Case Report

Robot-Assisted Laparoscopic Approach in a Patient of Zinner Syndrome with Hematuria: A Rare Presentation

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ABSTRACT

Congenital malformations of the seminal vesicles (SVs) are rare and are often associated with abnormalities of the ipsilateral upper tracts as embryologically both the ureteral buds and SVs arise from the mesonephric ducts. The triad of SV cysts, ipsilateral renal agenesis, and ejaculatory duct obstruction is known as the Zinner syndrome. We herein present the case of Zinner syndrome with hematuria as the mode of presentation. A 52-year-old gentleman presented with complaints of gross and painless hematuria for 3 months. An initial ultrasound revealed absent right kidney with a cystic structure abutting the urinary bladder. Cystoscopic examination revealed a high bladder neck. Magnetic resonance imaging of the abdomen revealed an absent right kidney and a large tubular structure in the region of the right ureter extending up to right SV with blood content and stones within. It was distally seen communicating with one of the cysts of the right SV. The cystic structure was removed with a robot-assisted laparoscopic approach. The console time was 110 min with minimal blood loss. Postoperative course was uneventful. Histopathology of the cyst wall revealed chronic inflammation. The patient is doing well on 6 months follow-up. This case was unique in terms of it presenting with a large intra-abdominal cyst with sharp stones within, probably first of its kind to be ever reported. Surgery is mandated for such symptomatic cysts and the daVinci robot with its minimally invasive approach offers the perfect platform for treating such challenging cases.

KEYWORDS: *Excision, hematuria, robotics, syndrome, Zinner*

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INTRODUCTION

Congenital malformations of the seminal vesicles (SVs) are rare and are often associated with abnormalities of the ipsilateral upper tracts as embryologically both the ureteral buds and SVs arise from the mesonephric (Wolffian) ducts. Zinner *et al*.[1] were the first to report the association of SV cysts and ipsilateral renal agenesis in 1914 and subsequently the triad of SV cysts, ipsilateral renal agenesis, and ejaculatory duct obstruction came to be known as the Zinner syndrome. It is considered as the male counterpart of the Mayer–Rokitansky–Kustner–Hauser syndrome seen in females.[2]

Till date, around 200 cases of Zinner syndrome have been reported in the literature.[3] It is a congenital anomaly which is usually asymptomatic throughout the childhood and presents in the third or fourth decade of life. Most of them are discovered incidentally but may present as dysuria, lower urinary tract symptoms, urinary tract infections, painful ejaculation, and even infertility.[4]

To the best of our knowledge, no case of Zinner syndrome reported so far have had hematuria as the initial mode of presentation. We herein present the case of Zinner syndrome presenting with hematuria who was found to have a large SV cyst with stones within and was managed by a robot-assisted laparoscopic approach.

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Case Report

Our index case was a 52-year-old gentleman who presented with the complaints of gross, intermittent, and painless hematuria for 3 months. Digital rectal examination revealed a grade 1 enlarged benign prostate. On ultrasonography (USG), a cystic structure abutting the urinary bladder was seen, and right kidney was not visualized. Urine culture was sterile. Urine cytology was negative for malignant cells.

A computed tomography (CT) scan [Figure 1a and b] revealed right renal agenesis and a tubular elongated structure extending from the right renal fossa to the pelvis 21 cm long and 6 cm wide closely abutting the urinary bladder. The distal end of the lesion was opening into the right SV. Hyperdense calcified structures were seen in the distal portion of the lesion. A cystopanendoscopy was done as a part of work up for hematuria [Figure 2]. Urethra and verumontanum were normal. No abnormal opening or utricle was appreciated on the verumontanum. There was a Grade II enlargement of the prostate and a high bladder neck. Prostatic urethra was deviated to the left. Right ureteric orifice was not visualized, and an indentation was noted on the right lateral wall. Bladder mucosa was healthy. A magnetic resonance (MR) urography [Figure 1c and d] was also performed to better characterize the lesion before taking up the patient for invasive surgery. It revealed the findings of a large cyst with blood content within and an absent right kidney. The lesion was seen communicating distally with one of the cysts of the right SV. Right ejaculatory duct was prominent with its distal end in close proximity with urethra. With a diagnosis of Zinner syndrome in mind, the patient was planned for the excision of the large cyst in view of persisting hematuria. The patient underwent a prior cystopanendoscopy and bladder neck incision in view of high bladder neck. The cyst was removed with a robot-assisted laparoscopic approach. The console time was 110 min with minimal blood loss. Cutting the cyst open revealed hemorrhagic fluid and the diagnosis of stones within the cyst was confirmed. There were multiple stones with spiky configuration [Figure 3]. Histopathology of the cyst wall revealed chronic inflammation. The postoperative course was uneventful, and the patient was discharged on the

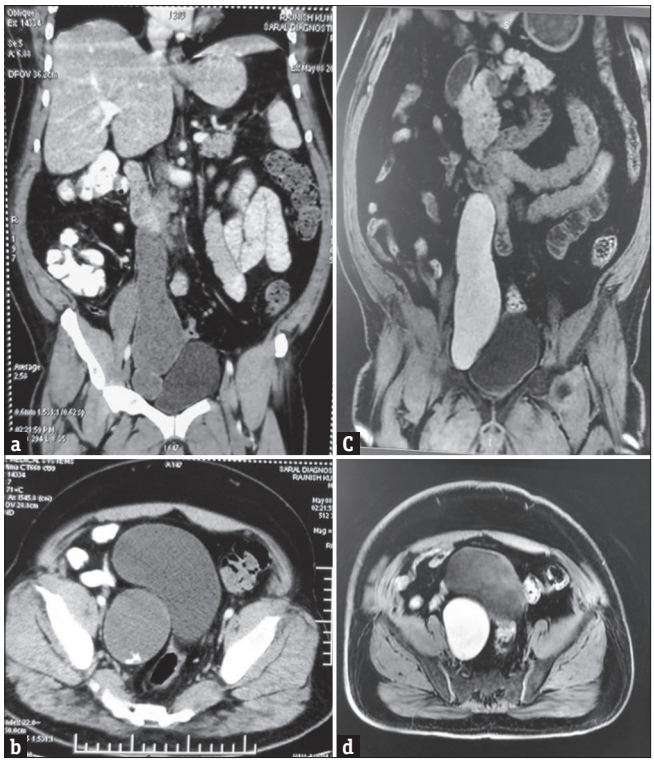

Figure 1: Computed tomography urography (a and b) magnetic resonance urography (c and d) showing a tubular cystic structure with stones within abutting the bladder and right renal agenesis

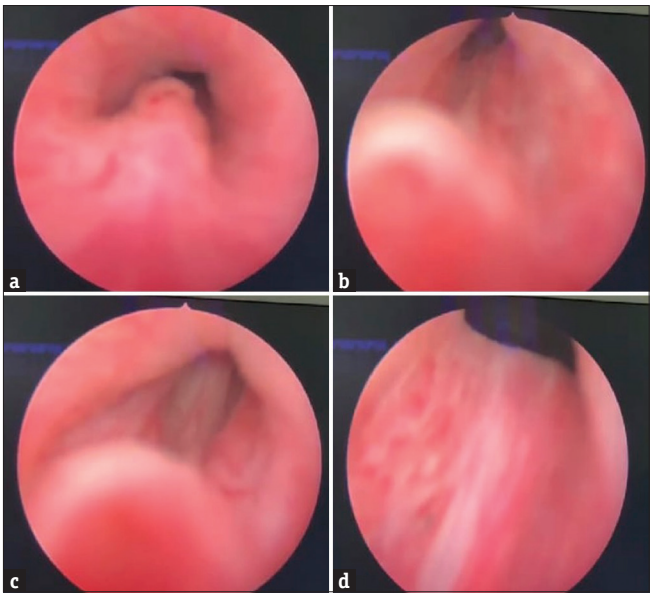

Figure 2: (a-d) Cystopanendoscopy revealing a normal verumontanum, enlarged prostate, and a high bladder neck

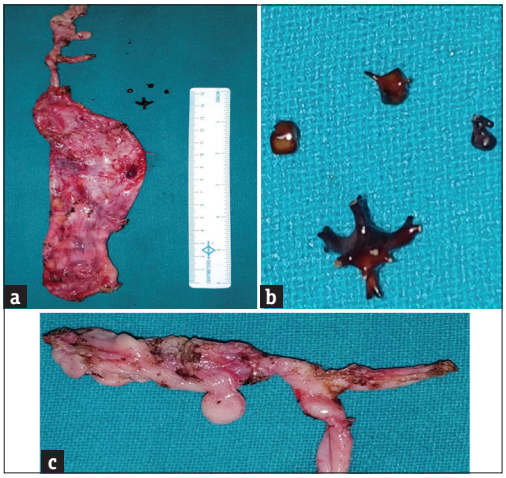

Figure 3: (a-c) Specimen depicting a large cyst with extension into ipsilateral seminal vesicle and stones present within the lower end of the cyst

second day. On 6 months follow-up, patients doing well.

Discussion

Zinner syndrome is a rare congenital anomaly that occurs as the result of disturbance in the development of mesonephric duct before the 7th week of gestation.[5] The distal end of the duct separates from the ureteric bud during 6th–8th week. Under the influence of testosterone and anti-Mullerian hormone, the distal end of the duct forms the hemitrigone, bladder neck, urethra, and organs of the male reproductive tract, namely SVs, vas deferens, ejaculatory ducts, and epididymis.[6] On the other hand, under the influence of growth factors, ureteric bud migrates toward the metanephric blastema to form the primitive kidney.[7]

Mutations in the metanephric blastema, retinoic acid pathway signaling hampers the fusion of the ureteric bud to the metanephros resulting in either hypoplasia or complete agenesis of the kidney. Simultaneously, if the distal end of the mesonephric duct does not grow, it results in the obstruction of the SVs and ejaculatory ducts resulting in the accumulation of secretions and cystic dilatation of SV as well as ejaculatory duct obstruction.[8]

Dysuria, frequency, painful ejaculation, and perineal pain are the most common presenting complaints due to pressure of the SV cyst and are well documented in the earlier reports of Zinner syndrome.[4] Patients might also present later with oligozoospermia and azoospermia leading to infertility.[4] Our case was unique as the patient had no complaints apart from the two episodes of painless gross hematuria. The hematuria can be attributed to the sharp configuration of stones in the cyst along with superimposed possible vesiculitis. The presence of hematuria might also point to a possible partial obstruction of the ejaculatory duct.

Although USG and CT scans are useful, MR is the imaging modality of choice as it would better delineate all the anatomical details in the pelvis that could be missed on a CT scan.[9] The periprostatic location helps its identification as SV cysts. They appear hypo or hyperintense on T1 and hyperintense on T2 sequences. They are usually small in size and rarely grow to a large size. Again our index case of unique as in presented with a large cyst reaching around 15 cm in size extending from the true pelvis to the renal fossa.

Surgical management is recommended for symptomatic patients and various options ranging from aspiration of cyst to cyst excision and various approaches, including transvesical, transperineal, transrectal, transperitoneal, retroperitoneal etc., have been described. All these approaches are limited by them being not versatile as the cysts are situated deep in the pelvis and also difficulty might arise if there are coexisting upper tract anomalies.[10] Thus, minimally invasive techniques such as laparoscopic and robotic excision with their more versatile approach and better postoperative outcomes should become the new standard of care for dealing such complex anomalies.

Conclusion

Zinner syndrome can have a wide variety of presentations, and although rare, can present with intractable hematuria. This case was unique in terms of it presenting with a large intra-abdominal cyst with sharp stones within, probably first of its kind to be ever reported. Surgery is mandated for such symptomatic cysts and the daVinci robot with its minimally invasive approach offers the perfect platform for treating such challenging cases.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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