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Case Report

A variant of Zinner syndrome with ectopic ureteral insertion into the seminal vesicle ☆

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ABSTRACT

Zinner syndrome comprises a triad of unilateral renal agenesis, ipsilateral seminal vesicle cyst, and ejaculatory duct obstruction, which can be accompanied by additional abnormalities of the genitourinary tract in some cases. Patients may be asymptomatic or present with urinary, reproductive, and/or local pain symptoms. Diagnosis is most commonly achieved via MRI. Here, we present the case of an 18-year-old male previously diagnosed with unilateral renal agenesis, who presented with testicular and penile pain, along with urinary urgency and frequency. MRI of the abdomen and pelvis revealed all three components of Zinner syndrome as well as an ectopic ureter emptying into the seminal vesicle. Our case adds to the existing limited literature on this rare syndrome and broadens the understanding of how this syndrome can present both clinically and radiologically.

Introduction

Zinner syndrome is a rare congenital condition characterized by a triad of developmental abnormalities affecting the male genitourinary system: unilateral renal agenesis, ipsilateral seminal vesicle cyst, and ejaculatory duct obstruction [1]. While once considered exceptionally rare, documented cases of Zinner syndrome have increased in recent years, attributed to the widespread utilization of advanced imaging techniques, which has led to the inadvertent discovery of asymptomatic cases [1,2]. When patients do develop symptoms, they frequently present with reproductive issues, such as painful ejaculation, recurrent epididymitis, and infertility. They may also experience urinary symptoms, including dysuria, hematuria, and urinary urgency as well as pelvic or perineal pain and discomfort [3,4]. Symptomatic cases typically present in the third to fourth decade, especially during periods of sexual activity [4]. However, as in our case, patients' presentations may deviate from this conventional demographic, as sexually inactive patients with Zinner syndrome can present at a younger age with genitoperineal pain and urinary symptoms [4]. Although computed tomography (CT) and ultrasonography can be used for diagnosis, the diagnosis of Zinner

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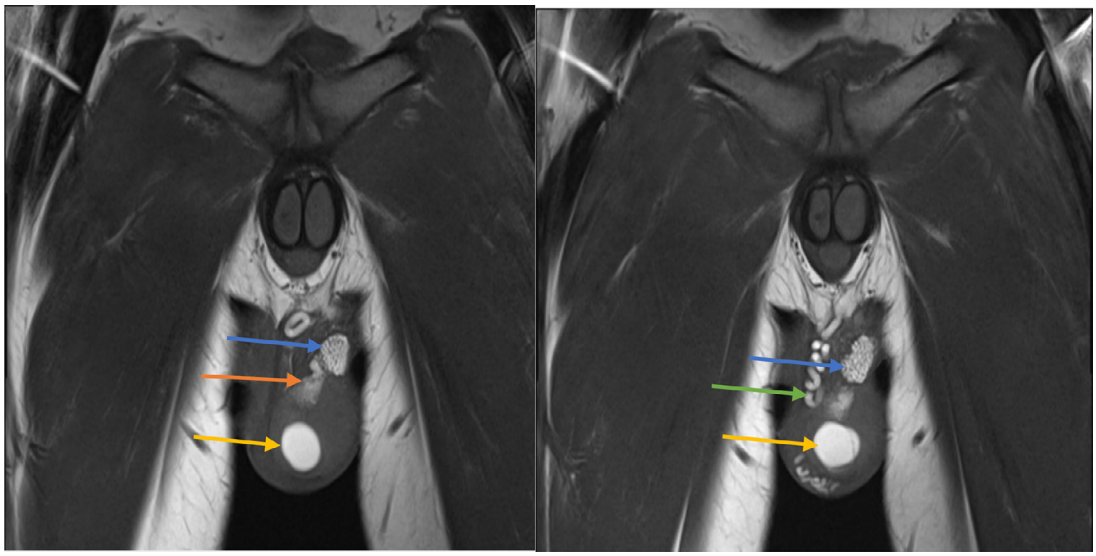

Fig. 1 – MRI T1-weighted images (coronal cut) showing dilated left testicular mediastinal tubules representing tubular ectasia of rete testis (orange arrow), along with enlargement of the left epididymis with multiple cystic interfaces (blue arrow) and dilated tortuous left ductus deferens along the medial side of epididymis posterior to the testis (green arrow); all of these structures elicit T1-hyperintense signal suggestive of proteinaceous/hemorrhagic contents. A well-defined rounded lesion (hematoma) can be seen at the inferior pole of the left testis (yellow arrow).

syndrome is most accurately confirmed by abdominal and pelvic magnetic resonance imaging (MRI), which can also detect coexisting anomalies such as an ectopic ureter [5].

Herein, we describe the case of an 18-year-old male diagnosed postnatally with congenital unilateral renal agenesis. Upon presenting with testicular and penile pain alongside urinary symptoms, further evaluation revealed the complete triad of Zinner syndrome—congenital renal agenesis, ejaculatory duct obstruction, and seminal vesicle cyst—along with an ipsilateral ectopic ureter emptying into the seminal vesicle.

Case presentation

A single, sexually inactive 18-year-old male with congenital unilateral renal agenesis presented to the clinic with the chief complaint of episodic pain in the penile and left testicular regions for 2 weeks. The pain was more prominent at night, intermittently alleviated by warmth, and worsened by cold stimuli. Over-the-counter analgesics, including paracetamol and nonsteroidal anti-inflammatory drugs, were ineffective. The pain was accompanied by left testicular swelling, retrograde ejaculation, and an array of urinary symptoms including frequent urgency that interfered with his sleep with no or minimal urination, intermittency, weak urine stream, and occasional dysuria. The patient also reported sudden bouts of nausea, dizziness, and diaphoresis as well as constipation. He had no surgical history, and family history was inconclusive.

Complete blood count, comprehensive metabolic panel, and tumor markers (beta-human chorionic gonadotropin and alpha-fetoprotein) were within normal limits. Hemoglobin A1c was normal. Urine studies were unremarkable aside from mucus in urine. Sperm studies revealed highly viscous sperm with a liquefaction test of 240 minutes and an alkaline pH of 8, but were otherwise normal.

Testicular ultrasound showed intact bilateral testes, homogenous with normal color flow. The left epididymis was enlarged with multiple cystic changes, representing tubular ectasia of the epididymis, and there was minimal hydrocele on the left side. Mirabegron was initially effective in controlling the pain, but the therapeutic effect eventually waned.

MRI of the pelvis and the inguinoscrotal region was obtained, and it showed abnormalities in the left testicular region, including tubular ectasia of the rete testis, an enlarged epididymis with cystic interfaces, and a dilated, tortuous ductus deferens. These structures demonstrated T1-hyperintense signals, indicating proteinaceous, and/or hemorrhagic contents. A hematoma was also noted at the inferior pole of the left testis. (Figs. 1–3). The left seminal vesicle cyst with a dilated ejaculatory duct were seen on axial view (Fig. 4). MRI of the abdomen and pelvis also confirmed the previously diagnosed left renal agenesis and demonstrated left ectopic ureter insertion into the left seminal vesicle. The atretic left ureter and the seminal vesicle both exhibit a high-intensity signal, suggesting the presence of hemorrhagic and/or proteinaceous contents. (Fig. 5).

Discussion

Zinner's syndrome, which was initially described by A. Zinner in 1914, is the male equivalent of Mayer-Rokitansky-Kuster-Hauser syndrome observed in females [6]. The syndrome is attributed to a developmental insult to the distal portion of the Wolffian (mesonephric) duct between the fourth and 13th weeks of gestation [1]. The Wolffian duct is a paired organ,

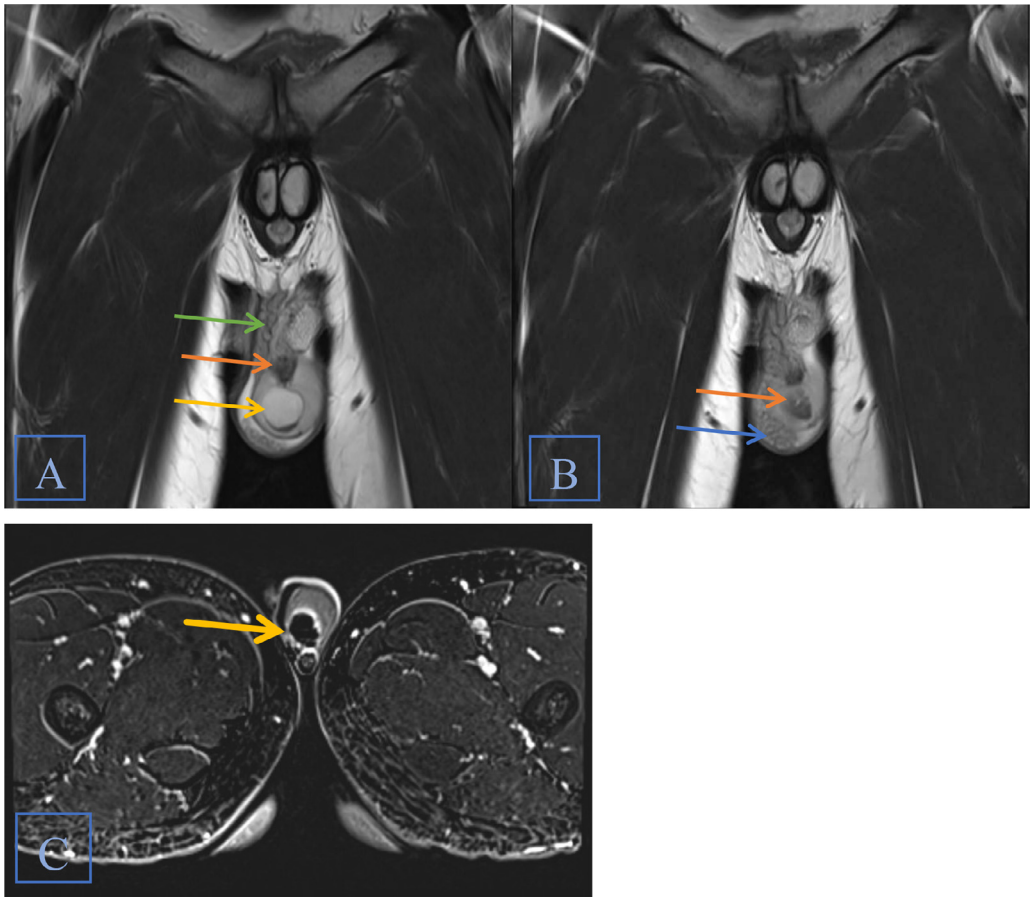

Fig. 2 – (A and B) MRI T2-weighted images (coronal cut) showing the tubular ectasia of left rete testis (orange arrow), enlarged epididymis (blue arrow), and dilated tortuous ductus deferens (green arrow). In addition, there is an intratesticular hematoma at the inferior pole of the left testis with a small air-fluid level (yellow arrow). (C) Subtracted postcontrast T1-fat suppression image demonstrating intratesticular hematoma with peripheral enhancement (yellow arrow).

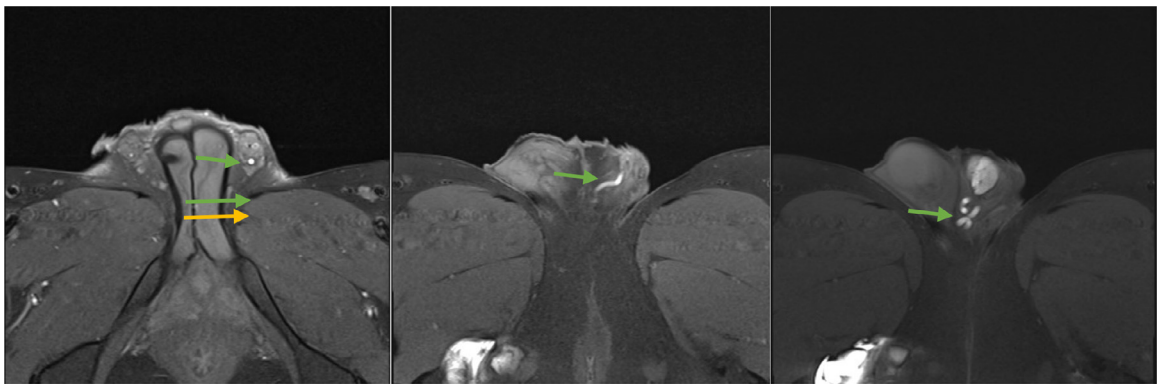

Fig. 3 – MRI T1-weighted images (axial cut) showing the dilated tortuous left ductus deferens at the spermatic cord and scrotum (green arrow).

and in males, it differentiates into the epididymis, vas deferens, ejaculatory ducts, seminal vesicle, hemitrigone, bladder neck, and proximal urethra [4,7]. The ureter is formed as a dorsal bud in the Wolffian duct, growing dorsocranially to fuse with the center of metanephric blastema to form the kidney; failure of this process can cause renal dysplasia or agenesis [7,8]. In addition, abnormal development of the distal part of the Wolffian duct can lead to atresia of the ejaculatory duct, causing its obstruction. Consequently, secretions would accumulate in the seminal vesicle, resulting in the formation of the seminal vesicle cyst [4]. If the ureter is formed from a more cranial location, it would instead meet one of the

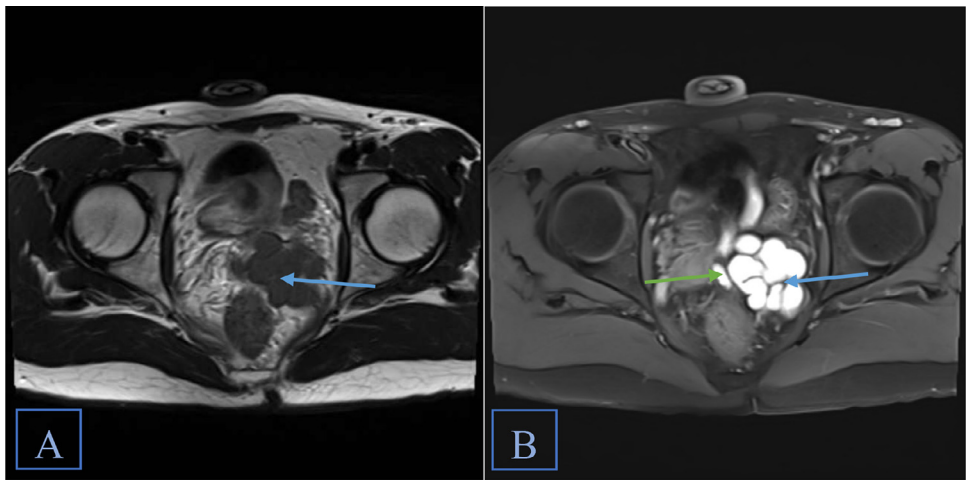

Fig. 4 – MRI axial T2 (A) and T1-fat suppression (B) weighted images showing the cystic dilatation of left seminal vesicle (blue arrow) with a dilated ejaculatory duct (green arrow) which elicit hypointense signal on T2-weighted imaging (A) and hyperintense signal on T1-fat suppression (B), suggestive of hemorrhagic/proteinaceous contents.

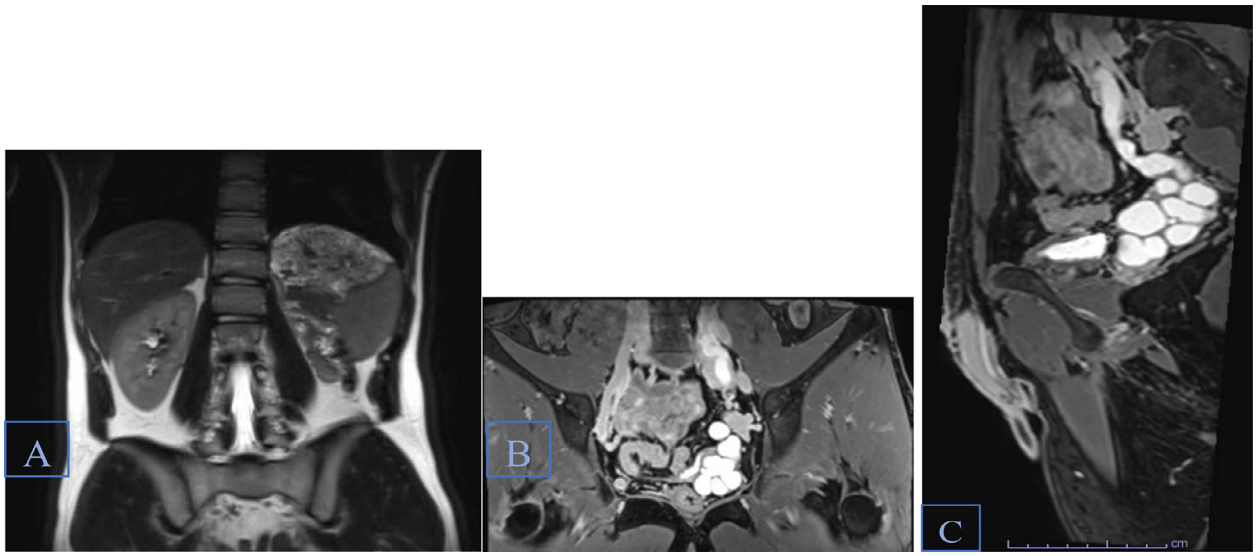

Fig. 5 – (A) Coronal T2-weighted MRI of the abdomen and pelvis showing the absent left kidney. (B,C) Coronal and sagittal T1-fat suppression images showing a tubular structure extending from the left side of seminal vesicle in the left pelvic and lumbar region that ends abruptly at the level of L4, representing the atretic left ureter. Both the atretic left ureter and the seminal vesicle show hyperintense signal content indicating hemorrhagic/high proteinaceous contents.

derivatives of the Wolffian duct, such as the ejaculatory duct, vas deferens, epididymis, or, as seen in our case, the seminal vesicle [8].

Although some patients are asymptomatic, a variety of symptoms can be associated with Zinner syndrome depending on the underlying mechanism [8,9]. Bladder irritation can lead to prostatitis-like symptoms or urethritis with urethral discharge, hematuria, and fever. Bladder outlet obstruction may manifest as lower urinary tract symptoms and painful, frequent urination of small volumes of urine, accompanied by a sense of urgency and a residual feeling of incomplete emptying. Cyst distention can cause genitoperineal and lower abdominal pain. Ejaculatory duct obstruction can cause painful ejaculation, decreased ejaculate volume and, in some cases, infertility [8]. In analyzing the relative frequency of symptoms, a systematic review spanning the medical literature published between 1999 and 2020 included 173 symptomatic cases of Zinner syndrome. The findings revealed that the most prevalent symptoms at presentation were dysuria (26.0%), urinary frequency (24.2%), perianal pain (20.2%), abdominal pain (14.5%), urinary urgency, and infertility (13.9% each). Interestingly, scrotal/testicular pain, our patient's primary complaint,

was present in less than 2% of reported symptomatic cases. The patient in our report also had constipation and nausea, symptoms observed in only 7 and 2 other cases, respectively, according to the systematic review. Furthermore, in our case, unilateral renal agenesis and ipsilateral seminal vesicle cyst were on the left side, which is slightly less commonly affected compared to the right side [10].

In cases of unilateral renal agenesis, individuals commonly maintain normal renal function due to compensation by the unaffected kidney [11]. This explains the normal renal function in our patient and in many similar cases in the literature. Nonetheless, the absence of one kidney may predispose individuals to specific renal pathologies, including nephrolithiasis, hypertension, or chronic kidney disease in later stages of life [11]. In addition, patients may occasionally develop renal impairment related to the other components of the triad. For instance, a 24-year-old diagnosed with Zinner syndrome developed hydronephrosis and renal dysfunction attributed to a sizable 14-cm seminal vesicle cyst obstructing the ureter. It should be noted, however, that such occurrences are rare, as seminal vesicle cysts typically do not exceed 5 cm in diameter [12].

MRI is the preferred modality for the diagnosis of Zinner syndrome [5]. No specific protocol is necessary for this purpose. Typically, a combination of imaging techniques including T1-weighted turbo spin-echo (TSE), T2-weighted TSE, and T1-weighted TSE with fat saturation is employed. These imaging sequences, conducted across the abdomen and pelvis in both transverse and coronal planes, are sufficient for establishing the diagnosis.

In this case, Zinner syndrome was also associated with an ectopic ureter emptying into the seminal vesicle. This variant has been documented in several studies, including a case series of 16 pediatric patients with Zinner syndrome, 11 of which had ipsilateral ectopic ureter emptying into the seminal vesical cyst; however, unlike our case, most were asymptomatic at the time of diagnosis [13]. The same variant was also reported in two 66-year old patients, one was completely asymptomatic, while the other required urgent surgical intervention following hospital admission due to fever, highlighting the variability in clinical presentations [7,14]. Two younger patients, aged 25 and 18, exhibiting this variant, presented with painful ejaculation [9,15]. Another patient, aged 21, initially misdiagnosed with chronic prostatitis, had the correct diagnosis revealed through subsequent investigations [16].

The treatment for Zinner syndrome is individualized, tailored to the presence or absence of symptoms and complications [8]. Observation and follow up is the best course for asymptomatic and mildly symptomatic patients [9]. However, symptomatic patients can be candidates for open or laparoscopic surgical interventions [14]. Our patient declined surgical intervention, citing fear of postoperative complications.

In conclusion, there is a diverse spectrum of clinical presentations for Zinner syndrome, spanning from asymptomatic cases to varying combinations of nonspecific, urinary, and reproductive manifestations. Diagnosis is largely based on imaging, preferably with MRI, which can help reveal additional anomalies like an ectopic ureter. The case we presented draws attention to the importance of considering Zinner syndrome in the differential diagnosis for young patients with unilateral renal agenesis who present with genitoperineal pain and/or urinary symptoms.

Patient consent

Written informed consent for the publication of this case report was obtained from the patient.

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