Ejaculatory Duct Obstruction in the Setting of an Ectopic Ureter

Joseph Shawn Smith, MD,1 and William Patrick Springhart, MD1,2

ABSTRACT

Introduction: A 17-year-old man was referred to a tertiary care urologist for hematuria, hematospermia, and testalgia. The patient had a history of right-sided renal dysgenesis at birth.

Methods: Fluoroscopic investigation with retrograde seminal vesiculography demonstrated the right ureter implanted ectopically into the right ejaculatory duct. Magnetic resonance imaging of the pelvis showed the ectopic ureter present proximally from the level of the external iliac vessels and implanting into a cystic dilatation on the posterolateral aspect of the prostate. Robot-assisted laparoscopic nephroureterectomy was performed finding an ectopic right ureter forming a confluent cyst with the right ejaculatory duct composed of the seminal vesicle (SV) duct, ureter, and ductus deferens. Incision of the SV revealed mucin balls obstructing the ejaculatory duct.

Results: The nephroureterectomy and removal of the mucin balls resolved the patient’s pain at the 1-month follow-up.

Conclusion: Painful ejaculation or hematospermia in the presence of known renal dysgenesis may constitute Zinner’s syndrome and be an indication for pelvic magnetic resonance imaging or retrograde seminal vesiculography investigation. The classic presentation of pain with contraction of hollow organs and associated proximal dilation on imaging can be applied to the ejaculatory duct. Robot-assisted laparoscopic nephroureterectomy with preservation of the effected SV was a suitable surgical approach for both diagnostic and therapeutic purposes in this case of Zinner’s syndrome. Joseph Shawn Smith, William Patrick Springhart. Ejaculatory Duct Obstruction in the Setting of an Ectopic Ureter. Sex Med 2020;8:574—576.

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Key Words: Hematospermia; Nephroureterectomy; Seminal Vesicles; Ureter

INTRODUCTION

Hematospermia is an unusual complaint comprising only 1%—1.5% of all urologic referrals that deserves appropriate investigation especially outside of clear traumatic incidents.1 In patients older than 40 years, it is usually related to self-limited inflammation. However, the causes of hematospermia are numerous with obstructive causes including calculi, post-inflammatory seminal vesicle (SV) cysts, SV diverticula, urethral strictures, utricular cysts, SV tumors, and benign prostatic hyperplasia.2 Painful ejaculation is a much more common complaint than hematospermia with an even broader differential. The investigation into dysejaculation alone rarely reaps a successful diagnosis.3 The forming ureter can implant ectopically anywhere along the Wolfian duct system where it originates. Ureteric bud insertion into an inappropriate location will likely not find a supportive mesenchyme resulting in renal dysgenesis and 10—15% having associated genital tract malformations.4 The lack of apoptosis of the common nephric duct will also maintain patency of the Wolfian duct.5

CASE

The patient was a Caucasian man who presented to a tertiary referral center in the United States at 17 years of age complaining of hematuria, hematospermia, and testicular discomfort. He had a history of right-sided dysplastic kidney at birth, and a renal ultrasound at 3 years of age confirmed absence of the right kidney on prior imaging. His physical examination showed no causes of his symptoms. He was placed on empiric antibiotics for possible infection, and his testalgia and hematuria resolved. However, he was still experiencing hematospermia and painful ejaculations. The patient then underwent cystourethroscopy with
cannulation of the ejaculatory duct, retrograde seminal vesiculography (Figure 1), retrograde ductusogram, and left retrograde pyelogram. Pelvic magnetic resonance imaging was then performed, which demonstrated the cystic collection in the posterolateral aspect of the prostate but unable to differentiate exactly the course and position of the ureter, ductus deferens, and SV duct (Figure 2). No other causes of the hematospermia were found. At this point, his symptoms were intermittent and not bothersome, so the decision was made to delay surgical treatment until he finished college.

He presented again at 22 years of age with continued mild, intermittently painful ejaculations and hematospermia. Before transitioning to surgery, he underwent complete semen analysis to investigate existing infertility and to bank his sperm in the event of an iatrogenic injury causing infertility. The evaluation showed normospermia, but the results were confounded by the large amount of blood present in the sample. The patient then underwent robotic-assisted laparoscopic nephroureterectomy. The ureter was identified at the level of the iliac vessel bifurcation and dissected proximally until the non-functional renal structure was identified. The ureter was then dissected distally tracking medially to the medial umbilical ligament. The right ductus deferens was identified and noted to be abnormally tortuous; the right SV was noted to be in the orthotopic position. The dissection of the ureter was carried until its insertion came into question as possibly posterior to the prostate. Concern for disrupting the external urethral sphincter dictated that the ureteral dissection had been carried to the furthest point possible. The ureter was clamped proximally and transected distally. The transection revealed the dilated cavity through the ureteral stump and was examined closer. Dark masses were noted to be present within this cavity and were extracted freely with graspers (Figure 3). The duct of the SV was identified within the cavity and cannulated with a 4 Fr. open-ended catheter to ensure patency. The catheter was removed, and the confluence was sutured closed tightly in a running fashion.

Final pathology of the kidney showed diffuse cystic dysplasia and no signs of malignancy. The ureter only had signs of chronic inflammation. The masses grossly resembled stones and were sent for stone analysis which showed 100% mucin composition with no crystal nidus.

DISCUSSION

The constellation of ipsilateral renal agenesis, SV cyst, and ipsilateral ejaculatory duct obstruction is a rare congenital abnormality of the mesonephric duct referred to as Zinner’s syndrome that has been recorded in approximately 100 cases. The patient presented with painful hematospermia consistent with complaints associated with Zinner’s syndrome. The entrapment of mucinous secretions has also been described previously as seen here. Various imaging modalities revealed a patent ejaculatory tract with proximal dilation in this patient. The use of retrograde seminal vesiculography via cannulation showed that this technique is a valuable imaging modality that can assess patency of the ejaculatory duct,
ductus deferens, and examine for filling defects or communications without risking possible scarring of the ductus deferens with antegrade vasography, especially in young men. Carbone et al reported 2 similar cases of renal dysplasia with associated ectopic ureter implantation into the Wolffian tract. Contrary to our case, magnetic resonance imaging was able to determine the communication of the ureter with the SV. Such a communication was not found in our patient but was seen during vesiculography at the time of endoscopic evaluation. Various treatments have been suggested for the patient with symptomatic Zinner syndrome. One of the cases reported by Carbone et al demonstrated a potential concern that was discussed both preoperatively with the patient and intraoperatively: implantation of the ectopic ureter possibly occurring caudally in the prostate. Implantation at this site may prohibit excision owing to risk of urinary incontinence. If excision is not possible, the SV cyst can be treated with transrectal aspiration or transurethral unroofing without significant recurrence. Considering the exact implantation of the ureter will be important for future clinicians offering operative management for anatomic aberrations such as these. In all, robotic-assisted laparoscopic surgical management has become the preferred treatment option for these rare patients with positive outcomes. In this case, the presence of the ectopic ureter added to the volume of the SV cyst and allowed for easier operative technique within the cyst. In cases without this additional volume, it would be more difficult to relieve duct obstruction and ensure recapitulation of the orthotopic anatomy to attempt fertility preservation. Our decision to incise the SV cyst, remove the obstructing mucin balls, and restore the SV appears to be a less explored technique than complete excision of the SV or the SV cyst. Future research can be carried out into developing such preservative techniques and then following up these patients for recurrence of symptoms and successful conception.

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REFERENCES

1. Mathers MJ, Degener S, Sperling H, et al. Hematospermia—a symptom with many possible causes. Dtsch Arztebl Int 2017;114:186-191.

2. Kumar P, Kapoor S, Nargund V. Haematospermia - a systematic review. Ann R Coll Surg Engl 2006;88:339-342.

3. Parnham A, Serefoglu EC. Retrograde ejaculation, painful ejaculation and hematospermia. Transl Androl Urol 2016;5:592-601.

4. Kowalczyk K, Baka-Ostrowska M, Felberg K, et al. Unilateral renal dysplasia associated with ectopic ureter opening into ipsilateral ejaculatory duct. Cent Eur J Urol 2009;62.

5. Uetani N, Bouchard M. Plumbing in the embryo: developmental defects of the urinary tracts. Clin Genet 2009 Apr;75:307-317.

6. Mehra S, Ranjan R, Garga UC. Zinner syndrome—a rare developmental anomaly of the mesonephric duct diagnosed on magnetic resonance imaging. Radiol Case Rep 2016;11:313-317; Published 2016 May 24.

7. Carbone A, Palleschi G, Tomiselli G, et al. Renal aplastic dysplasia and ipsilateral ectopic ureter obstructing the seminal via: a possible cause of male infertility; Part 2. Eur Urol 2007;52:600-601.

8. Kiremit MC, Acar O, Sag AA, et al. Minimally Invasive management of Zinner’s syndrome with Same-Session Robotic-assisted seminal Vesiculectomy and ipsilateral nephroureterectomy using a Single Geometry of Trocars. J Endourol Case Rep 2018;4:186-189.

9. Scarcia M, Maselli FP, Cardo G, et al. Robot-assisted excision of seminal vesicle cyst associated with ipsilateral renal agenesis. Arch Ital Urol Androl 2016;87:325-326.