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A rare presentation of Zinner syndrome as recurrent epididymitis: A case report

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Abstract

Zinner syndrome is a rare congenital malformation characterized by ipsilateral renal agenesis, atresia of the ejaculatory duct, and cystic distension of the seminal vesicles. Symptoms typically arise in adolescence or adulthood and may include painful urination, perineal discomfort, and post-ejaculatory pain. Diagnosis, often made between the ages of 20 and 50 years, involves differentiating Zinner syndrome from other cystic disorders using techniques such as cystoscopy, MRI, intravenous urography, and transrectal ultrasound. Treatment varies based on symptom severity, ranging from conservative approaches to invasive surgeries. Regular follow-up is essential to identify complications and preserve fertility. Herein, we present a 27-year-old male with recurrent scrotal swelling and erythema incidentally diagnosed with Zinner syndrome. The patient had a history of recurrent epididymitis and was found to have cystic dilatation of the seminal vesicle, ipsilateral renal agenesis, and obstruction of the left ejaculatory duct on imaging. Given the patient's mild symptoms, he was managed conservatively with antibiotics and pain medication, resulting in significant improvement.

Keywords

Infertility, hemospermia, epididymitis, solitary kidney, rare diseases

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Introduction

Zinner syndrome (ZS) is a rare congenital malformation associated with an embryogenic abnormality of the genitourinary tract in the distal part of the mesonephric (wolffian) duct that appears between 4th and 13th gestational age.[1] It is characterized by a triad of ipsilateral renal agenesis, atresia of the ejaculatory duct, and cystic distension of the seminal vesicles.[2] Typically, asymptomatic until adolescence, ZS may cause dysuria, perineal pain, and post-ejaculatory pain. Infertility is common in ZS, often necessitating surgical intervention.[2] We herein describe a young male presenting with chronic scrotal swelling and erythema associated with hemospermia. The patient was incidentally found to have ipsilateral renal agenesis prompting the diagnosis of ZS.

Case presentation

A 27-year-old male patient was referred to our radiology department for the evaluation of left scrotal swelling and erythema. Upon further questioning, he reported recurrent episodes of left-sided epididymitis during the past 6 months. He also reported undocumented fever with diffuse swelling and redness of the left scrotum 3 days before admission. There was no history of urgency, frequency, dysuria, perineal and abdominal pain, or infertility. His past surgical and family histories were both unrevealing. Physical examination revealed painful erythema of the left testicle with relative improvement with elevation and a positive Prehn's sign. Digital rectal examination was normal, and no lymph nodes were palpated. The right testicular examination was unrevealing.

Sexual history revealed no issues of intercourse or fertility. The patient also had a 2-year-old male child with no complaints or signs of infertility. Notably, the patient was empirically treated for epididymitis three times in the previous 6 months with ceftriaxone and doxycycline, without

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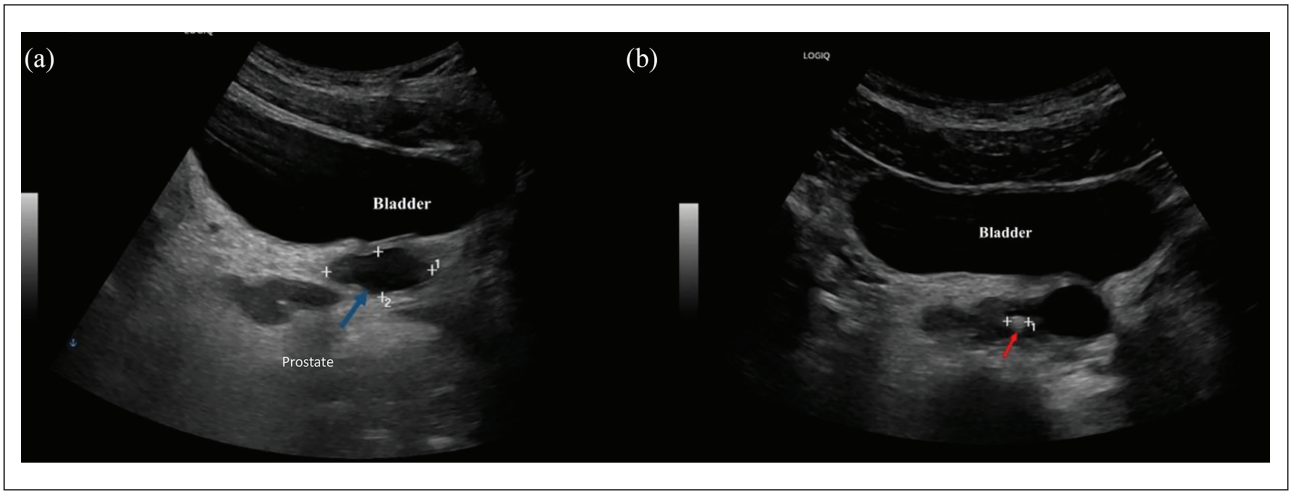

Figure 1. (a) A pelvic ultrasound image showing a cystic structure measuring approximately 3 cm × 1.4 cm located to the left of the prostate gland (blue arrow), which represents cystic dilatation of the left seminal vesicle. Note the urinary bladder is anterior and the prostate is posterior to the structure. (b) The above image shows a calcified focus measuring approximately 0.5 cm located at the terminal segment of the left seminal vesicle (red arrow), resulting in obstruction of the left ejaculatory duct.

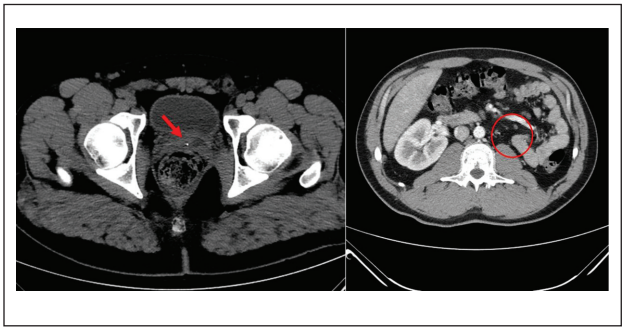

Figure 2. The CT scan displays the confirmed ultrasound findings, revealing an impacted stone in the left ejaculatory duct (red arrow), absence of the left kidney, and hypertrophied right kidney (red circle).
CT, computed tomography.

improvement in his condition. Consequently, a scrotal ultrasound revealed a thickened and hypoechoic left epididymis with normal vascularity and no signs of a hydrocele or varicocele. Pelvic ultrasound showed a 3 cm × 1.4 cm cystic structure on the left side of the prostate consistent with cystic dilatation of the left seminal vesicle (Figure 1). Furthermore, a calcified mass measuring 0.5 cm was detected at the terminal segment of the left seminal vesicle, which was obstructing the left ejaculatory duct (Figure 1). Laboratory studies including complete blood count, c-reactive protein, nucleic acid amplification tests for chlamydia and gonorrhea, serum prolactin, testosterone, luteinizing hormone, and follicular stimulating hormone were all within normal limits. Urinalysis was normal apart from true hematuria.

Semen analysis revealed normal sperm count, morphology, motility, and normal volume of 1.7 mL (*N*: 1.5–5 mL) along with hemospermia. A non-contrast computed tomography (CT) of the abdomen and pelvis confirmed left seminal vesicle dilation and an impacted stone in the ejaculatory duct, consistent with the ultrasound findings (Figure 2).

Given these results, the diagnosis of ZS was strongly suspected, prompting the performance of a contrast-enhanced CT scan to exclude any associated urological abnormalities. The CT scan confirmed left renal agenesis, with compensatory hypertrophy of the right kidney observed (Figure 2). Percutaneous seminal vesicle cyst drainage was performed with successful resolution of his symptoms at the 7-month follow-up. With conservative management, the stone spontaneously passed, subsequently leading to the cessation of the recurrent episodes of epididymitis. Considering the patient's clinical improvement, stone passage, and normal sperm count, we believe obstruction of the ejaculatory duct was alleviated.

The patient then received counseling and was prescribed antibiotics, and naproxen once daily for 2 weeks, resulting in significant symptomatic improvement. He was discharged with instructions to take naproxen as needed for ejaculatory pain, and regular follow-up. His symptoms progressively improved over time. In addition, he was offered sexual health counseling.

Discussion

ZS is a rare deformity that simultaneously affects the seminal vesicle and the upper urinary tract. Embryonic developmental abnormalities contribute to the occurrence of ipsilateral renal agenesis, ejaculatory duct atresia, and the formation of cysts in the seminal vesicle. This condition was first identified by Zinner in 1914.[3] The prevalence of ZS is difficult to estimate, given its rarity. In a systematic review of publications on ZS between 1999 and 2020, 214 cases were identified by a group of Chinese researchers.[4] Typically,

the diagnosis of this condition is commonly established between the ages of 20–50 years, as symptoms arise when seminal vesicular cysts reach a size of at least 5 cm causing obstruction and prompting medical attention.[5]

In some cases, the condition may present with voiding symptoms following the initiation of sexual activity, often appearing in the second and third decades of life.[3] However, in our specific case, the patient was asymptomatic and fertile, indicating that the condition did not affect his sexual activity. The most commonly observed symptoms, reported by van den Ouden et al.[6] include discomfort after ejaculation (21%), perineal pain (29%), increased frequency of urination (33%), episodes of epididymitis (27%), and dysuria (37%).[6] Recurrent episodes of epididymitis have also been documented in the literature as a presentation of Zinner's syndrome.[7,8] We postulate that the obstructed ejaculatory duct leads to the accumulation of fluid, resulting in stasis. This stagnant condition may trigger an inflammatory response, ultimately contributing to the occurrence of recurrent epididymitis episodes.

The differential diagnosis includes several cystic disorders affecting different pelvic organs, including diverticula of the ampulla of the vas deferens, prostatic cysts, ejaculatory duct cysts, ureteroceles, and abscesses.[9] The location of the cyst and associated developmental abnormalities, such as renal agenesis or malformations of the external genitalia, play a crucial role in clinching the diagnosis of ZS.[10]

Diagnostic tools for ZS include cystoscopy, MRI, intravenous urography (IVU), and transrectal ultrasound (TRUS). IVU shows smooth filling defects and no contrast excretion in the affected bladder side. TRUS reveals an anechoic cystic pelvic lesion. For accurate diagnosis and surgical planning, MRI is preferred.[8] In our case, a cystic structure measuring 3 cm × 1.4 cm was observed adjacent to the left side of the prostate gland. It was believed to be the source of the patient's dull pain and persistent obstructive symptoms.

For moderate cases, conservative therapy with antibiotics or cyst aspiration is recommended. However, for patients with severe symptoms, more invasive procedures like laparoscopic vesiculectomy, exploration, and transurethral resection of the ejaculatory duct (TURED) are advised.[11] Treatment should be considered only for symptomatic cases and is mainly surgical. Various surgical approaches are available, ranging from minimally invasive methods such as transrectal or transperineal cyst aspiration, to more invasive techniques. A more aggressive option involves transurethral unroofing of the cyst through TURED, leading to enhanced semen quality and increased paternity rates. However, this procedure carries the risk of complications, including potential harm to the rectum, bladder neck, external sphincter, and the possibility of causing retrograde ejaculation and epididymitis.[12] Thus, aggressive management of seminal vesicle cysts should be considered on a case-by-case basis, especially in the context of recurrent epididymitis. Our patient received conservative management due to mild symptoms. Surgical options and potential complications were discussed, including impotence, urinoma, and pelvic organ damage. Annual follow-up was recommended for monitoring cyst progression and assessing cancer development.

Fortunately, most individuals with ZS have a good prognosis. However, a significant subset of patients experiences complications and comorbidities that could rarely lead to death. The most common comorbidities were tumors arising from the seminal vesicle cyst, such as adenomas or adenocarcinomas among other malignancies.[13,14] On the other hand, some cases of ZS had also been associated with developmental deformities, as well as syndromic and inherited disorders.[7]

In summary, ZS is a rare potential diagnosis in young patients presenting with renal agenesis and nonspecific pelvic symptoms. It can present as infertility or chronic pelvic pain with unclear etiology not responding to typical medical therapy. Moreover, it can also present as recurrent episodes of epididymitis. We aim to portray the rarity and importance of including ZS in the differential diagnosis of refractory chronic pelvic pain and recurrent epididymitis, as well as the importance of utilizing renal imaging in such cases.

Conclusion

Although rare, ZS should be considered as a potential diagnosis in young patients presenting with renal agenesis and nonspecific pelvic symptoms. Symptoms typically result from the mass effect of the seminal vesicle cysts and irritation to the surrounding urinary bladder. MRI is preferred for evaluating pelvic masses and surgical planning. Conservative treatment with regular follow-up is recommended for mild presentations, while invasive procedures may be necessary for complex cases. Given the rarity of this condition, radiologists should remain vigilant for ZS in the appropriate clinical setting.

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Author contributions

N.K. contributed to data collection and article drafting. M.A. contributed to article drafting and editing. M.A. contributed to article drafting, editing, and final revision. A.A. contributed to article editing and final revision. M.Q. contributed to article editing and final revision. All authors read and approved the final article.

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Research ethics and patient consent

This case report did not require review by the Ethics Committee. Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor of this journal if requested.

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