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

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Abstract

Zinner's syndrome (ZS) was first described by Zinner in 1914. It includes unilateral renal agenesis, ipsilateral seminal vesicle cyst, and ejaculatory duct obstruction. ZS treatments ranging from medical drug therapy to laparoscopic interventions have been tried in the literature. The 21-year-old patient presented with scrotal pain after ejaculation. The diagnosis was ZS, and the patient underwent transperitoneal laparoscopic left seminal vesicle cyst excision. After two 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.

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

Introduction

Zinner's syndrome (ZS) was first described by Zinner in 1914. Unilateral kidney agenesis is a syndrome associated with an ipsilateral seminal vesicle cyst and ejaculatory duct obstruction (1). They are usually diagnosed within 3 or 4 decades. Patients may present asymptomatic or with 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 reported.

The 21-year-old patient presented with scrotal pain after ejaculation. The diagnosis was ZS, and the patient underwent transperitoneal laparoscopic left seminal vesicle cyst excision. After 2 years, we performed transurethral ejaculatory duct resection (TUR-ED) 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.

Case Presentations

Case 1

In 2019, a 21-year-old patient was admitted to the emergency department with severe scrotal pain. It was learned that the patient had post-ejaculation pain for approximately three years and had experienced very severe pain attacks 3 times. There was no known history of disease or surgery, smoking, or alcohol use. No pathological findings were detected in the scrotal and abdominal examinations. Scrotal color Doppler ultrasound and urinalysis were normal. On digital rectal examination, there was minimal mass formation in the prostate.

Computed tomography (CT) showed 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 to the bladder and 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 the 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, ZS was diagnosed. Laparoscopic left seminal vesicle cyst excision was performed. There were no complications during the operation. The 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 reapplied two years later. The patient reported pain in the perineal area that worsened after ejaculation. During digital rectal examination, a fluctuating lesion of approximately 3 cm in diameter was palpated on the left side at the base of the prostate. Urinalysis was normal.

In the contrast-enhanced pelvic MRI image, the seminal vesicles and ejaculator duct were dilated on the leftside. A cystic structure with hyperintense content was observed on T1A,

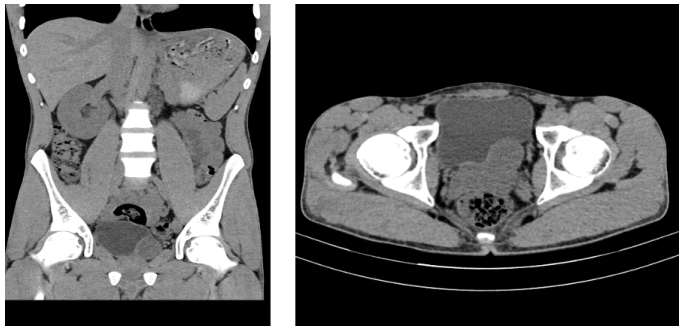

Figure 1. 2019 CT image (coronal and axial sections) (left seminal vesicle cyst and left renal agenesis)

CT: Computed tomography

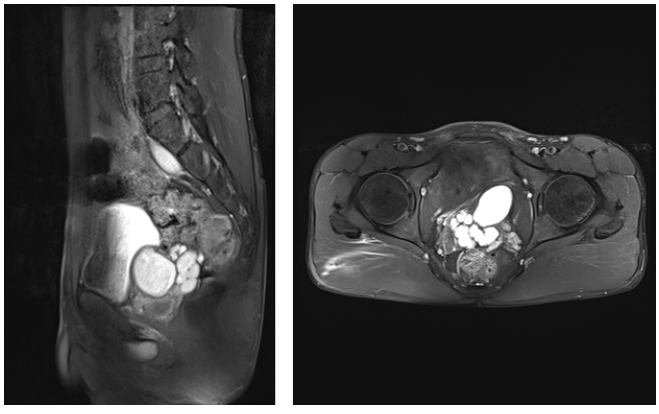

Figure 2. 2019 MR images (sagittal and axial sections)

MR: Magnetic resonance

which was thought to be a hemorrhage. No pathology could be distinguished at the verumontanum level (Figure 3).

Semen analysis revealed low ejaculation volume (sperm vol: 1 mL sperm concentration 23.7M-Progressive motility 39%-Kruger 5%). The patient, who was previously diagnosed with ZS, was believed to have developed ejaculatory duct obstruction based on the current imaging and laboratory findings. The decision for TUR-ED was made. TUR-ED was performed using a 22-Fr resectoscope. The resected tissue pathology was reported as a "benign fibromuscular tissue" sample. During resection, brown seminal fluid was observed to pass into the urethra. In the rectal examination performed in the same session, the cyst disappeared dramatically after resection. Seminal vesicle lumens were enlarged and 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 the 14th day after surgery. It was learned that he had hematospermia lasting 1 week and then 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%).

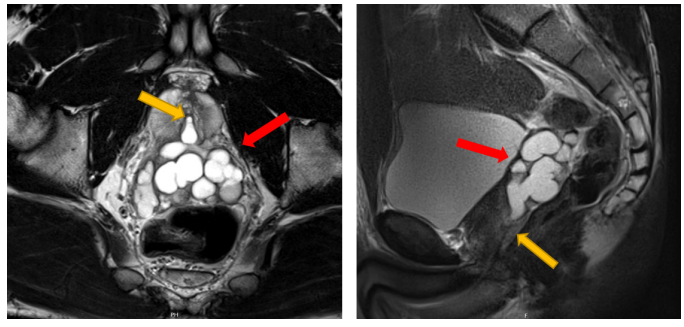

Figure 3. 2021 Pelvic MRI (axial and sagittal sections): Dilation in the left seminal vesicle (red arrow), ejaculator duct obstruction (yellow arrow)

MRI: Magnetic resonance imaging

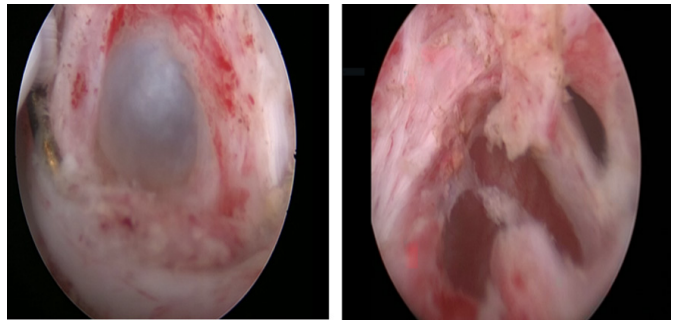

Figure 4. Ejaculatory duct image before TUR-ED (left) and post-resection images (right)

TUR-ED: Transurethral ejaculatory duct resection

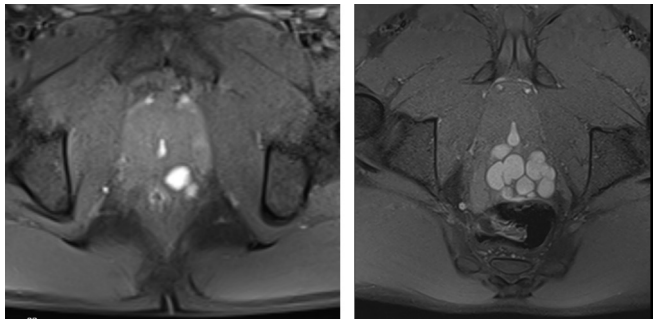

Figure 5. 2019 MR images on the left and 2021 MR images on the right (enlargement of the ejaculatory duct)

MR: Magnetic resonance

Discussion

ZS was first described by Zinner in 1914. It is characterized by a triad of unilateral renal agenesis, ipsilateral seminal vesicle cyst, and ejaculatory duct obstruction. In our literature research, incidence information on ZS could not be 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). The underdiagnosis of ZS may be because of 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.

It is associated with an anomaly in the development of the distal part of the mesonephric or Wolffian duct during early embryological life. ZS is also thought 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 because of defects in the interaction of 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 second or third decade and present to the physician (5). Patients should be informed about the rare possibility of malignant cysts (6).

Seminal cysts smaller than 5 cm are usually asymptomatic and diagnosed at a later 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. Cases of incontinence have also 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 can be used for diagnosis. Abdominal ultrasound and transrectal ultrasound can be used to detect renal agenesis and seminal vesicle cysts. It can provide information about the location of the cyst and other pathologies that may accompany. Scrotal ultrasound can be used to show scrotal pathologies in the differential diagnosis. In the first case, epididymitis, orchitis, and other scrotal pathologies were excluded by scrotal ultrasound. IVP may show renal agenesis but will not be sufficient to visualize a seminal vesicle cyst. 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 scan showed renal agenesis and a seminal vesicle cyst, but no urinary tract stone disease or obstruction.

MRI is a good diagnostic tool because it is radiation-free, shows the relationship with surrounding tissues well, provides information about the site of origin, and differentiates malignancies well. 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 there is hemorrhage 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 thought to be due to hemorrhage.

Transrectal-transperineal cyst aspiration, TUR-ED, and cyst excision should be considered in the treatment. Because cyst aspiration is 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 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 this study, the patient's complaints did not recur in the 2-year postoperative follow-up. (14,15)

Two years after the operation, the patient presented with perineal pain. When the MRI images at the first and second visits were analyzed, dilatation of the ejaculatory duct developed 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 seen in our patient, seminal vesicle cyst excision is considered 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 considered for symptomatic treatment in these patients, and it should be considered that TUR-ED may be required primarily if there is dilatation in the ejaculatory duct on imaging (16). Because modalities such as USG-mediated antegrade seminal vesicle flushing are rarely used today in the diagnosis andtreatment 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 pre-operative and postoperative spermiograms of our patient, we observed an increase in semen volume and sperm count. However, unexpectedly, Kruger decreased while forward motility increased. Although the patients showed anatomical and symptomatic improvement, 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 could 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 effectively as diagnostic methods. The patient should be evaluated multidisciplinarily, and the possibility of infertility–subfertility despite treatment should be explained the patient. First-line cyst aspiration and seminal cyst excision may be considered 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: Informed consent was obtained from the patient.

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: İ.E.D., E.E., Literature Search: İ.E.D., E.E., Writing: İ.E.D.

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