

CASE STUDIES

Prolapsed, Ectopic Cecoureterocele

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This case study presents the prenatal diagnosis of a rare prolapsed, ectopic cecoureterocele. The diagnosis was confirmed by postnatal urethro-cystography, sonography, and surgical intervention.

Key words: ureterocele, ectopic ureterocele, cecoureterocele

Ureteroceles occur in approximately 1 in every 4000 children and occur most commonly in Caucasians. Females are affected four to seven times more often than males. Although there appears to be a slight left-sided preponderance, approximately 10% of ureteroceles occur bilaterally. Ureteroceles may be classified as intravesicular, defined by their presence entirely within the bladder, or extravesicular, defined by the permanent presence of some portion of the ureterocele at the bladder neck or urethra. They are also classified based on the location of insertion of the ureter into the bladder or based on their association with single or duplicated renal systems.¹

Extravesicular ureteroceles are also termed *ectopic ureteroceles*. This case report demonstrates a rare presentation of a prolapsed ectopic cecoureterocele. Ectopic ureteroceles typically arise from the upper pole moiety of a duplicated collecting system and are more common in the pediatric population.²

Case Presentation

A woman in her mid-20s, gravida 2, para 2, presented at 22 weeks gestation for a routine anatomic survey. All prenatal sonographic examinations were performed on a GE prologic 400 system, using a phased-array 5.0-MHz transducer. The examination was unremarkable with the exception of a thin-walled membrane/septation within the urinary bladder. This was thought to represent a

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FIG. 1. A 2.5-cm cystic mass extending outward between the fetal labia.

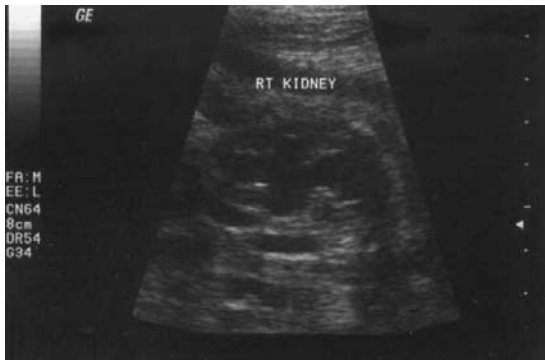


FIG. 2. Fetal right kidney demonstrating moderate hydronephrosis.

ureterocele. There was no evidence of hydronephrosis or hydroureter on either side. The woman returned at 32 weeks gestation. At this time, the urinary bladder membrane/septation was more prominent, and there was now a 2.5-cm cystic mass extending outward between the fetal labia (Fig. 1). The right kidney demonstrated moderate hydronephrosis (Fig. 2) with a tortuous hydroureter, whereas the left kidney showed similar but milder dilatation. The differential diagnosis included a prolapsed ectopic ureterocele, urethra prolapse, or periurethra cyst with a moderate urinary outlet obstruction. The patient was followed weekly until term to monitor the degree of urinary obstruction as well as amniotic fluid levels. Throughout this time, amniotic fluid remained within normal limits, and the bilateral hydronephrosis and hydroureter did not change significantly.



FIG. 3. Fetal bladder with an intravesicular filling defect.

At 38 weeks gestation, the woman delivered a 7-lb, 10-oz female infant. There was a 2.5-cm cystic mass extending outward between the labia. The mass appeared to originate from the perineum between the urethra and vagina. A Foley catheter was inserted into the urinary bladder. Urine output appeared normal.

An urethrocytogram was performed. Instillation of contrast material into the bladder demonstrated an intravesicular filling defect (Fig. 3) without evidence of ureteral reflux either with contrast filling or infant voiding. The cystic mass was diagnosed as a prolapsed, dumbbell-shaped ectopic ureterocele. The ureterocele appeared to originate from the right side of the bladder. Forty-eight hours later, renal and bladder sonograms were performed using an Acuson Sequoia system with a 6.5-MHz phased-array transducer. Examination of the right kidney demonstrated a duplicated collecting system with a small dysplastic upper pole moiety (Fig. 4). The upper pole appeared to have a single small collecting system. The lower pole of the duplicated system exhibited moderate to marked dilatation (Fig. 5). There was a redundant, tortuous, dilated, peristalsing ureter extending from the right kidney down to the bladder (Fig. 6). It was unclear whether its origin was the upper or lower pole system. The left kidney appeared to have a moderately dilated single collecting system without evidence of hydroureter. Except for the right upper pole, both renal cortices appeared normal in texture and appearance. Examination of the bladder and perineum showed a dumbbell-shaped ectopic

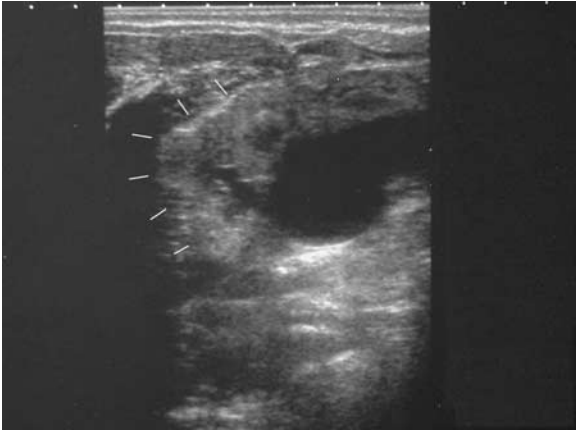


FIG. 4. Fetal right kidney with a small dysplastic upper pole moiety.



FIG. 6. Redundant, tortuous, dilated, peristalsing right ureter.

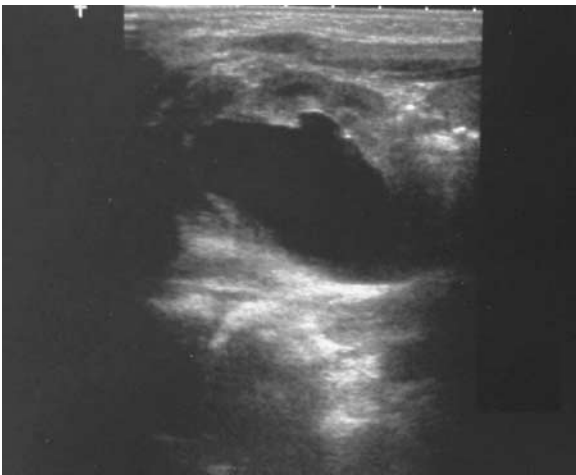


FIG. 5. Lower pole of the fetal right kidney exhibiting marked dilatation.



FIG. 7. Ureterocele within the urinary bladder.

ureterocele with a portion extending into the urinary bladder (Fig. 7), tracking parallel and posterior to the urethra and extending outward to the interlabial region (Fig. 8). The sonographic findings concurred with the urethrocytogram conclusion.

After reviewing the diagnostic testing and performing a thorough physical examination, a pediatric urologist further classified the ureterocele as a rare cecoureterocele. He then incised and deflated the external component of the ureterocele at the infant's bedside without complications. The infant's urinary tract will be monitored over several months to assess the degree of hydronephrosis and

hydroureter. If the dilatation does not resolve satisfactorily over time, surgical reimplantation of the ureter may be considered. In addition, surgical removal of the external ureterocele membranes will be scheduled at a later date.

Discussion

A ureterocele is a submucosal cystic dilatation of the terminal segment of the ureter. The true etiology of ureteroceles is not known. Several postulated theories include the incomplete dissolution of the Chwalla membrane during embryologic devel-

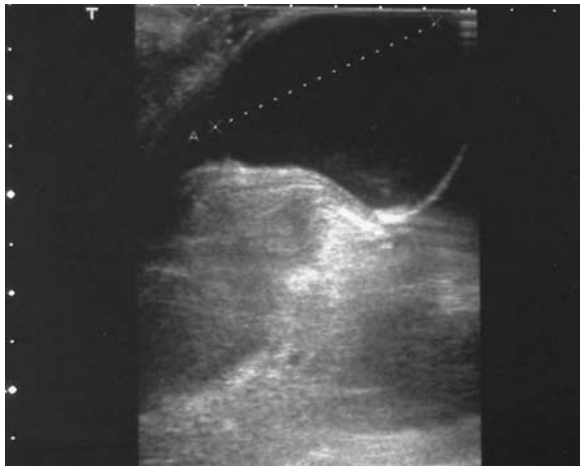


FIG. 8. Ureterocele extending outward to the interlabial region.

opment, altered development of the ureteral bud, inadequate muscularization, *Schistosoma haematobium* infection, excessive dilation of the intramural ureter during the development of the bladder and trigone, and trauma. Single-system ureteroceles are those associated with a single kidney, a single collecting system, and a solitary ureter, whereas duplex-system ureteroceles are associated with kidneys that have a completely duplicated collecting system and two ureters. Orthotopic ureteroceles are located in a normal position in the bladder. They usually arise from a single renal unit with one collecting system and are more common in adults, whereas ectopic ureteroceles are located in an ectopic position, such as the bladder neck or urethra.

In contrast to intravesicular ureteroceles, the incidence of ectopic ureteroceles has been reported to be approximately 80% in most pediatric series. Similarly, approximately 80% of ureteroceles are associated with the upper pole moiety of a duplex system.³ When ectopic ureteroceles are associated

with duplicated collecting systems, the upper pole moiety may be dysplastic or poorly functioning.^{2,3}

Mandell et al⁴ proposed a classification based on the features of the affected ureteral orifice as follows:

- Stenotic ureteroceles are defined as ureteroceles that are located inside the bladder with an obstructing orifice.
- Sphincteric ureteroceles lie distal to the internal sphincter. The ureterocele orifice may be normal or patulous, but the distal ureter leading to it becomes obstructed by the activity of the internal sphincter.
- Sphincterostenotic ureteroceles have characteristics of both stenotic and sphincteric ureteroceles.
- Cecoureteroceles are elongated beyond the ureterocele orifice by tunneling under the trigone and the urethra.

Conclusion

This case report demonstrates a rare presentation of a prolapsed ectopic cecoureterocele. It offers a valuable opportunity for review of the clinical and diagnostic features of this urinary system abnormality and its many variations.

References

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