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Trauma and reconstruction

An unusual variant of Zinner syndrome with ureteral ectopia from an atrophied supernumerary kidney

Meghan Cooper, DO *, Lucas Wiegand, MD

University of South Florida, Tampa, FL, USA

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ABSTRACT

This case presents an unusual variation of Zinner syndrome with a patient found to have an ectopic ureteral insertion of the seminal vesicle from a supernumerary pelvic kidney and complete duplication of the ipsilateral, otherwise normal renal unit. The case offers an interesting variant of a previously established syndrome with successful treatment involving robotic resection of an ectopic ureter and seminal vesiculectomy, resulting in resolution of the patient's symptoms.

Introduction

Zinner syndrome was first described in 1914 as a congenital Wolffian duct abnormality including renal agenesis or dysgenesis, ipsilateral seminal vesicle cyst and ejaculatory duct obstruction.[1] Presentation is usually in the third or fourth decade with symptoms including dysuria, perineal pain, painful ejaculation and infertility.[2] Depending on the severity of the symptoms patients can often be managed conservatively, particularly if it was an incidental finding on imaging. However, if significant symptoms are present, then surgical intervention is required, often with laparoscopic modalities being the more favored approach.[3] This case presents a unique variation of Zinner syndrome with ureteral ectopia into the seminal vesicle from a supernumerary kidney that was successfully managed with robotic-assisted laparoscopic ectopic ureterectomy, as well as right seminal vesiculectomy.

Case presentation

A 55-year-old Caucasian male presented to the emergency department with a two-day history of right lower quadrant and flank pain radiating to his groin. He also reported dysuria, diarrhea and decreased oral intake with one episode of non-bilious emesis just prior to presentation. He denied hematuria, urgency, frequency, hesitancy, fevers, or chills. He had a history of Type 2 diabetes and erectile dysfunction but reported no other medical problems. His surgical history included a right inguinal hernia repair. He was a never smoker and used alcohol occasionally. Family history was non-contributory. His current medications included 81mg aspirin, metformin and sildenafil. On exam, the patient was a well-developed middle-aged male with normal digital rectal exam and a normal genitourinary exam. Complete blood count, comprehensive metabolic panel and urinalysis were unremarkable, with the exception of a mild leukocytosis of 11.9. He was admitted to rule out appendicitis, and computed topography (CT) scan showed normal left collecting system and right-sided duplicated system with the ureters intimately involved with an inflammatory mass-like structure arising from the right prostate-seminal vesicle junction (Figs. 1 and 2). Renal scan was negative for obstruction bilaterally and no function was seen within this mass or at its origin. Given the appearance of the seminal vesicles on CT, magnetic resonance imaging (MRI) of pelvis was then ordered to further evaluate area, which revealed peripherally enhancing tubular blind-ending fluid-filled structure extending from the right hemipelvis to the prostato-vesical junction possibly terminating in the urinary bladder or prostatic urethra (Fig. 3). A prostate specific antigen was also ordered, which was 0.49 ng/mL.

At this point, the differential diagnosis included seminal vesicle abscess or calcified seminal vesicle cyst. The patient was discharged home on a one-month course of ciprofloxacin 500mg twice daily. He followed up one month later with improvement, but not resolution of his pain. Repeat non-contrast CT showed resolution of the inflammation, but the suspected seminal vesicle cyst persisted. He was taken to operating room and underwent cystoscopy, right retrograde pyelogram of upper and lower pole collecting systems, attempted cannulation of ejaculatory duct, transrectal ultrasound aspiration of seminal vesicle fluid and seminal vesicle biopsy, as well as injection of contrast into the right seminal vesicle. The seminal vesiculography revealed remnant ectopic ureter inserting into the right seminal vesicle. Biopsy and fluid aspirate

* Corresponding author. Department of Urology, University of South Florida, Morsani College of Medicine, 2 Tampa General Circle, Tampa, FL, 33606, USA.
E-mail address: Meghan.a.cooper@gmail.com (M. Cooper).

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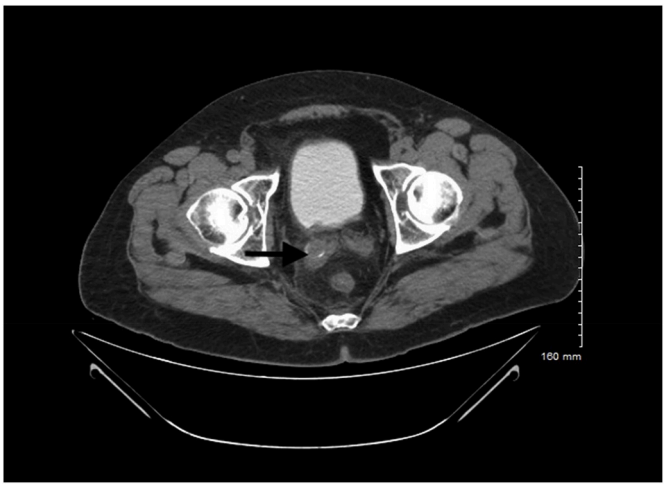

Fig. 1. Delayed axial CT with IV contrast showing right seminal vesicle with inflammatory changes surrounding a mass-like structure (black arrow) arising from the right prostate-seminal vesicle junction.

cytology of the seminal vesicle were both negative for malignancy. Given his significant symptoms, and after discussion of the risks, benefits, and alternatives, the patient agreed to undergo robotic-assisted laparoscopic excision of right seminal vesicle with right ectopic ureter.

The patient was placed in low dorsal lithotomy position. Intraoperative findings included normal cystoscopy except for complete right-sided duplicated collecting system with two right ureteral orifices noted. Placement of two double J stents into the right collecting system was performed prior to proceeding with robotic portion of the procedure. Once beginning our pelvic dissection we noted an ectopic ureter extending supero-laterally from the right seminal vesicle, which was dissected proximally to a blind ending. We proceeded to remove these structures en bloc without complication, carefully excising the ectopic ureter away from the duplicated ureters associated with the normal renal unit, essentially within a common sheath. He was discharged home on postoperative day one and at follow up had no further pain. Pathology was negative for malignancy and his stents were removed 4 weeks post operatively. Follow up renal scan was performed a year later, which again showed no obstruction. At most recent follow up one year after surgery, he continued to do well with no residual pain or symptoms.

Discussion

The prevalence of congenital genitourinary abnormalities is estimated to be less than 1%.[4] Although these anomalies are relatively rare and often subclinical in nature, they can initially present incidentally during a workup for other pathologies. Patients are often diagnosed via ultrasound during a workup for urinary tract infection, epididymitis or flank pain in prepubertal males or the symptoms often present as prostatitis, pain on ejaculation or infertility in older males.

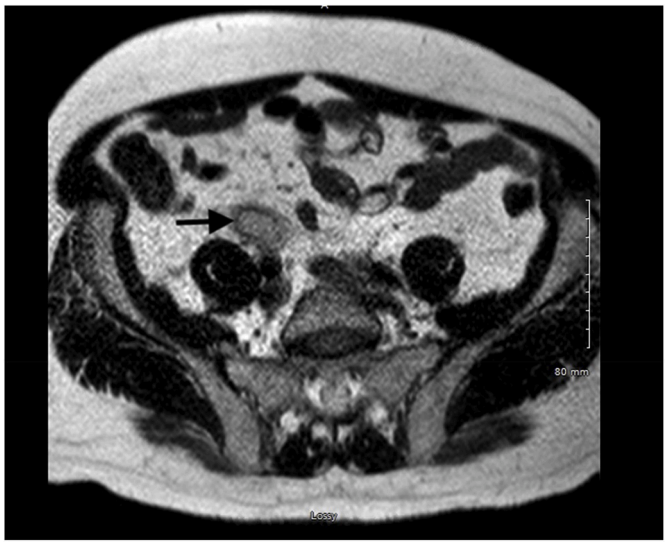

Fig. 3. Pelvic MRI showing peripherally enhancing tubular blind-ending fluid-filled structure (black arrow) extending from the right hemi pelvis to the prostato-vesical junction possibly terminating in the urinary bladder or prostatic urethra.

Ectopic ureters with ureterovasal fusion are uncommon and often associated with ipsilateral renal agenesis. In our current case, there was noted to be an ectopic ureteral insertion into the Wolffian duct structure in the setting of a supernumerary kidney. To date there are no cases in the literature with this interesting variant of Zinner's syndrome.

Zinner's syndrome, which is a rare constellation of congenital findings including seminal vesicle cyst, ejaculatory duct obstruction and ipsilateral renal agenesis was first described in 1914. It occurs with anomalous growth of the Wolffian duct between the 4th and 13th week of gestation and results in the aberrant embryological development of the kidney and ejaculatory duct. This syndrome is considered the male counterpart to Mayer-Rokitansky-Kuster-Hauser syndrome found in females. It is often diagnosed with the onset of sexual activity as that is when the urinary and ejaculatory symptoms usually present.[5]

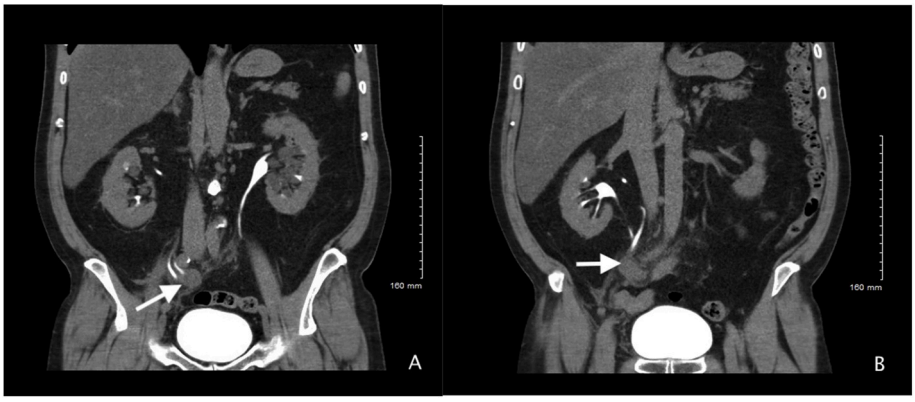

Fig. 2. A & 2B: Delayed coronal CT (A) showing normal left collecting system and relationship of distal duplicated right ureters in relation to blind ending ectopic ureter (white arrow) arising from seminal vesicle. (B) showing proximal duplicated ureters in relation to ectopic ureter (white arrow).

Conclusion

This case showcases a unique presentation of Zinner syndrome with an ectopic ureter originating from an atrophic supernumerary kidney and inserting into the right seminal vesicle. The successful robotic approach used in this case allowed us to meticulously dissect the area without risk to the duplicated ureters traveling to the normal ipsilateral kidney.

Author contributions

Meghan Cooper: Data curation; Formal analysis; Investigation; Roles/Writing – original draft; Visualization.

Lucas Wiegand: Conceptualization; Supervision; Writing – review & editing; Project administration.

Declaration of competing interest

None.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.eucr.2020.101160.

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