Case 267: Cecureterocoele Manifesting in a Man with Infertility

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Conflicts of interest are listed at the end of this article.

Radiology 2019; 292:263–266 • https://doi.org/10.1148/radiol.2019162781 • Content code: GU • ©RSNA, 2019

History A 28-year-old man presented with lifelong anejaculation, which had become an issue because of family planning. The patient had a history of normal erections and experienced the sensation of orgasm without ever ejaculating. On physical examination, both testes were present in the scrotum, with normal dimensions and a normal epididymis bilaterally. The patient had a slightly tender left testicle, and digital rectal examination findings were normal.

The patient underwent further investigation for the possibility of retrograde ejaculation with urine cytology, the results of which were negative. Genetic testing was performed to exclude Y chromosome microdeletions. Serum-luteinizing and follicle-stimulating hormone levels were normal, with a borderline low level of testosterone (7.6 nmol/L; normal range, 8.0–29.0 nmol/L). All other pertinent laboratory results were noncontributory.

Pelvic MRI was requested to exclude an anatomic cause of anejaculation. MRI was performed in accordance with the standard clinical prostate protocol, with a dynamic contrast material–enhanced study. CT of the upper abdomen was also performed. The patient subsequently underwent cystoscopy, which revealed an intravesicular fluid-filled mass near the left ureteric orifice.

Part one of this case appeared 4 months previously and may contain larger images.

Imaging Findings

MRI showed a lobulated cystic structure within the bladder that was contiguous with the ureter and extended submucosally from the left ureteric orifice into the prostatic urethra (Figs 1, 2). The structure measured approximately 5 × 4 cm, with homogeneous intermediate T2 and high T1 signal, consistent with hemorrhage or proteinaceous content (Fig 3). In addition, the cystic lesion tunneled inferiorly through the bladder neck, anterior to the urethra. No hydroureter was visible on the MRI, and the distal portion of the left ureter was mildly dilated and contained blood products or proteinaceous content. Imaging of the upper abdomen showed an atrophic left kidney and a normal right kidney (Fig 4). A subsequent mertiatide (or MAG_3) renogram (not shown) enabled us to confirm there was no functional contribution from the left kidney.

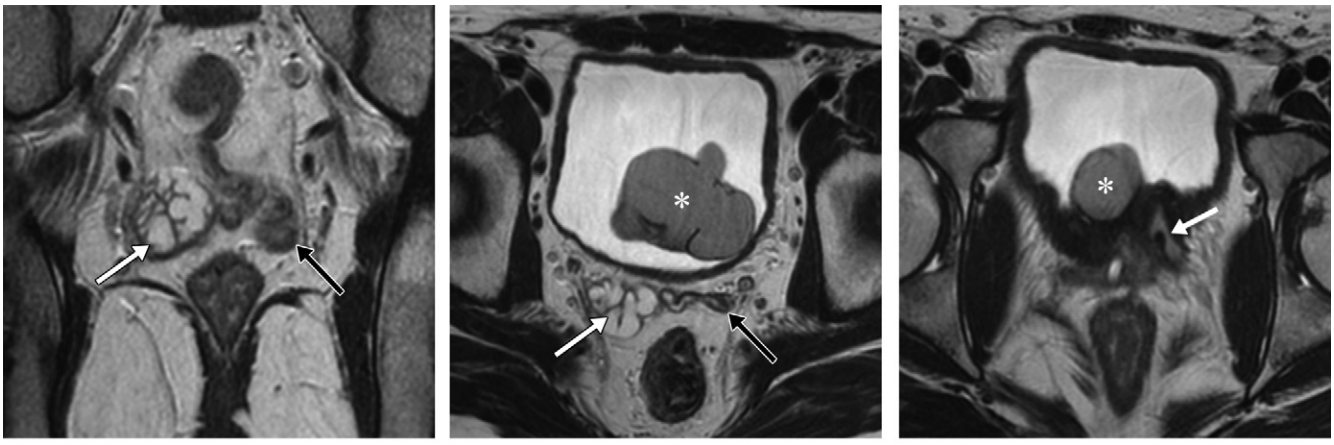
The right seminal vesicle was prominent in appearance, while the left seminal vesicle was atrophic (Fig 1b), with both ejaculatory ducts appearing dilated (Fig 2c). Additionally, there was a tubular structure adjacent to the left bladder wall corresponding to the dilated left vas deferens, which together with the left ejaculatory duct demonstrated high T1 signal in keeping with internal blood products (Fig 3a). The described findings are in keeping with a cecoureterocele, with an atrophic nonfunctioning left seminal vesicle and presumed obstruction of the right ejaculatory duct resulting in anejaculation.

Subsequent flexible cystoscopy was performed, which revealed a large cystic mass at the bladder neck arising primarily from the left side. This finding enabled us to confirm there was no communication with the urethra, indicating cecoureterocele was the most likely diagnosis (Fig 5). The patient was referred to a specialist center for further management of his infertility.

Discussion

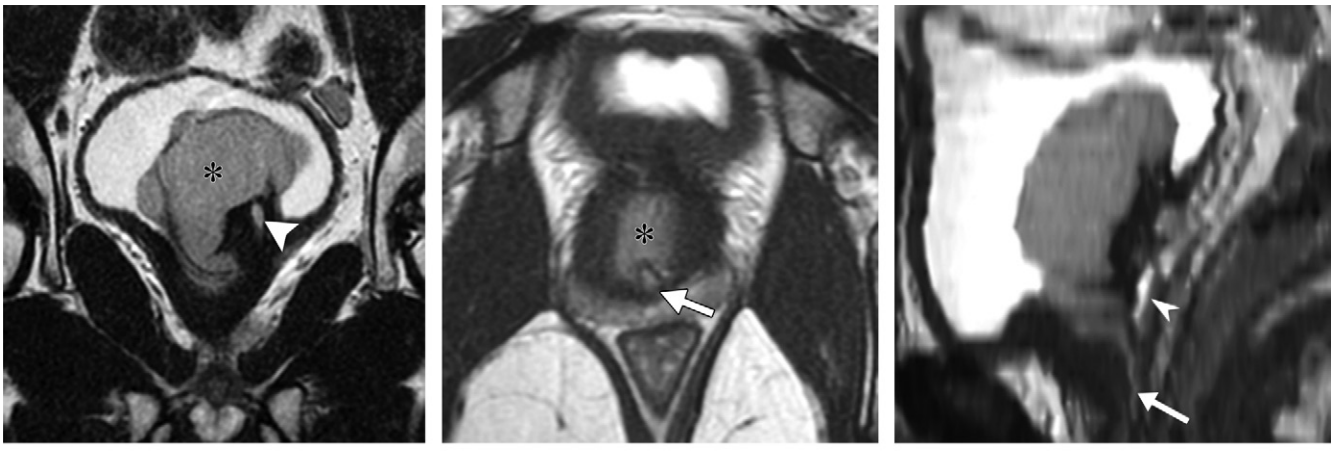
The initial differential diagnosis would include Zinner syndrome, ureterocele with secondary complications (hemorrhage, infection, or both), and cecoureterocele.

Zinner syndrome is a congenital anomaly of the Wolffian ducts in men and is considered equivalent to Mayer-Rokitansky-Kustner-Hauser syndrome in women. The findings in this syndrome comprise a triad of a seminal vesicle cyst, ipsilateral renal agenesis, and ejaculatory duct obstruction (1). Initial diagnosis of congenital urinary anomalies is usually made with perinatal US, which may delineate a cystic structure in the bladder lumen or otherwise depict megaureter secondary to obstruction. A relevant history was not present in this patient. He did not report perineal pain, chronic or recurrent genitourinary infections, or painful ejaculation, all of which have been previously reported by patients with this syndrome (2). The syndrome consists of a triad of renal agenesis, a seminal vesicle cyst, and ejaculatory duct obstruction (1). The syndrome is hypothesized to be caused by an insult during fetal development between the 4th and 13th gestational weeks. During this time, there may be incomplete migration of the ureteric bud, leading to a failure in the development of the metanephric blastema, and in turn leading to renal and concomitant ejaculatory duct agenesis (2,3). In this case, the left kidney is visualized; however, it is atrophic, likely due to chronic obstruction at the vesicoureteric junction. The key finding that lets us rule out Zinner syndrome



a. b. c.

Figure 1: Images from coronal **(a)** and **(b, c)** axial fast spin-echo T2-weighted MRI of the pelvis, with **b** being superior to **c**, show the atrophied left seminal vesicle (black arrow) and prominent right seminal vesicle (white arrow in **a** and **b**). Ectopic insertion of the left ureter is seen (arrow in **c**) here and joins the lobulated intravesicular cecoureterocele (∗) in Figure 2c.



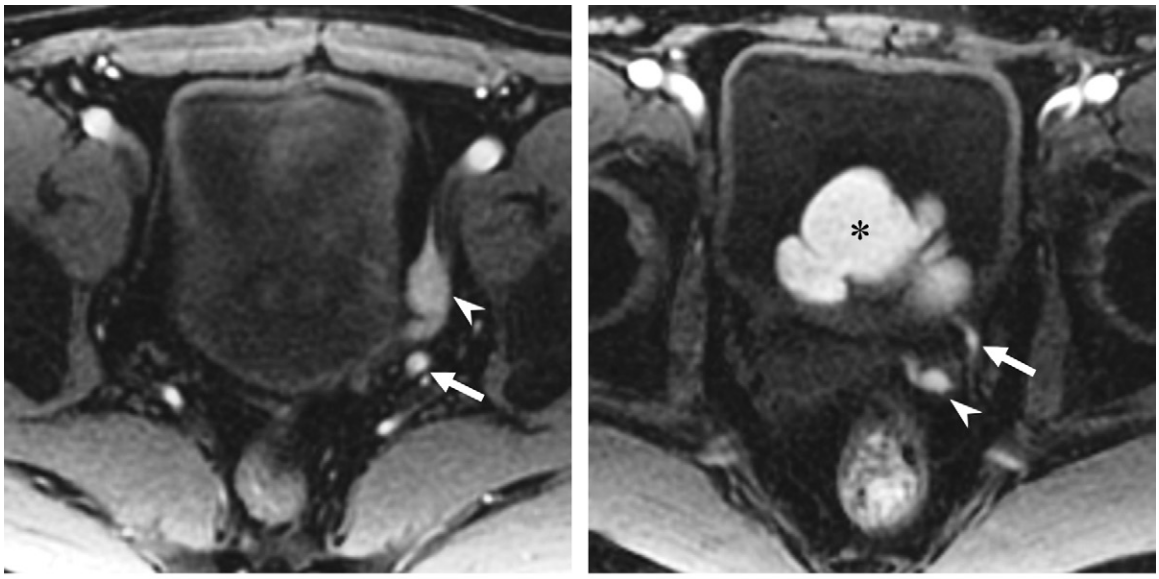
a. b. c.

Figure 2: **(a)** Coronal T2-weighted (repetition time msec/echo time msec, 4574/86.5) MRI of the pelvis shows the left ureter (arrowhead). **(b)** Axial T2-weighted (3000/85.4) MRI of the pelvis shows the urethra (arrow). Note the cecoureterocele (∗), which is anterior to and does not communicate with the urethra. **(c)** Sagittal reformatted MRI from **b** shows the right ejaculatory duct (arrowhead) and urethra (arrow).

in this case is the lack of ipsilateral seminal vesicle cysts, which were previously consistently reported in Zinner syndrome. The left seminal vesicle visible in Figure 1b demonstrates atrophy and is clearly separated from the reported lesion found lower in the prostatic urethra. In this case, the ejaculatory duct obstruction was caused by a cystic structure tunneling through the prostatic urethra and not by a cyst in the seminal vesicle. Additionally, there is a normal Y-shaped rather than a discoid adrenal gland on the left side, indicating normal in utero development of the kidney (4). These presented features enabled us to exclude Zinner syndrome in this case.

A ureterocele is a relatively common congenital anomaly and represents cystic dilatation of the ureter at the insertion to the urinary bladder. The diagnosis is usually made by identifying the ureteric insertion on fluoroscopic or cross-sectional studies, and it is estimated that around 75% of patients with ureteroceles have a duplicated collecting system (5). Most patients with this condition present during infancy with recurrent urinary tract infections and are rarely asymptomatic until adulthood (6). In this patient, the cystic structure was present inferior to the ureteric orifice, tunneling caudally alongside the prostatic urethra. One collecting system was present, which was not consistent with the diagnosis of an ectopic ureterocele. The findings in the present case cannot be explained by a simple ureterocele.

Cecoureterocele is a subtype of ureterocele with an intravesicular orifice and an additional blind-ending submucosal pouch that extends alongside the urethra caudally in the submucosal plane. It is rarely diagnosed in adults because the presenting symptoms are usually evident in early childhood. In children, these symptoms include vesicoureteric or bladder outlet obstruction with prolapse. Additionally, current screening with prenatal US for congenital anomalies



a. b.

Figure 3: Images from **(a, b)** unenhanced axial fat-saturated T1-weighted (6.2/3.1) MRI show the left ureter (arrow), left vas deferens (arrowhead in **a**), left seminal vesicle (arrowhead in **b**), and cecoureterocele (*).

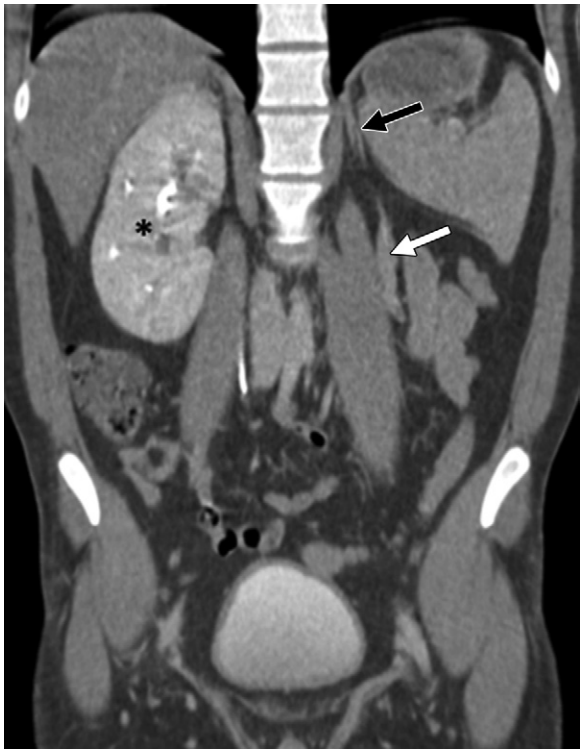


Figure 4: Coronal CT urogram shows remnant left kidney (white arrow), normal Y-shaped left adrenal gland (black arrow), and normal right kidney (*).

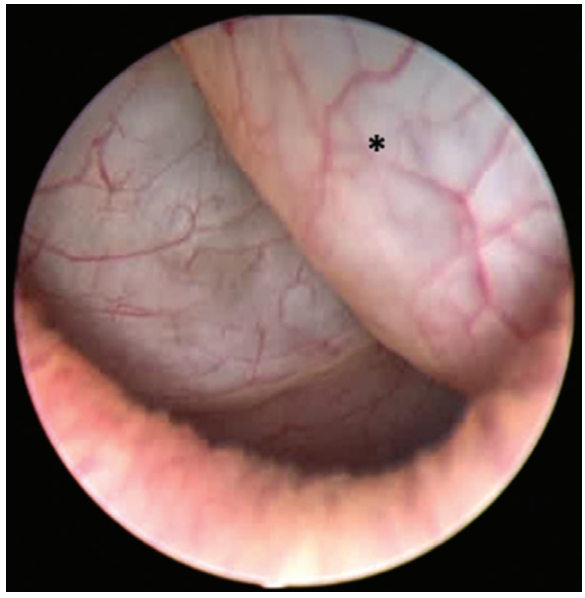


Figure 5: Image obained at cystoscopy shows the cecoureterocele (*) as a dilated cystic structure.

usually enables identification of this type of lesion much earlier (5,6). On US images, a cystic structure may be visible in the vicinity of the vesicoureteric junction, with associated reflux of obstruction. In adults, case reports of cecoureteroceles describe a different presentation, which accounts for a more indolent course. The few reports of cecoureterocele in adults have described patients presenting with prolapse of the cecoureterocele through the external genitalia and have occurred in women, which is more likely because of the short female urethra (7–10). In this case, the nonfunctioning kidney was likely not diagnosed in childhood, and atrophy occurred later. The unusual symptom of anejaculation was initially investigated for association with hormonal or genetic diseases and retrograde ejaculation. On the MRI (Fig 1c), the left ejaculatory duct is visible and markedly dilated, indicating secondary obstruction causing atrophy,

as opposed to primary atresia of the duct, which is the case in patients with Zinner syndrome.

In most cases, the presentation includes recurrent urinary tract infections, bladder outlet obstruction, or renal failure, leading to an earlier diagnosis (10). In this case, the patient presented with anejaculation, which was an important issue for family planning. Given the nonfunctioning left kidney and the risk of intervention, a conservative management plan was chosen for this patient. The patient was referred for in-vitro fertilization, with surgical sperm retrieval via microsurgical testicular sperm extraction.

In summary, a diagnosis of cecoureterocele was possible based on the constellation of MRI and CT findings, although the presentation of this entity in a man with anejaculation is rare.

Disclosures of Conflicts of Interest: **M.C.** disclosed no relevant relationships. **T.B.** disclosed no relevant relationships. **N.T.** Activities related to the present article: disclosed no relevant relationships. Activities not related to the present article: is a consultant to Coloplast and GTU Urological; gave lectures for and was reimbursed for travel expenses by Astellas. Other relationships: disclosed no relevant relationships. **O.W.** disclosed no relevant relationships.

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Congratulations to the 26 individuals who submitted the most likely diagnosis (cecureterocoele manifesting in a man with infertility) for Diagnosis Please, Case 267. The names and locations of the individuals, as submitted, are as follows:

Individual responses

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