CASE REPORT
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Is Laparoscopic Approach Adequate for Zinner's Syndrome? One Patient, Two Cases

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Abstract

Zinner syndrome (ZS) was first described by Zinner in 1914. This condition includes unilateral renal agenesis, ipsilateral seminal vesicle cyst, and ejaculatory duct obstruction. ZS treatments ranging from medical drug therapy to laparoscopic interventions have been investigated in the literature. A 21-year-old patient presented with scrotal pain after ejaculation. The diagnosis was Zinner's syndrome, and the patient underwent transperitoneal laparoscopic excision of the left seminal vesicle cyst. After 2 years, transurethral ejaculatory duct resection (TUR-ED) was performed at a single center because of symptomatic dilatation in the seminal vesicles. The patient's 1-year urological follow-up after TUR-ED remained normal. This presentation is a case report of a single patient and two cases that are rare in the literature. Cyst aspiration and seminal cyst excision may be considered as first-line treatment options, but the possibility of recurrence should not be forgotten. Even if seminal cyst excision is performed, it should be kept in mind that TUR-ED may be required in the future.

Keywords: Zinner's syndrome, andrologia, congenital, laparoscopy, ejaculator ductus, case reports

Introduction

Zinner syndrome (ZS) was first described by Zinner (1) in 1914. Unilateral kidney agenesis is a syndrome associated with ipsilateral seminal vesicle cyst and ejaculatory duct obstruction (1). They are usually diagnosed in 3 or 4 decades. Patients may be asymptomatic or have symptoms such as painful ejaculation, urgency, hematuria, tenesmus, chronic pelvic pain, and hematospermia. Infertility is also reported in a significant proportion of cases. Although the definitive treatment of this syndrome is unknown, treatments ranging from medical drug therapy to laparoscopic interventions have been investigated in the literature.

A 21-year-old patient presented with scrotal pain after ejaculation. The diagnosis was Zinner's syndrome, and the patient underwent transperitoneal laparoscopic left seminal vesicle cyst excision. After 2 years, we performed transurethral ejaculatory duct resection (TUR-ED) due to symptomatic dilatation in the seminal vesicles. The patient's 1-year urologic follow-up after TUR-ED was normal. This presentation presents a case report of a single patient and two cases that are rare in the literature.

Case Presentations

Case 1

In 2019, a 21-year-old man was admitted to the emergency department because of severe scrotal pain. It was learned that the patient had postejaculation pain for approximately 3 years and had experienced very severe pain attacks three times. No known history of disease or surgery, smoking or alcohol use. No pathological findings were detected on scrotal and abdominal examination. Scrotal color Doppler ultrasound (USG) and urinalysis findings were normal. Digital rectal examination revealed minimal mass formation in the prostate.

Computed tomography (CT) scan revealed hypertrophy in the right kidney, agenesis in the left kidney, and a low-density, smooth-contoured hypodense lesion, which may belong to a cyst with a diameter of approximately 43 mm, indenting the bladder adjacent to the anterior seminal vesicle on the left (Figure 1).

Radiological evaluation was completed using abdominal magnetic resonance imaging (MRI). On MRI, a 44x31 mm cystic structure was observed anterior to the seminal vesicles on the

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left, indenting into the bladder and with hyperintense content on T1A (Figure 2).

The low ejaculation volume in the spermiogram suggested obstruction of the ejaculatory duct. Other parameters in sperm analysis were observed naturally (semen volume 1 mL, sperm concentration 24.7 million/mL, progressive motility 43%, Kruger 9%).

According to the patient's current imaging and laboratory findings, Zinner's syndrome was diagnosed. Laparoscopic left seminal vesicle cyst excision was performed. There were no complications during the operation. Histopathology of the cyst resulted as "seminal vesicle cyst". The patient's complaints did not recur during the 2-year postoperative follow-up.

Case 2

The same patient was re-applied 2 years later. The patient reported pain in the perineal area that worsened after ejaculation. During digital rectal examination, a fluctuating lesion, approximately 3 cm in diameter, was palpated on the left side at the base of the prostate. Urine analysis was normal.

In contrast-enhanced pelvic magnetic resonance imaging, the seminal vesicles and ejaculator duct were dilated on the left. A cystic structure with hyperintense content was observed on T1A, which was believed to indicate hemorrhage. No pathology was detected at the verumontanum level (Figure 3).

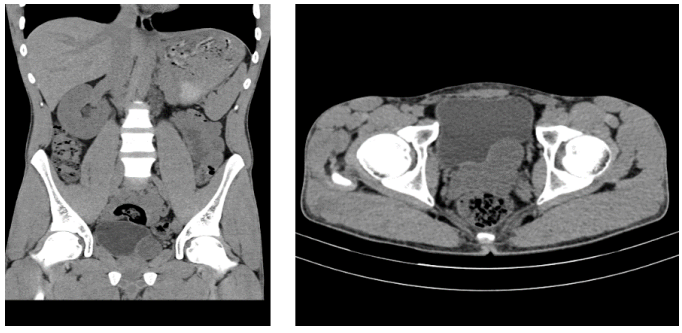

Figure 1. 2019 CT image (coronal and axial sections) (left seminal vesicle cyst and left renal agenesis)

CT: Computed tomography

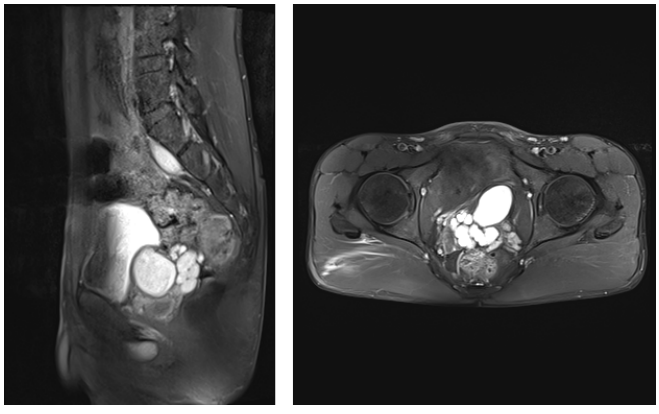

Figure 2. 2019 MRIs (sagittal and axial sections)

MRI: Magnetic resonance imaging

Semen analysis revealed low ejaculation volume (sperm vol: 1 mL sperm concentration 23.7M-progressive motility 39%, Kruger 5%). The patient was previously diagnosed with Zinner's syndrome and was believed to have developed ejaculatory duct obstruction based on the current imaging and laboratory findings. The decision to perform TUR-ED was made. TUR-ED was performed with a 22-Fr resectoscope. The pathology of the resected tissue was reported as a "benign fibromuscular tissue" sample. During resection, brown seminal fluid passes into the urethra. In the rectal examination performed in the same session, the cyst disappeared dramatically after resection. The seminal vesicle lumens were enlarged and the cyst walls were thickened (Figure 4). The surgery was completed without any complications, and a urethral catheter was inserted.

The patient's catheter was removed on the first postoperative day and he was discharged after his complaints were resolved. The patient was called for control on postoperative day 14. It was learned that he had hematospermia that lasted for 1 week, and it ended. There was no recurrence of the patient's complaints during the 1-year follow-up, and in the semen analysis performed in 2022, it was observed that the ejaculate volume increased (semen volume: 6 mL, sperm density 8.7 million/mL, progressive motility 50%, Kruger 3%).

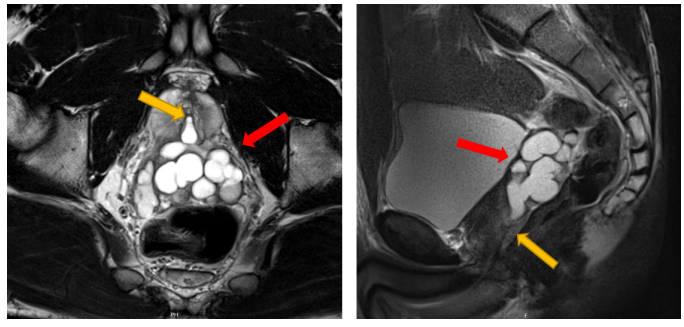

Figure 3. 2021 pelvic MRI (axial and sagittal sections): Dilation in the left seminal vesicle (red arrow), ejaculator duct obstruction (yellow arrow)

MRI: Magnetic resonance imaging

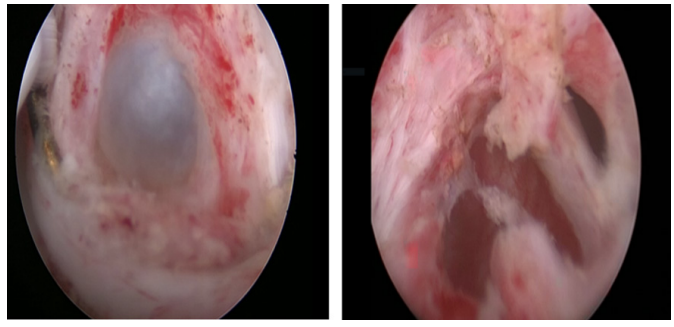

Figure 4. Ejaculatory duct image before TUR-ED (left) and post-resection images (right)

TUR-ED: Transurethral ejaculatory duct resection

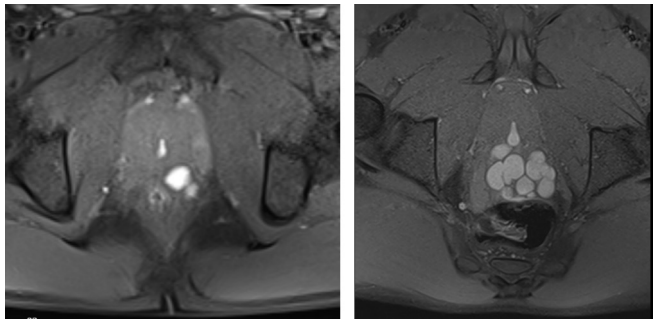

Figure 5. 2019 MRIs on the left and 2021 MRIs on the right (enlargement of the ejaculatory duct)

MRI: Magnetic resonance imaging

Discussion

ZS was first described by Zinner (1) in 1914. The condition is characterized by a triad of unilateral renal agenesis, ipsilateral seminal vesicle cysts, and ejaculatory duct obstruction. In our literature review, incidence information on ZS was not found. In a study conducted by Sheih et al. (3) in children, the incidence of the coexistence of two elements of Zinner's triad (unilateral renal agenesis and ipsilateral seminal vesicle cyst) was 0.0046% (1,2). The underdiagnosis of ZS may be due to the absence of symptoms or because symptomatic patients may respond to medical treatment and not undergo further evaluation. Another reason may be that clinicians do not consider it a prediagnosis.

This condition is associated with an anomaly in the development of the distal part of the mesonephric or Wolffian duct in early embryonic life. ZS is also believed to be similar to Mayer-Rokitansky-Kuster-Hauses syndrome in women (4). Seminal vesicle cysts may be congenital or acquired. Congenital defects occur during embryological development due to defects in the interaction between the mesonephric duct and urogenital sinus and defects in the development of the ureteric bud. Congenital cases are usually unilateral. Patients who are asymptomatic in the early stages of life usually become symptomatic in the sexually active period in the 2nd or 3rd decade and present to the physician (5). Patients should be informed about the rare occurrence of malignant cysts (6).

Seminal cysts smaller than 5 cm are usually asymptomatic and are diagnosed at a late stage. The most common and frequent symptoms are abdominal, perineal, and pelvic pain. Dysuria, hematuria, urinary tract infection, infertility, epididymitis, painful ejaculation, and prostatitis may also be associated. In addition, cases of incontinence have been reported. At the first presentation of our patient, abdominal and extragenital system examinations were normal, and digital rectal examination revealed minimal mass formation. Although the size of the cyst was 44x31 mm, the patient complained of severe scrotal pain after ejaculation. In patients with ZS, findings such as epididymal tenderness on physical examination and palpable mass on rectal examination may be present or may be completely normal (7).

Multiple imaging modalities are available for diagnosis. Abdominal and transrectal USG can be used to visualize renal agenesis and seminal vesicle cysts. This tool can provide information about the location of the cyst and other pathologies that may accompany it. Scrotal USG can be used to identify scrotal pathologies in the differential diagnosis. In the first case, epididymitis, orchitis, and other scrotal pathologies were excluded by scrotal USG. Intravenous pyelogram may show renal agenesis, but it is not sufficient to visualize seminal vesicle cysts. If it is large and compresses the bladder from the outside, the appearance of the filling defect may suggest a cyst (8-10).

CT is more effective in the differential diagnosis of renal pathologies, cysts, and urinary tract stone disease in terms of explaining scrotal radiating pain, which is one of the symptoms of the disease. In this case, CT showed renal agenesis and seminal vesicle cyst, but no urinary tract stone disease or obstruction.

MRI is a good diagnostic tool because it is radiation-free, shows a relationship with surrounding tissues, provides information about the site of origin, and differentiates malignancies. Prostatic cysts, seminal vesicle adenoma, müllerian duct cysts, and malignancies should be considered in the differential diagnosis. On MRI, seminal vesicle cysts are usually non-contrast enhancing, hypointense on T1, and hyperintense on T2 (11). If hemorrhage is present in the cyst, or if there is dense proteinaceous fluid, it may also be hyperintense. In the second case, a cystic structure with hyperintense content was observed on T1A, which was attributed to hemorrhage.

Transrectal-transperineal cyst aspiration, transurethral ejaculatory duct resection, and cyst excision should be considered in treatment. Since cyst aspiration will be accompanied by ejaculatory duct defects, the possibility of recurrence is high, and some authors recommend sclerosing material injection after aspiration (12). TUR-ED is not chosen as the primary approach because of the accompanying ductus agenesis-hypoplasia. Cyst excision can be performed using open-laproscopic and robotic methods. The success rate of the laparoscopic and robotic systems is similar to that of open surgery, and the recovery period is shorter (13). The patient's symptoms resolved after cyst excision. In this case, treatment options were presented, and laparoscopic seminal vesicle cyst excision was performed. In our case, the patient's complaints did not recur during the 2-year postoperative follow-up (14,15).

Two years after the operation, the patient presented with perineal pain. The MRI images at the first and second visits showed dilatation of the ejaculatory duct in addition to the seminal vesicle cyst at the second visit (Figure 5). Subsequently,

cystoscopy revealed ductus ejaculatorius obstruction, and TUR-ED was performed. As observed in our patient, seminal vesicle cyst excision is a treatment option for ZS. However, the possibility of recurrence of seminal vesicle pathologies is high because possible ejaculatory duct defects may accompany or develop. For this reason, TUR-ED should definitely be kept in mind for symptomatic treatment in these patients, and it should be kept in mind that TUR-ED may be required primarily if there is dilatation in the ejaculatory duct on imaging (16). Since modalities such as USG-mediated antegrade seminal vesicle flushing are rarely used today in the diagnosis and treatment of obstructive infertility in patients with ZS, TUR-ED operation may be preferred as the primary treatment method according to MRI findings (9,17,18).

When comparing the preoperative and postoperative spermiogram of our patient, we observed an increase in semen volume and sperm count. However, unexpectedly, Kruger's motility decreased while forward motility increased. Although the patients showed anatomical and symptomatic improvement, it is important to note that decreased sperm quality and abnormalities may occur in patients with ZS. This may be due to cellular, congenital, and endocrinological causes. It is important to conduct endocrinological and andrological follow-up in patients with ZS because of the potential deterioration of sperm quality in the future (19,20). Increasing research in this field can enhance sperm quality and prevent infertility.

Conclusion

The association between renal agenesis and seminal vesicle cysts is extremely rare. Patients may present with many non-specific symptoms. Therefore, differential diagnosis should be made carefully. Digital rectal examination, USG, CT, and MRI can be used as diagnostic methods. The patient should be evaluated multidisciplinaryly, and the possibility of infertility-subfertility despite treatment should be explained the patient. First-line cyst aspiration and seminal cyst excision may be considered as treatment options, but the possibility of recurrence should be considered. Even if seminal cyst excision is performed, it should be kept in mind that ejaculatory duct resection may be required in the future.

Ethics

Informed Consent: Verbal and written informed consent was obtained from the patient for the study.

Footnotes

Authorship Contributions

Surgical and Medical Practices: İ.E.D., Y.Ş., Concept: İ.E.D., Y.Ş., Design: E.E., Data Collection or Processing: İ.E.D., E.E., Analysis or Interpretation: Y.Ş., Literature Search: İ.E.D., E.E., Writing: İ.E.D.

Conflict of Interest: No conflict of interest was declared by the authors.

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