder was reconstructed.

Case report
The patient, a boy 9 years and 4 months of age, presented with no neurological abnormalities. MRI findings of the spine were normal. For right vesicoureteral reflux (VUR) Grade V (Fig. 1-a), the patient had undergone right ureteroneocystostomy at another hospital 8 months after birth. Dimercaptosuccinic acid (DMSA) renal scintigraphy showed right split renal function of 25%. During the postoperative course, bilateral VUR developed and febrile UTI recurred (Fig. 1-b,c). Therefore, at the same hospital, the patient underwent bilateral ureteroneocystostomy including right ureteroplasty (resection tapering) at the age of 2 years. However, recurrent right pyelonephritis could not be controlled. Thus, the patient was referred to our hospital at the age of 5 years for detailed examination and treatment. VUR was found in the bilateral dilated ureters and no anatomical urethral obstruction was found on voiding cystourethrography (VCUG) (Fig. 2-a-c). While the bladder capacity was small at only 50 mL, it was confirmed that the patient could void without post-void residual urine. Uroflowmetry (UFM) showed normal (Bell-shaped) curve and voided urine volume and post-void residual urine volume were 56 ml and 5 mL respectively.

On cystometrogram (CMG), the bladder capacity was 70 mL, and the intravesical pressure was 6 cmH₂O. The patient was diagnosed with mildly increased intravesical pressure. Timed voiding was started in combination with anticholinergic administration. Voided urine volume increased to 100-150ml on bladder diary. However, post-void residual urine volume also increased and, recurrent right pyelonephritis could not be controlled. DMSA renal scintigraphy showed right split renal function of 19%. Radical surgery for bilateral VUR was considered to be necessary, and the patient underwent bilateral ureteroneocystostomy (psoas bladder hitch procedure with bilateral ureteroplasty (folding) at the age of 6 years and 11 months. VCUG was performed 5 months after surgery, revealing a heart-shaped bladder deformity due to the effect of bilateral psoas bladder hitch procedure and the disappearance of the left VUR, while the right VUR worsened, resulting in the formation of a urine reservoir consisting of the bladder and the right ureter with a urine storage capacity of 280 mL (Fig. 2-d). When urine was stored to the maximum capacity, the pressure in the entire reservoir involving the bladder and right ureter was low at 8 cmH₂O. In addition to urotherapy, clean intermittent catheterization (CIC) was started twice daily because post-void residual urine in bladder and right renal pelvis was confirmed after spontaneous voiding. However, the patient refused the CIC due to urethral pain associated with catheter insertion, subsequently experiencing recurrent right pyelonephritis. DMSA renal scintigraphy showed that recurrent pyelonephritis exacerbated the right renal function, resulting in the non-functioning kidney.

At the age of 9 years and 4 months, the patient underwent right nephrectomy and ureterocystoplasty to enable spontaneous
voiding. The right kidney was first removed transabdominally (Fig. 3-a) and the renal pelvis was separated at the border between the kidney and the extrarenal pelvis (Fig. 3-b). The vessels from the renal artery to the renal pelvis were severed. A Pfannenstiel incision was then made to reach the prevesical space and the bladder was identified (Fig. 3-b). Since the right lower ureter had previously been used 3 times ureteroneocystostomies, excessive detachment from the surrounding tissue was considered likely to be the cause of poor blood

Fig. 1. Voiding cystourethrography before loop ureterocystoplasty: a) Before the first right ureteroneocystostomy; Right VUR Grade V and no bladder deformity are noted; b, c) Before the second ureteroneocystostomy; Bilateral VUR is noted. The bladder is mildly deformed.

Fig. 2. Voiding cystourethrography before loop ureterocystoplasty: a, b, c) Before the third ureteroneocystostomy; VUR is noted in the lower segments of the bilateral ureters. The bladder is mildly deformed. The patient can void without post-void residual urine; d) After the third ureteroneocystostomy; Severe right VUR and heart-shaped bladder deformity are noted.
supply to the right ureter, and detachment of the middle to distal portion of the right ureter from the surrounding tissue was therefore avoided. The blood flow in the right ureter was considered to possibly be exacerbated by detubularization of the ureter during augmentation cystoplasty. Thus, the right ureter was looped without being detubularized, and the right renal pelvis was reverted and anastomosed to the bladder by continuous suturing with 3-0 Vicryl suture (Fig. 3-c). A suprapubic catheter was placed for 14 days after surgery. On postoperative day (POD) 18, the patient was discharged from our hospital. VCUG at 12 months after surgery showed that the bladder capacity was 250 mL. Spontaneous timed voiding by abdominal straining was performed; 200 mL could be drained from the bladder, and the post-void residual urine volume was 50 mL on US (Fig. 4). At and after postoperative 1 year, voiding function was assessed by UFM, because the patient rejected VCUG and CMG due to urethral pain associated with catheter insertion, which revealed that, after 3 years, the patient’s bladder capacity had increased to 350 mL and subsequently remained unchanged, and no bladder overdistension was observed. To date, for the 7 years since the surgery, the postoperative course has been favorable. Intermittent spontaneous voiding by abdominal straining has been performed, enabling the drainage of 300-350 mL of urine, and the post-void residual urine volume has

**Fig. 3.** Intraoperative findings during loop ureterocystoplasty: a) The non-functioning right kidney is removed transabdominally via a para-rectus muscle incision; b) The dilated renal pelvis separated from the right kidney is reverted caudally; d) The anastomotic site between the right renal pelvis and bladder is visible. The black, white, and yellow arrows indicate the right kidney, right renal pelvis, and bladder, respectively.

**Fig. 4.** Voiding cystourethrography after loop ureterocystoplasty: a) A loop shaped bladder consisting of the bladder anastomosed to the right renal pelvis is observed during voiding (maximum bladder capacity: 250 mL); b) Post-void residual urine volume is 50 mL on ultrasonography.
remained as small as 10–25 mL without urinary incontinence. There have been no occurrences of febrile UTI postoperatively, and on DMSA renal scintigraphy, the unaffected left kidney has been functioning normally without renal scarring.

**Discussion**

To our knowledge, this is the first report of ureterocystoplasty in which the ureter after multiple ureteroneocystostomies was looped without being detubularized and was then anastomosed to the bladder. The various techniques of ureterocystoplasty have been reported [3-8]. As shown in this case, ureterocystoplasty is considered to be the most reasonable method in patients who have a small bladder with a markedly dilated ureter associated with a unilateral non-functioning kidney and who may be judged to be capable of spontaneous voiding.

VCUG before the first ureteroneocystostomy showed no bladder deformity (Fig. 1-a). Bladder deformity occurred and progressed during infancy after the first ureteroneocystostomy, resulting in decreased bladder capacity (Fig. 1-b,c and Fig. 2). Regarding the cause of the small bladder in this patient, no neurological abnormalities and no anatomical/functional urethral obstruction were detected and it remains unclear whether the patient’s condition was due to a detrusor disorder associated with multiple ureteroneocystostomies or to nonneurogenic neurogenic bladder [10, 11]. We should had been regarded his bladder function as low compliant rather than relatively high on the first CMG, because the patient had bilateral high grade VUR. Recurrent febrile UTI resulted in a non-functioning right kidney. Early cutaneous vesicostomy [10, 11] or early augmentation cystoplasty should have been performed before the loss of the right renal function. In the patient before the loss of the right renal function, the distal segment of the right ureter should be used for ureterocystoplasty, and the proximal segment of the right ureter should be anastomosed to the bladder [3, 5] or to the contralateral left ureter [4,7]. It is assumed that transureteroureterostomy [4,7] might have been selected for this patient because a 4th ureteroneocystostomy may well have increased the risk of complications such as VUR and ureteral obstruction.

Churchill et al. reported ureterocystoplasty used the ureter in a patient who had undergone a single ureteroneocystostomy [9]. In our patient with multiple ureteroneocystostomies, the blood supply from the iliac artery and the bladder to the lower segment of the ureter was considered to be insufficient. It was also considered to be preferable that the blood vessels from the renal artery to the renal pelvis and upper segment of the ureter be preserved during ipsilateral nephrectomy. However, it was essential to sever these vessels in order to pull the renal pelvis caudally to the bladder without tension. In this patient, the main blood supply to the right ureter was highly likely to be dependent only on the blood vessel distribution from the gonadal artery to the middle segment of the ureter. Churchill et al. reported that, even in patients presenting with damaged blood vessels from the iliac artery to the ureter due to prior surgery, blood supply to the ureter can be maintained even after detubularization of the dilated ureter used for ureterocystoplasty if blood supply from the renal and gonadal arteries to the ureter are preserved [9]. In our case, it appeared to not be feasible to preserve the blood vessels not only those from the iliac artery but also the vessels from the renal artery. Therefore, it was
determined that, for preservation of blood supply to the ureter, the dilated ureter to be used for augmentation cystoplasty not be detubularized. Accordingly, loop ureterocystoplasty was performed. The persistent of reflux to the ureteral stump after nephrectomy is often symptomatic after ureterocystoplasty. In the patient, no persistent of reflux to the ureteral stump after nephrectomy was observed. This is the advantage of our loop ureterocystoplasty over standard ureterocystoplasty. The difference between ileum and ureter is bladder compliance after augmentation cystoplasty. The bladder compliance after ureterocystoplasty is higher. Therefore, some of the patients who underwent ureterocystoplasty can void spontaneously. In the case we present here, the persistence of massive post void residual urine in the bladder due to the urine in refluxing ureter was observed before loop ureterocystoplasty. Therefore, the patient was required CIC for the purpose of preventing from recurrent UTI. However, after loop ureterocystoplasty, the urine in refluxing ureter flowed into bladder automatically. The patient was not required CIC and satisfactory spontaneous voiding could be achieved employing timed voiding by abdominal straining. We didn’t perform a construction of abdominal continent catheterizable stoma, because some of the patients who underwent ureterocystoplasty could void spontaneously. However, if the patient had not voided spontaneously after ureterocystoplasty, we would have constructed abdominal continent catheterizable stoma in reoperation. Further long-term follow-up examinations are considered to be essential for monitoring of the functions of the unaffected left kidney and the bladder function.

Conclusion

When ureterocystoplasty is performed on patients with a history of multiple surgeries on the distal side of the ureter, loop ureterocystoplasty might be one of the treatment options. However, it must be recognized preoperatively that, if poor blood flow in the renal pelvis and ureter to be used for augmentation cystoplasty is observed intraoperatively, ureterocystoplasty should be discontinued and switched to enterocystoplasty with or without abdominal continent catheterizable stoma.

Compliance with ethical statements

Conflicts of Interest: None.
Financial disclosure: None.
Consent: All photos were taken with parental consent.

ORCID ID of the authors
Shigeru Nakamura /0000-0002-3607-5974
Taiju Hyuga / 0000-0002-3455-9094
Kazuya Tanabe /0000-0001-9052-4059
Satoru Inoguchi /0000-0002-3528-6585
Shina Kawai /0000-0002-5938-5377

References


Epidermoid inclusion cyst after urethroplasty: A rare complication

Volkan Sarper Erikci¹, Merve Dilara Oney¹, Gokhan Koyluoglu²

¹Departments of Pediatric Surgery, University of Health Sciences, Tepecik Training Hospital, Izmir, Turkey
²Departments of Pediatric Surgery, Katip Celebi University, Tepecik Training Hospital, Izmir, Turkey

ABSTRACT

Epidermoid inclusion cysts (EIC) is a disease caused by the nidation of the epidermal tissues and sebaceous materials into the dermal and subcutaneous layers. This entity is usually observed after trauma and surgical procedures. A 5-year-old boy with a ventral penile mass was admitted to our department. The history of the patient revealed that he was operated at the age of 1 year for hypospadias in a different medical center. The patient’s history also revealed that during the initial operation, a skin graft was used for covering urethroplasty. One year later the operation, a ventral penile mass was observed by his mother during wash. The mass continued to increase in size reaching of 1 cm in diameter. The mass was excised and the histopathological study revealed that the mass was compatible with an EIC. With 2 years of follow up the patient is disease free. EICs after penile surgical interventions may be a factor of anxiety for both the children and their parents. In order to avoid this unwanted complication, implantation of the superficial tissue layers into the deep the dermal and subcutaneous tissues should be avoided.

Key Words: Epidermoid inclusion cysts, hypospadias, complication, children.

Introduction

Epidermoid inclusion cysts (EIC) are rare in children. The pathophysiological mechanism in this disease is postulated to be the implantation of superficial tissue layers including the epidermal cells and sebaceous glands into the deep dermal and subcutaneous tissues [1, 2]. The typical occurrence is a painless swelling at the affected body sites. These cysts can occur anywhere on the body and typically present as nodules directly underneath the child's skin. They are usually mobile and the size of these cysts can be variable ranging from a few millimeters to several centimeters in diameter. These lesions may remain stable or progressively enlarge over time. These cystic masses usually become painful to the patient and may present as a fluctuant filled nodule below the patient's skin. Herein, after an unsuccessful surgical intervention for hypospadias, a 5-year-old boy
with an EIC located at the ventral aspect of the penis is presented and the topic is discussed and a brief literature review is given.

Case report
A 5-year-old boy complained of a penile mass located at the ventral aspect of the penis. The history of the patient revealed that he had been operated for hypospadias at the age of 1 year. At the initial operation a skin graft had been used in order to cover urethroplasty suture line. The family of the patient declared that initially the mass was small, 1 year after the operation it started to grow rapidly (Fig. 1).

Fig. 1. Preoperative appearance of penile EIC.

Under general anesthesia a surgical intervention was performed with a vertical incision on the ventral aspect of the penis. The mass with a dimension of 2x1.5x0.7 cm was totally excised (Fig. 2, 3). The urethra was not catheterized intra-operatively and the fashion of penile skin closure after EIC removal was interrupted simple suture using 5/0 polyglicolic acid. Histopathological examination findings were compatible with epidermal inclusion cyst with a capsule at the outermost and inside of which there was keratinized material. With an uneventful postoperative follow-up of 2 years, he is disease free without any symptoms of recurrence.

Fig. 2. Operative view of EIC during excision.

Fig. 3. Postoperative view of the penis after removal of EIC.

Discussion
As an intriguing clinical entity, EIC is supposed to occur due to implantation of the superficial squamous cells and oleaginous materials into deep tissues following an injury and surgical procedure [1, 2]. Accepted as real cysts, these masses contain keratinized material inside and have an outer capsule composed of keratinized squamous epithelial cells. EICs may either be congenital or acquired in origin. Congenital forms of penile EIC have been supposed to result from
abnormal embryologic closure of the median raphe [3]. Penile surgery and trauma are two of these factors. It has been suggested that during penile surgical interventions like circumcision or hypospadias surgery, epidermal cells are embedded within the dermis [4,5]. In some of these cases no initiating factor for EIC development is present and these are called as idiopathic forms of penile EIC [6].

EICs may be single or multiple and the size of the mass varies. Accumulation of epidermal materials including secretions and debris leads to formation of a cyst. This painless swelling gradually increases as the time passes [7]. In accordance with those reported previously, the cystic mass in the presented case was initially small, but later it became larger rapidly. Physical examination is enough in diagnosing these masses. In doubtful cases imaging modalities such as ultrasonography and other means of radiological studies may be useful in conforming the diagnosis. There are various diseases in the differential diagnosis of penile EICs and these are diverticula or fistulae at urethra, dermoids or teratomas [3]. Physical examination findings in our case was enough to diagnose EIC and no other diagnostic imaging studies were performed.

There are various complications that can be observed during the surgical intervention for EIC. These are rupture of the cyst, infection, hematoma and rarely carcinomas [8,9]. Rupture deserves special attention because rupture of the cyst almost always ends up with an inflammatory reaction due to release of keratin inside the cyst. The time interval in our case is relatively long with a time period of 4 years between the initial hypospadias surgery and surgical excision of the EIC. Hopefully, despite rather long delay for treatment of EIC in our case, no complication was observed in our case during pre- and postoperative period.

Nevertheless, once EIC is diagnosed at the penis, there should not be delay in the management of these masses and gentle surgical excision should be the choice in order to avoid above mentioned complications. Complete surgical excision is curative in the management of penile EIC. In order to avoid local recurrences, meticulous dissection of the mass is paramount [1]. One of the important issues in the management of these masses is that there should not be violence against to the capsule of the EIC during surgical excision. Otherwise, local seeding of the surface materials into the dermal subcutaneous tissues may lead to local recurrence.

Histopathological examination is the cornerstone in confirming the diagnosis of EIC. These masses are typically lined by keratinized squamous epithelium and contain sebaceous material inside [7]. Histopathological examination findings were compatible with EIC containing cheesy material inside in our patient. The patient is disease free after a 2 year-follow up period.

Penile EICs are rare during childhood and can pose diagnostic dilemmas for health providers. Timely and appropriate surgical treatment is important in preventing complications such as infection, hematoma and rarely carcinomas. Most of the reported cases of EICs are children with penile masses after circumcision [10,11]. As a cause of urethro-cutaneous fistula, a penile EIC following urethroplasty has also been reported [3]. After an English language literature search for complications following hypospadias surgery in children, we did not find out EIC following hypospadias surgery in childhood. To our knowledge, after hypospadias surgery, our case is unique and probably the first case of EIC having an isolated EIC without any other urological complication.
Conclusions
Surgical interventions like circumcision and hypospadias repairs should be performed carefully. This is important in avoiding psychological and surgical trauma that the child may face following penile surgical interventions. During surgery, it is also important to avoid seeding of superficial tissue materials into the dermis and subcutaneous tissue. The diagnosis of EIC in children with penile cystic masses should be kept in mind and a gentle management including total excision of EIC without violating the capsule should be performed.

Compliance with ethical statements
Conflicts of Interest: None.
Financial disclosure: None.
Consent: All photos were taken with parental consent.

Acknowledgement

ORCID ID of Authors
Volkan Sarper Erikci /0000-0002-9384-2578
Merve Dilara Oney / 0000-0002-2895-2578
Gokhan Koyluoglu / 0000-0002-1140-169X

References
Bilateral single system vaginal ectopic ureters: A rare variant

Shyamendra Pratap Sharma, Sarita Chowdhary, Pranaya Kumar Panigrahi, Shiv Prasad Sharma

Department of Pediatric Surgery, Institute Of Medical Sciences, Banaras Hindu University, Varanasi, India

ABSTRACT

Bilateral single-system vaginal ectopic ureter (BSSVEU) is a very rare entity in pediatric urology. A 1-year-old girl was brought to outpatient clinic with continuous dribbling without normal voiding. The patient was diagnosed as BSSVEU by clinical findings, CT intravenous urography, genitoscopy and cystoscopy. Bilateral ureteric reimplantation was performed. Postoperative dry time was 2-3 hours after 3 months of follow-up and renal function was preserved at 3 months. Here, we report such a case and briefly discuss its diagnosis and management.

Key Words: Single-system ectopic ureter, vaginal ectopic ureter, ureteric re-implantation.

Introduction

An ectopic ureter opens other than the posterolateral aspect of the trigone, sometimes the ectopic opening is outside of the urinary system. Ectopic ureters are usually associated with a duplex kidney and, in general, 80% of ectopic ureters arise from the upper pole of the kidney. If an ectopic ureter drains a single kidney, it is called a single system ectopic ureter, which occurs in only 20% of cases [1]. Bilateral single-system ectopic ureters (BSSEU) are even rare. This is the first case of opening into the vagina with normal urethral opening and normal bladder neck. In this case, both ureters were opening in the vagina leading to the very small unused urinary bladder with very small capacity (< 5 ml). The objective of reporting this case is to describe the challenge of management in a young girl whose small urinary bladder has never been exposed to urine.

Case report

A 1-year-old female presented with continuous dribbling of urine and never passed urine in the stream since birth. Parents were of poor socioeconomic status and an illiterate rural background. There was no history of high-grade fever, chills & rigor, swelling over back, altered bowel habit. Mother was unbooked and antenatal history was uneventful.

On general examination, child laying comfortably in mother lap, afebrile, heart rate 110/min, Blood pressure 90/60 mmHg,
respiratory rate 38/min, no pallor, icterus, cyanosis or lymphadenopathy present. There were no skeletal defects, neurological examination and other systems were normal. The abdomen was soft, nontender, no organomegaly, Bowel sounds were normal. On per rectal examination anal tone normal, no mass felt, no sacral agenesis. Three orifices in the perineum (urethra, vagina, anus). On separating the labia majora, no urine coming per urethrae and urine pouring out through vaginal orifice (Fig. 1).

Fig. 1. Perineum of the child showing three openings (urethra, vagina, anus).

All routine investigations such as hemoglobin, total leucocyte count, renal function tests, and urine microscopic examination were within the normal range. Abdominal sonography revealed bilateral moderate hydroureteronephrosis (Right >> Left), small contracted urinary bladder, uterus, and ovaries were normal, no other anomaly detected. Micturating cystourethrogram was not possible. Genitogram was done showing ectopic ureteric opening in the vagina with reflux on the left side, the right system not visualized (Fig. 2). A CT intravenous pyelography showed right hydro-ureteronephrosis is more in comparison to the left kidney, bilateral ureters inserted to the lateral vaginal wall and contrast collection in the vaginal cavity and no calculus (Fig. 3).

Fig. 2. Genitogram showing ectopic ureteric opening in the vagina with reflux on the left side.

On genitoscopy and cystoscopy, the bladder neck was developed with an ill-defined trigone. Bladder capacity was approximately < 5 ml, normal vagina and ureteric orifice could not be visualized in the bladder or vagina. Bladder and vagina were catheterized for 24 hours, urine came out only from the vagina. The definitive diagnosis of Bilateral Single System Vaginal Ectopic Ureter (BSSVEU) was made and planned for single stage bilateral ureteric re-implantation.
Fig. 3. CT intravenous pyelography showing right hydroureteronephrosis more in comparison to the left kidney, bilateral Ureters inserted to the lateral vaginal wall and contrast collection in the vaginal cavity.

At surgery, the ureters were identified, traced and found to be opening into the postero-lateral wall of the upper vagina [Fig. 4]. Ureters were dissected and isolated from vaginal insertion, bilateral reimplantation was done over DJ stents. The bladder was too small to accommodate both DJ stents and urethral catheter so a 6 Fr infant feeding tube inserted and fixed. Infant feeding tube comes out on day 2, both DJ stents come out per urethra first on day 5 and the second one on day 7 itself.

Fig. 4. Intraoperative picture showing ureters opening into the postero-lateral wall of the upper vagina.

Fig. 5. Post-operative MCU at 3 months showing increased bladder capacity (30ml) with bilateral VUR, but no contrast retention after micturition.

On follow-up, after 1-month child passing urine in the stream and dry time for the child was 30 min. After 3 months the child remained dry for two hours. Renal function preserved. MCU show increased bladder capacity (30ml) with B/L VUR, but no contrast retention after micturition (Fig. 5). Antibiotic prophylaxis was given to the child. The mother was very happy to report that the excoriation around the perineum had healed, and the child was passing urine in the stream.

Discussion
We report a case of bilateral single-system ectopic ureters opening into the vagina associated with the hypoplastic bladder, well developed vesical neck and urethra, and bilateral ureteric reflux. This is an unusual location for bilateral single-system ectopic ureters. There is 3 other reports of bilateral single-system ectopic ureters
opening into urogenital sinus but not in well-developed vagina. First by Sheldon et al. [2], in their case, urogenital anomalies were more severe i.e., the vagina was rudimentary and both kidneys were dysplastic, resulting in end-stage renal disease. Second by Bhupendra P. Singh et al. [3], in their case, ectopic ureters opening into urogenital sinus associated with the absence of the urethra, hypoplastic bladder, incontinent vesical neck, and bilateral ureteric reflux. Third by Farzeen Sharaf et al. [4], in their case anatomy was similar to Sheldon and Welch except for the renal abnormalities. All three case reports urethral openings were in urogenital sinus. However, in our case the vagina was normal, urethra and vesical neck were well developed and both kidneys were normal.

The primary management of BSSEU may vary from bilateral ureteric re-implantation only for the attainment of continence, to adding a bladder augmentation and bladder neck reconstruction, at the same sitting or subsequently. Kesavan, et al. [5] showed the vesical neck and trigone not well developed in 54% of unilateral and 75% of bilateral ectopic ureters.

Bhupendra P. Singh et al. [3] stated that ureteric re-implantation alone may not attain continence in patients with BSSEU. On contrary to them Kumar et al. [6] report that a bladder with BSSEU may not necessarily be useless and bladder capacity increases with the passage of time without the need for augmentation.

In this case, the increase in bladder capacity and improvement in continence was achieved only by ureteric re-implantation. The well-developed vesical neck and adequate urethra helped us to consider this option. Hence, in cases of bilateral single-system ectopic ureters, management modalities should be individualized from patient to patient depending upon the anomalies in the lower urinary tract as well as the reproductive tract. A child presenting with dribbling of urine since birth, with or without normal micturition, needs to be investigated early and thoroughly. In our opinion, primary ureteral re-implant is an option to avoid staged procedure and augmentation surgery complications. Adequate time should be given and after re-implantation, bladder response to urine challenge should be observed.

**Compliance with ethical statements**

**Conflicts of Interest:** None.

**Financial disclosure:** None.

**Consent:** All photos were taken with parental consent.

**ORCID ID of the authors**

Shyamendra P Sharma /0000-0003-3518-1836
Sarita Chowdhary /0000-0001-8436-7544
Pranaya K Panigrahi /0000-0002-4072-202X

**References**


Multiple urethroperineal fistula in a boy with posterior urethral valve: A case report of an extremely rare association

Md. Samiul Hasan, Ipsita Biswas, Umama Huq
Department of Pediatric Surgery, Dhaka Shishu (Children) Hospital, Shere Bangla Nagar, Dhaka, Bangladesh

ABSTRACT

We describe a case of posterior urethral valve with multiple urethroperineal fistulas. This association is extremely rare. The crucial point in the diagnosis is to distinguish these fistulas from the urethral duplications that actually guide the treatment. The presence of these fistulas in patients with posterior urethral valve may have a beneficial effect on renal function as it reduce the pressure on the bladder. On the other hand, incomplete valve ablation may contribute to the recurrence of the fistula.

Key Words: Urethroperineal fistula, posterior urethral valve, urethral duplication.

Introduction

Congenital urethroperineal fistula (CUPF) is a very rare anomaly of the lower urinary tract in children. In addition to the normal orthotopic urethra, there exists an extra channel draining from prostatic urethra to perineum. This term sometimes creates confusion with hypospadiac variety of urethral duplication especially Y duplication (Effmann type IIA2). In contrast to duplication, the orthotopic dorsal urethra in cases of CUPF is of normal caliber and function [1,2]. On the other hand, posterior urethral valve (PUV) is a relatively common urinary tract anomaly in boys. Several urinary tract anomalies have been described in association with both of these two but the association of CUPF with PUV is extremely rare. In fact, we did not find them together in extensive literature review. We intend to describe a patient with posterior urethral valve who had multiple urethroperineal fistulas draining urine from the posterior urethra proximal to the valve, thereby reducing pressure on bladder and upper tract.

Case report

One and a half-year-old boy, second issue of his non consanguineous parents from a low socio economic background presented to us with dribbling of urine and straining during micturition since birth. Mother noticed three small openings in the perineum leaking urine during micturition for one year. There was no known congenital anomaly in the family. On
clinical examination, there were three openings in the perineum, right to the midline, surrounded by scar tissue (Fig. 1). The urinary bladder was palpable and both the kidneys were ballotable.

![Fig. 1. Three opening in the perineum surrounded by scar tissue.](image1)

Sonography revealed bilateral hydroureteronephrosis, thickened irregular bladder wall, and dilated posterior urethra. Micturating cystourethrogram (MCUG) showed normal anterior urethra, dilated and elongated posterior urethra. There were two extra channels leading from the posterior urethra to the perineum. The urinary bladder was thick, irregular and had multiple diverticulums. High-grade vesicoureteric reflux (VUR) was present on the right side (Fig. 2).

We did cystoscopic valve ablation and excision of all three fistulas. Postoperatively, the boy is doing well. The perineum is dry. We have a plan to check MCUG after 6 months.

**Discussion**

Multiple urethroperineal fistulas are metaphorically called ‘watering can’ perineum. This condition is usually associated with a chronic and severe infection of urethra and perineum. Most commonly occur in gonorrhea, schistosomiasis, tuberculosis and Crohn’s disease. Its association with the posterior urethral valve has never been found [3].

Congenital urethroperineal fistula is usually single, often deviates on either side of the midline but there is no side preponderance [1]. In this case, there were three opening visible in perineum right to the midline though; MCUG visualized two, maybe due to inadequate pressure in the third tract.

Differentiation of CUPF from urethral duplication is critical which directs the treatment of these conditions. In CUPF, the normally positioned orthotopic urethra is of normal caliber and function. The excision of the fistulous tract is usually curative. On the other hand, ventrally placed urethra in duplication is the main urethra and excision of...
which could be devastating [4]. MCUG along with Cystoscopy are enough to differentiate between these two. Some authors used MRI and fistulogram for confirmatory diagnosis [5,6]. Associated upper tract anomalies have been reported in patients with urethroperineal fistula. Most common are vesico-ureteral reflux (VUR), renal dysplasia and agenesis [4,5]. This patient also had right sided VUR which might be due to the presence of PUV. Urethroperineal fistula in patients with posterior urethral valve could have some beneficial effects also. It reduces the pressure on the bladder and upper tract thereby helps to protect renal function like a pressure pop-off. Different pressure pop-off mechanisms have been described before like unilateral reflux & renal dysplasia, bladder diverticulum, urinoma, and urinary ascites but urethroperineal fistula has never been described in association with posterior urethral valve [7]. Our patient had bladder diverticulum and unilateral reflux and serum creatinine was normal at presentation. Fistulous tracts in this patient were excised after ablation of posterior urethral valve. It is accepted that reduced pressure in posterior urethra after valve ablation will prevent recurrence of fistula. Recurrence of fistula has been reported requiring re operation [4]. Long term follow up is necessary for this patient to comment on recurrence. Incomplete valve ablation or post-operative urethral stricture may contribute to recurrence of fistula.

**Conclusion**

Urethroperineal fistula is an extremely rare association with posterior urethral valve which could have a beneficial effect on renal function but it should be carefully differentiated from urethral duplication which requires completely different management.

**Compliance with ethical statements**

**Conflicts of Interest:** None.

**Financial disclosure:** None.

**Consent:** All photos were taken with parental consent.

**ORCID iD of the authors**

Md. Samiul Hasan /0000-0001-5470-2241

Ipsita Biswas /0000-0001-7519-4899

Umama Huq /0000-0002-7438-783X

**References**


