Robot-Assisted Laparoscopic Resection of the Mesonephric Duct Remnant in a Patient with Zinner Syndrome

Elizabeth P. Kwenda, BS,¹ Rachel A. Locke, BS,¹ Jeremy S. Archer, MD,¹ Li-Ming Su, MD,¹
Archana Shenoy, MD,² Romano T. DeMarco, MD,¹ and Christopher E. Bayne, MD¹

Abstract

Introduction: A 17-year-old male with Zinner syndrome, a right seminal vesicle cyst, and a solitary left kidney presented with chronic pelvic pain. Previous surgeons had attempted robot-assisted laparoscopic seminal vesicle cyst aspiration and transurethral resection of the ejaculatory duct. Neither surgery provided sustained symptom relief. Abdominal and pelvic MRI showed a cystic structure lodged between the prostate and bladder. The right seminal vesicle, kidney, and ureter were not observed.

Materials and Methods: A robot-assisted laparoscopic seminal vesiculectomy was planned. Dissection distal to the right vas deferens and between the bladder neck and prostate revealed a cystic seminal vesicle-like structure. Attached to this was a tubular structure coursing deep to the vas deferens from the right renal fossa. This was presumed to be a dysplastic ureter. The dysplastic ureter was transected from the seminal vesicle and the seminal vesicle was marsupialized to the deep pelvis. Proximally, the dysplastic ureter was transected and left open.

Results: Histologic assessment of the specimen revealed an ~12.1 cm tubular mesonephric remnant. The postoperative course was uncomplicated. At 6 months follow-up, the patient remains free of symptoms with preserved ejaculatory volume.

Conclusions: Mesonephric duct abnormalities and symptoms present on a spectrum. We present a safe and effective resection of a mesonephric duct remnant from a 17-year-old male with Zinner syndrome. A robotic approach localized to the right allowed for excellent observation without compromising left-sided genitourinary anatomy. In males presenting with renal agenesis and pelvic symptoms, clinicians should be suspicious of Zinner syndrome and other mesonephric abnormalities.

Keywords: Zinner syndrome, mesonephric remnant, pediatric urology, robot assisted

Introduction

ZINNER SYNDROME is a rare mesonephric duct abnormality whose phenotypic triad consists of unilateral renal agenesis, seminal vesicle malformation, and ejaculatory duct obstruction. Diagnosis is often made in older adolescents, usually by suspicious clinical history and confirmatory imaging.¹ This case describes a unique resection of a mesonephric duct remnant in a patient with Zinner syndrome.

Case Report

A 17-year-old male with Zinner syndrome presented to the pediatric urology clinic for chronic pelvic pain that worsened

with urination and ejaculation. Previous surgeons had attempted to alleviate the patient's symptoms with a robot-assisted laparoscopic seminal vesicle cyst aspiration followed by transurethral resection of the ejaculatory duct 1 year later. The operative report for the seminal vesicle cyst aspiration stated the surgeons followed the right vas deferens to a dilated structure low within the pelvis and performed laparoscopic needle aspiration of the contents. Neither of the previous surgical interventions provided substantial or sustained relief of his pelvic pain.

Vital signs and physical examination were unremarkable on initial clinic visit. The patient reported pain with urination and ejaculation as well as urinary urgency, frequency,

Departments of ¹Urology and ²Pathology, University of Florida College of Medicine, Gainesville, Florida, USA.

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and straining. Pain was elicited with depression of the prostate on rectal examination. Pelvic MRI of the pelvis with gadolinium revealed a cystic structure confined in the deep pelvis between the bladder and prostate. A right seminal vesicle was not observed on imaging, but the left seminal vesicle was identified in the correct position. The findings are depicted in Figure 1. Abdominal MRI showed absence of right kidney and ureter. Interventional radiology at our institution did not feel the cystic structure was accessible from a percutaneous approach.

The patient and family were counseled on treatment options and elected for robot-assisted laparoscopic right seminal vesiculectomy. Preoperative counseling focused on the risks of erectile and ejaculatory dysfunction along with bladder neck injury associated with the procedure. Preoperative semen analysis was normal according to WHO 2010 criteria.

The patient was aligned in a supine position with arms secured and legs gently flexed in a steep Trendelenburg position. Trocar placement was similar to a transperitoneal robot-assisted laparoscopic prostatectomy. A 12 mm robotic trocar was placed at the umbilicus for the robotic camera. Secondary trocars were placed in an inverted U-shaped configuration with an additional 12 mm assistant port in the right lower quadrant. The surgical dissection began with a peritoneal incision from the right pelvic sidewall to the midline overlying the presumed location of the right seminal vesicle. The dissection was not extended beyond the midline because of an effort to preserve the left vas deferens, seminal vesicle, and neurovascular bundle. The right vas deferens was identified and tracked to a dilated ampulla of the vas deferens; however, a right seminal vesicle was not initially identified. Using preoperative MRI findings, we decided to dissect deeper into the pelvis, beyond the ampulla. Distal dissection revealed a structure separate from the vas deferens that appeared to be a right seminal vesicle cyst with an attached tubular structure. The tubular structure became increasingly atretic as it ran directly beneath the vas deferens and high into the retroperitoneum, where it coursed medial to the ascending colon. Intraoperative findings are shown in Figure 2. Proximally, we transected the structure with cold scissors and left it open. Distally, at the point where the structure

FIG. 1. MRI scan demonstrating patient's cystic mesonephric duct remnant.

FIG. 2. Operative image. The vas deferens is located inferior to the right arm (white asterisk). The left arm is showing the mesonephric duct remnant (yellow arrow).

entered the seminal vesicle cyst, we transected the cyst and left it marsupialized to the deep pelvis. The specimen was secured in a laparoscopic bag and removed through the umbilical trocar.

The resected specimen was 12.1 cm in length. Histologically, the proximal aspect was solid and demonstrated mesonephric remnants and thick-walled vessels. The middle aspect showed fibromuscular tubular tissue with rudimentary lumina lined by pseudostratified ciliated epithelium. Distally, the lumen was relatively dilated and lined by cuboidal epithelium and muscle. Given the pathologic findings, duplication of vas deferens was a diagnostic consideration. Ectopic ureteral tissue could not be diagnosed with confidence based on the histology alone, given the lack of transitional epithelium (urothelium); however, a dysplastic ureter could not be excluded. The pathologic and histologic sections are shown in frame A–D of Figure 3.

The postoperative course was uneventful. The patient reported unchanged ejaculation sensation, volume, and velocity, so a formal repeat semen analysis was not collected. All pain symptoms were resolved with preserved ejaculatory volume at 6-month follow-up.

Conclusions

Zinner syndrome is a rare triad of unilateral renal agenesis, ejaculatory duct obstruction, and seminal vesicle cysts.^{1,2} Current hypotheses regarding the pathogenesis of Zinner syndrome revolve around the maldevelopment of the mesonephric duct or Wolffian duct. In normal embryologic development, the mesonephric duct performs some rudimentary kidney function. It persists after the rest of the mesonephros has regressed and opens to the cloaca. In a process called reciprocal induction, the metanephric blastema secretes growth factors to induce growth of the ureteral bud from the caudal portion of the mesonephric duct. Once formed, the ureteral bud produces growth factors that stimulate differentiation of the metanephric blastema into the future renal glomeruli and tubules. Concurrently, the presence of testosterone

FIG. 3. Clockwise from *top left* (A) Pathologic segment of the mesonephric duct remnant. (B) Histologic section from the proximal aspect of the remnant, demonstrating mesonephric remnants and thick-walled segments. (C) Histologic section from the middle aspect demonstrated fibromuscular tubular tissue with a rudimentary lumen lined by pseudostratified ciliated epithelium. (D) Histologic section of the distal portion of the remnant was lined by cuboidal epithelium and muscle.

in males stimulates the distal end of the mesonephric duct to differentiate into the epididymis, vas deferens, seminal vesicles, and trigone of the bladder. Failure of signaling pathways during the development of the distal mesonephric duct can result in agenesis or dysplasia of ureteral bud (and subsequently the ipsilateral kidney) or genital adnexal structures (such as the ipsilateral seminal vesicle). Our patient presented with right renal agenesis and a right seminal vesicle cyst with histologic findings consistent with mesonephric remnants, confirming the diagnosis of Zinner syndrome.

Zinner syndrome was first reported in 1914.³ Some patients remain asymptomatic and are diagnosed incidentally, whereas the remainder of patients typically present with pain during adolescent and adulthood. Although nonspecific, the most common complaints are dysuria and ejaculatory difficulty related to the compressive effect of the seminal vesicle cysts. Other commonly reported symptoms include abdominal and pelvic pain, urinary tract infections, and urinary frequency.²

Smaller cysts in Zinner syndrome are generally asymptomatic and can be managed expectantly. For symptomatic patients, a conservative nonsurgical approach is usually taken, although larger cysts or more severe symptoms may require laparoscopic and robotic exploration/excision of the cystic lesion.² Possible complications of surgical

intervention include erectile and ejaculatory dysfunction as well as infertility.

Sundar and Sundar reported a case of a 25-year-old male who presented with urinary frequency. A CT with delayed contrast showed an empty left renal fossa and nonexcreting left ureter. Transrectal ultrasound guided imaging suggested a remnant left ureter draining to a left seminal vesicle. MRI confirmed a communication between the left ureter remnant and a seminal vesicle cyst. Laparoscopic excision of the cyst was effectively undertaken with no symptoms reported on 1-year clinical follow-up.²

In a pooled analysis of 52 cases of seminal vesicle cysts associated with ipsilateral renal agenesis, van Den Ouden et al. noted ureteral remnants in 14 patients. Open exploration provided relief in all symptomatic patients. Of note, cystic aspiration had a lower 30% success rate compared with transurethral derofing, which provided symptomatic relief in 75% of patients. The authors subsequently recommended cystic aspiration for diagnostic purposes only.⁴ Other studies also noted minimal therapeutic effects and recurrence when aspiration of the cystic seminal vesicle was used for management. Our patient most likely fell in the same category of patients who reported no symptomatic relief after seminal vesicle cystic aspiration because of the presence of the previously unidentified mesonephric duct remnant, which enabled fluid reaccumulation and

cyst persistence. Accordingly, we urge clinicians to consider the possibility of Zinner syndrome and exclude the presence of mesonephric duct remnants in patients with persistent pelvic symptoms and renal agenesis.

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Address correspondence to:

Christopher E. Bayne, MD

Department of Urology

University of Florida College of Medicine

1600 SW Archer Rd

PO Box 100247

Gainesville, FL 32610-0247

USA

E-mail: christopher.bayne@urology.ufl.edu

Abbreviations Used

CT = computed tomography

MRI = magnetic resonance imaging

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