## Prolapsing ectopic ureterocele presenting as a vulval mass in a newborn girl

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ABSTRACT

Prolapse of the ureterocele and its presentation as a vulval mass is an extremely rare condition. There are no detailed imaging findings of such cases in the literature. We present voiding cystourethrography and MRI findings of a newborn girl with a ureterocele extending through the urethra.

*Key words:* • *ureterocele* • *pediatrics* • *urogenital abnormalities* • *magnetic resonance imaging* 

A n ectopic ureterocele with ureteral duplication is a relatively frequent urinary system anomaly in children; however, prolapsing of the ureterocele through the urethra is a rare presentation. Diagnosis based on ultrasonography (US) or intravenous urography (IVU) may be inconclusive (1). We report imaging findings of a prolapsed ureterocele that was presented as a vulval mass in a newborn girl. To the best of our knowledge, magnetic resonance imaging (MRI) findings in such a case have not been previously described.

## Case report

A 1-month-old girl was referred to our hospital for evaluation of recurrent urinary tract infections and a cystic vulval mass (Fig. 1). Abdominal US revealed dilatation of the upper pole collecting system and draining ureter of the right kidney, with an ureterocele in the bladder. Transperineal US showed a simple cyst without an internal solid structure. Voiding cystourethrography (VCUG) showed an ovoid, well-defined filling defect distal to the widened urethra (Fig. 2). No vesicoureteral reflux was seen. MRI was performed to demonstrate the full extent of abnormality and to clarify the vulval cystic mass. MRI showed a markedly dilated right upper pole collecting system associated with a large ipsilateral ureterocele, which was outpouching from the urethral meatus (Fig. 3). With these imaging findings the diagnosis of prolapsed ureterocele (through the urethra to the extrameatal space) was reached. Right upper nephroureterectomy and ureterocele excision were performed.

## Discussion

An ureterocele is a cystic dilatation of the submucosal distal ureter. It can be either intravesical or extravesical (ectopic). An ureterocele prolapsing through the urethra is a rare presentation. While Scott (2) reported an incidence rate less than 5%, Shekarriz et al. (3) did not find any cases of prolapse in a review of 106 children. Weakness of the urethral wall distal to the ureterocele may cause such an abnormality (4). It is more common in infants and may be complicated by urinary obstruction and infection. There are only a few reported cases of prolapsed ureterocele presenting as a vulval mass (1, 5, 6), but detailed imaging findings of this abnormality have not been previously described in the literature.

US is a relatively inexpensive and widely available imaging technique; therefore, it may be used initially to reveal an intravesical ureterocele and associated duplex system. However, it may only provide limited information about the extravesical segment of the ureterocele (7). We attempted to perform transperineal US in our patient; however, the cyst was relatively small and it was difficult to reveal the relationship

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Received 6 October 2006; revision requested 6 December 2006; revision received 8 December 2006; accepted 29 December 2006.

between the intravesical segment and urethra. IVU may demonstrate pelvicalyceal and ureteral anatomy. It may also show an ureterocele as an ovoid intravesical filling defect. Nonetheless, it is not an adequate method for evaluating the urethra. Furthermore, ionization radiation limits its common use in the pediatric population. VCUG is an ideal method for evaluating urethral abnormalities. Dilatation of the urethra with a cystic urethral diverticula like structure may be a clue suggesting ureterocele prolapse; however, VCUG is an invasive method and exposes patient to radiation. MRI seems to be the best possible non-invasive method for a definitive diagnosis. It produces high quality images for assessment of the upper and lower urinary tracts. It may demonstrate the extension of the ureterocele through the urethra. A cystic vulval mass and its association with an ureterocele and duplex system can easily be identified. It is also a valuable technique for patients who need combined morphologic assessment of the entire urinary tract. Major disadvantages of MRI include its relatively high cost and the need for sedation in young children.

The differential diagnosis includes cystic lesions of the vulva, such as an epidermal inclusion cyst, Skene's duct cyst, and hidradenoma papilliferum. Presence of an ureterocele in the bladder, a connection between the 2 portions of the ureterocele extending between the bladder and external urethral meatus through a wide urethra should suggest the diagnosis of a prolapsing ureterocele.

In conclusion, a prolapsing ureterocele may, on rare occasions, present as a vulval cystic mass. US, IVU, and VCUG may be inconclusive for making a definitive diagnosis. We suggest that MRI may be a good alternative modality for clarifying the exact abnormality in such cases.

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**Figure 1.** Photograph of an oval cystic mass protruding through the urethra.



**Figure 2.** Voiding cystourethrography reveals a wide urethra with a distal ovoid cyst-like outpouching filling defect (*arrowheads*).



**Figure 3.** *a*, *b*. Coronal single-shot turbo spin-echo T2-weighted MR image (a) shows dilatation of the collecting system with a large ureterocele in the bladder (*star*). An ovoid cyst is seen as an irrelevant structure distal to the bladder (*arrow*). Sagittal T2-weighted turbo spin-echo MR image (b) through the pelvis depicts the ureterocele (*star*) in the bladder with an ovoid cyst (*arrow*) protruding from the external urethral meatus.

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