

Novel application of da Vinci robotic system in patients of Zinners syndrome – case report and review of literature

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ALLAPARTHI S, BLUTE RD JR. Novel application of da Vinci robotic system in patients of Zinners syndrome – case report and review of literature. *The Canadian Journal of Urology*. 2010;17(2):5109-5113.

Seminal vesicle cysts combined with ipsilateral renal agenesis, ectopic ureter and giant right ampullary cyst of vas deferens represent a rare urological anomaly, Zinners syndrome. In symptomatic patients' seminal vesiculectomy along with enbloc excision of the ipsilateral ampullary cyst, ectopic ureter and dysplastic, renal tissue

is the preferred treatment option. We report robotic assisted removal of a large seminal vesicle cyst with ipsilateral renal agenesis, ectopic ureter and a giant right ampullary cyst of vas deferens in a 34-year-old male. We reviewed the literature about this rare urological anomaly and novel usage of da Vinci surgical robotic surgical system (DSRS) (Intuitive Surgical, Sunnyvale, California) in performing this procedure.

Key Words: seminal vesicle cyst, Zinners syndrome, da Vinci robotic system

Introduction

In 1914 Zinner first reported a rare urological anomaly, association of a seminal vesicle cyst with ipsilateral renal agenesis.¹ Association of this anomaly with ectopic ureter and ampullary cyst of vasdeferens was even rarer. A literature review showed many reports of this developmental anomaly, along with a pooled case analysis of 55 cases by Van den Ouden et al.²⁻⁵ In symptomatic patients, surgical excision is the treatment of choice. In the past two decades, the management has shifted from open surgical to laparoscopic and very recently even robot-assisted removal was performed. Experience with laparoscopic excision was also limited to series of case reports and two series of three cases each in high volume institutions due to rarity of the condition.⁶⁻⁹ Robotic assisted laparoscopic approach has been reported in three case reports; two adult and one pediatric with successful relief of symptoms and minimal morbidity.¹⁰⁻¹²

We report one additional case of symptomatic seminal vesicle cyst with ipsilateral renal agenesis, ectopic ureter and a giant right ampullary cyst of vas deferens in a 34-year-old male, which was successfully managed by robotic assisted surgical excision. Based on a review of the existing literature en bloc excision of the ipsilateral seminal vesicle, cyst, ectopic ureter and dysplastic renal tissue, is the treatment of choice in symptomatic patients.

Case presentation and management

A 34-year-old man presented with a 1 year history of symptoms of outlet obstruction including decrease in the force of stream, intermittency and straining to void. He appeared to respond to treatment of what was presumed to be a primary bladder neck outlet obstruction with alpha blockers. However, the symptoms worsened with vague perineal pain prompting further work up. A cystoscopy showed unremarkable left ureteral orifice but absent right ureteric orifice. This was followed by, a voiding cystourethrogram, CT scan, MRI of his abdomen, pelvis and perineum. Imaging studies demonstrated a giant (greater than 5 cm) right seminal vesicle cyst with right renal agenesis, ectopic ureteral remnant that emptied into the seminal vesicle and a giant cyst of the

Accepted for publication December 2009

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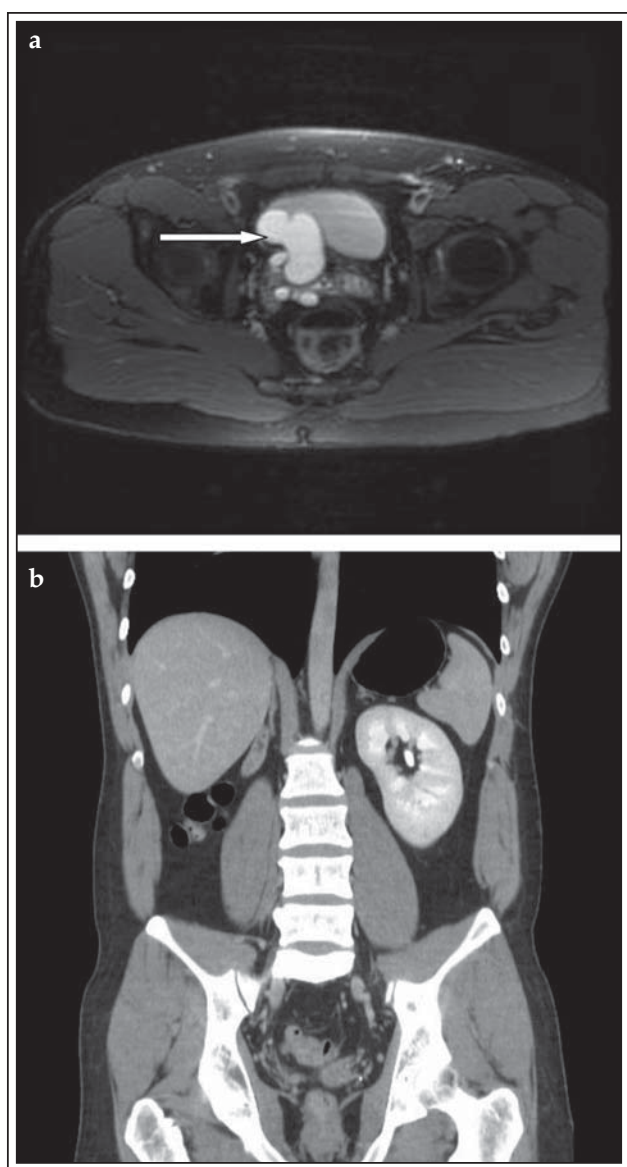


Figure 1. a) MRI, arrow showing right seminal vesicle cyst, b) CT urogram showing absent right kidney.

right ampulla of the vas deferens, Figure 1. The patient had several children and had normal erectile function. Rectal examination demonstrated the palpable cystic masses beneath the right hemitrigone, which elevated the trigone and obstructed the bladder neck. After discussing all the risks and benefits, including novel application of DSRS technology the patient provided fully informed consent for surgery.

Surgical technique

Under general anesthesia, the patient was securely positioned in low lithotomy and steep head down

position with all pressure points padded. A 20-French Foley urethral catheter with 30 mL in balloon was placed in a sterile fashion. The abdomen was insufflated with CO₂ using a Veress needle up to 15 mm Hg prior to inserting the right paraumbilical 12 mm Ethicon port (Ethicon Endosurgery Inc, Cincinnati, OH) for the camera. Under direct vision, two additional 8 mm da Vinci cannulas were placed approximately 8 cm lateral and inferior to the camera port. One additional working 12 mm Versa Step port (One-Step port; U.S. Surgical) was placed in the right lower quadrant about 2 inches medial and above the right anterior superior iliac spine, Figure 2. Da Vinci robot was docked with all ports in place. Anatomical landmarks were identified on entry into the abdomen with the 0° lens. Dissection started in the cul-de-sac identifying and dissecting out the seminal vesicles and vas deferens. Clearly there was no vas visible in the right internal ring and the left vas was easily followed to the cul-de-sac and up to the bladder base. Using this as a guide we opened up the peritoneum covering the right seminal vesicle. After meticulous blunt and sharp dissection we noticed a bulge, the bluish right seminal

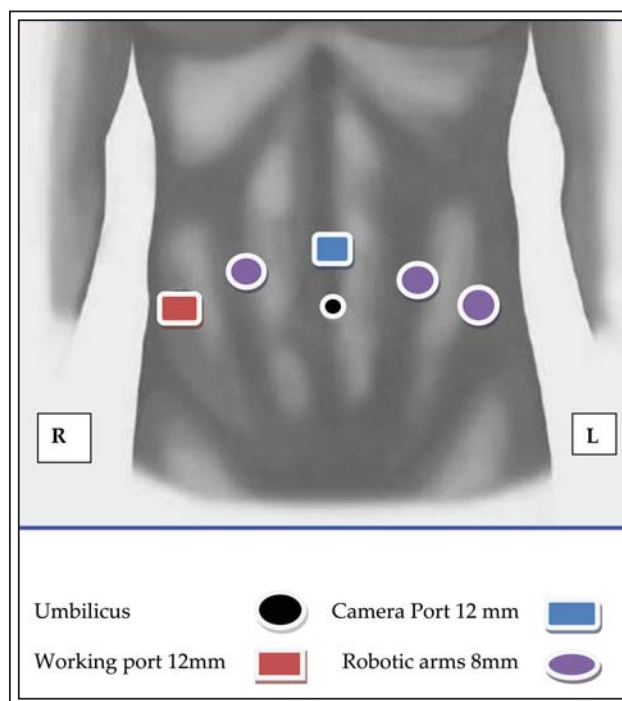


Figure 2. Port placement for robotic assisted removal of a large seminal vesicle cyst with ipsilateral renal agenesis associated with an ectopic ureter and giant ampullary cyst of the vas deferens using da Vinci S robotic system umbilicus camera port 12 mm working port 12 mm robotic arms 8 mm R L.

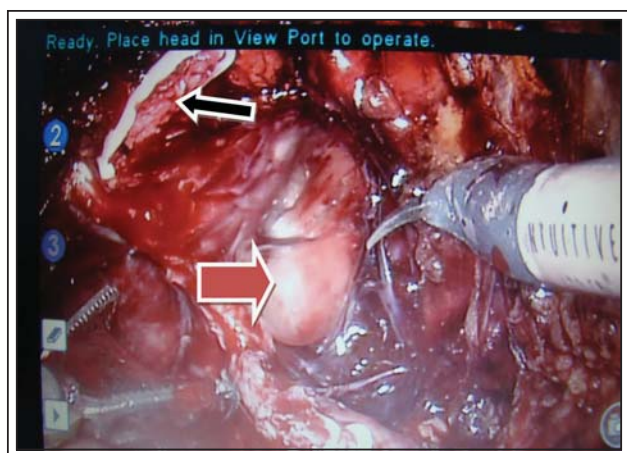


Figure 3. Intraoperative picture showing bluish cystic seminal vesicle (red arrow) and weck clip on ureteral remnant draining into seminal vesicle (black arrow).

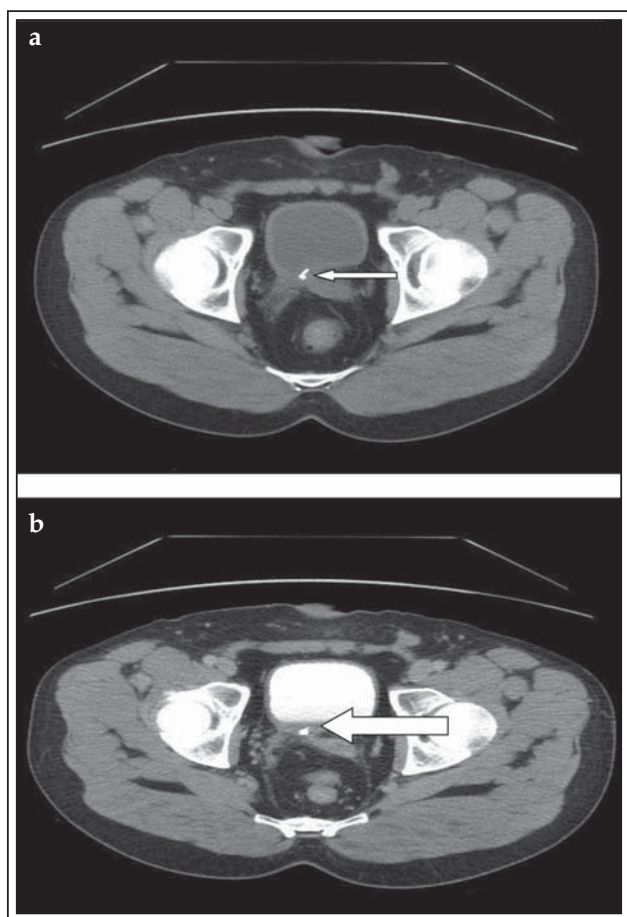


Figure 4. a) Post operative CT urogram arrow showing absent cyst with clip in place, b) in contrast phase with no extravasation.

vesicle cyst and then very shortly thereafter, visualized the centimeter ureteral remnant draining into the cyst that terminated at the level of the pelvic brim, Figure: 3. Careful dissection was done and ureter transected with large Weck (Weck Surgical Instruments, Teleflex Medical, Durham, NC) clips and dissected down to the cyst. En bloc mobilization of the specimen was possible and the insertion of the seminal vesicle in the prostate was sectioned last and over sewn with a 2-0 Vicryl suture. Specimen was removed intact through the camera port by using a retrieval bag and the peritoneal incision was sutured leaving a suction drain. The total operative time was 150 minutes with minimal blood loss of less than 25 mL. Patient was discharged on postoperative day two. The pathology was suggestive of benign seminal vesicle cyst with a portion of a tubular structure with muscular wall and cuboidal cell lining, possibly a Wolffian duct remnant. Patient had uneventful recovery with a follow up check cystoscopy in 8 weeks and repeat CT urogram 6 months after the procedure as shown in Figure 4.

Discussion

This rare urological anomaly embryogenesis is based on the intimate relationship of the urinary and reproductive systems. During 4-7 weeks of embryonic life the ureter develops as an outgrowth of the mesonephric (Wolffian) duct from the caudal end. Kidney develops when ureteric bud meets the metanephric blastema. Differential growth of the mesonephric duct and urogenital sinus explains the establishment of the ureteral orifice in its correct anatomical location on the bladder trigone. If the ureteric bud develops too early, it will be located more cranially in the mesonephric duct, and therefore it will meet the mesonephros, which will atrophy or disappear, resulting in agenesis or dysplasia of the kidney. The prolonged contact of the ureter with the mesonephric duct can result in an ectopic orifice in one of the derivatives of the duct. Additional structures that arise from the mesonephric duct system include the vas deferens, ejaculatory duct and lower two-thirds of the epididymis. Seminal vesicle cyst formation is thought to be due to build up of seminal vesicle fluid due to an abnormal or obliterated ejaculatory duct. The differential diagnosis of cystic pelvic masses in the male includes müllerian duct cysts and ejaculatory duct cysts, both of which are midline in location. The presence of spermatozoa in the aspirate may differentiate seminal vesicle cysts from müllerian duct cysts. Other possibilities include prostatic cysts and, more lateral in location, diverticulosis of the ampulla of the vas deferens, ectopic ureterocele, and abscess.

TABLE 1. Patient characteristics and diagnostic imaging

References	Van den Ouden et al ²	King et al ⁵	Cherullo et al ⁸
No. of patients	52	13	3
Mean age at presentation (range)	30.2 yrs (11-62)	21.9 yrs (5 mos-53 yrs)	35.7 yrs (30-42)
Presenting symptoms (%)	Dysuria (37), frequency (33), perineal pain (29), epididymitis (27), infertility (17), scrotal pain (13)	Abdominal pain (46), irritable LUTS (38), epididymitis/UTI (38), perineal/scrotal pain (23) painful ejaculation (8)	Perineal pain (100), testicular pain (33), painful ejaculation (33), dysuria (67), irritable LUTS (67)
Diagnostic evaluation (%)			
TRUS	27	62	67
IVP	100	92	33
CT	27	62	100
MRI	4	31	33
Vasography	46	16	67

Review of literature, about this rare urological anomaly on patient characteristics, presenting symptoms, physical findings, diagnostic tests and surgical treatment are shown in Table 1. The diagnosis is usually made either in early adulthood or in the third to fifth decades of life. Predominantly, symptoms develop due to irritation of adjacent organs by enlarged and inflamed seminal vesicle cyst. This includes irritative lower urinary tract symptoms (LUTS), perineal or suprapubic pain, postcoital discomfort, hematospermia, prostatitis, epididymitis and issues of fertility. Testicular ectopia and agenesis were also

reported. Initial evaluation of patient with suspected findings include thorough clinical examination, transrectal ultrasonography for evaluating the findings on digital rectal examination. CT and MRI may be useful in further evaluation of other urological abnormalities and pelvic pathology. Cystoscopy may be useful to uncover concurrent bladder pathology, extrinsic compression and ectopic ureteric orifice.

Although open surgery performed through a transvesical, retrovesical, transperineal, or transcoccygeal route has high success rate, its related morbidity was considerable given deep location of the seminal vesicles

TABLE 2. Experience with robotic and laparoscopic excision of seminal vesicle cyst with ipsilateral renal agenesis

References	Patient age (s)	Operative time (min)	Estimated blood loss (mL)	Hospital stay (days)
Carmignani et al ^{16*}	19 y	180	NR	2
Ikari et al ^{17*}	24 y, 10 mo	90, 120	NR	2
Cherullo et al ^{18*}	42 y, 35 y, 30 y	180, 210, 240	350, 300, 250	1
Moore et al ^{11#}	16 y	156	10	1
Selli et al ^{10#}	39 y	180	50	4
Carmack et al ^{12#}	28 y	120 ^a	25	2
Present case	34 y	150	25	2

*Laparoscopic excision of seminal vesicle cyst with ipsilateral renal agenesis

#Robotic assisted excision of seminal vesicle cyst with ipsilateral renal agenesis

^aExcision of seminal vesicle cyst without ipsilateral renal agenesis

in the retrovesical space. Transurethral resection with unroofing has been reserved for smaller cystic lesions in close contact with the prostate, and was associated with a significant failure rate.⁸ However, the large size attained by congenital cysts of the seminal vesicles restricts their surgical removal through the abdominal approach, particularly if associated ipsilateral ureterectomy or nephroureterectomy is required.

Since the late 1990's, laparoscopy has gained acceptance as the modality of choice when surgical excision of these malformations is necessary. Laparoscopy offers the absolute advantages of decreased postoperative pain, lower morbidity, shorter length of hospital stay, and quicker convalescence.^{6,7} However, the experience was limited because of the rarity of this condition. Although technological advances have been made in laparoscopic surgery, conventional laparoscopy is limited by lack of three-dimensional (3-D) vision, poor maneuverability and ergonomic movement of instruments. Robotic technology provides additional dimensions to laparoscopy by adding 3-D visualization, improving the degrees and freedom of movement, improved ergonomics and facilitation of intracorporeal suturing given the fact that seminal vesicles are deeply located in the pelvis.^{13,14}

However, a possible limitation of using robotic approach is difficulty accessing dysplastic kidneys, using the robotic ports centered in the prostate area. Table 2 represents a review of published robotic and laparoscopic data. Results include operative time, hospital stay, and estimated blood loss. Our results are comparable to those reported in the literature.

Conclusion

In conclusion, we report the novel usage, feasibility, technical aspects of DSRS in performing the minimally invasive treatment of the rare congenital cystic malformations of the seminal vesicle associated with ipsilateral ectopic ureter, renal agenesis and large ampullary cyst of vas deferens with minimal morbidity and acceptable outcomes. □

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