CASE REPORT

Zinner syndrome detected as an incidental ultrasonographic finding in a 38-year-old Rwandan man

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Abstract

Congenital lesions of the seminal vesicles are rare. Zinner syndrome is an uncommon congenital syndrome comprising seminal vesicle cysts with unilateral renal agenesis and ipsilateral ejaculatory duct obstruction. Patients can be asymptomatic or present with symptoms, usually between the ages of 11 and 40 years. The clinical presentation is nonspecific but abdominal pain, pelvic pain, urinary irritation symptoms, and infertility have been reported previously. Diagnosis is mainly through computed tomography and magnetic resonance imaging. The management strategy for Zinner syndrome depends on the severity of symptoms and the size of the cysts. Small and asymptomatic cysts are managed conservatively. Surgical management can proceed through open, laparoscopic, or robotic approaches. Herein, we present a rare case of Zinner syndrome managed at a private multispeciality hospital in Kigali, Rwanda.

Keywords: Zinner syndrome, seminal vesicles, seminal vesicle cysts, ejaculatory ducts, renal agenesis, dysuria, abdominal pain, pelvic pain, urology, congenital malformations, Rwanda

Introduction

Zinner described a rare congenital syndrome occurring as a triad of unilateral renal agenesis, ipsilateral seminal vesicle cysts, and ipsilateral ejaculatory duct obstruction.[1] Its frequency has been reported as 0.00464%.[2] About 200 cases have been documented, including 2 indigenous African patients.[3]-[5]

Case presentation

A 38-year-old African man was referred to our urology clinic after being evaluated and treated for dysuria at a peripheral facility, where a cystic pelvic mass was detected incidentally during ultrasonographic examination. His urination pattern was normal with no associated abdominal or perineal pain. He was married and a father of 3 children. He had been diagnosed with hypertension 1 year previously, and it was well controlled with medication. Upon his presentation to our urology clinic, his dysuria symptoms had resolved after a course of oral antibiotics prescribed at the peripheral facility.

Examination revealed normal external genitalia and bilateral nondilated vasa deferentia. Digital rectal examination revealed a normal prostate and no palpable seminal vesicle masses. His initial laboratory evaluation was normal with the following available results: creatinine, 0.5 mg/dL (normal range, 0.6-1.2 mg/dL); urea, 4 mmol/L (2.50-7.50 mmol/L); and urine analysis showing 0-1 white blood cells per high-power field with no bacterial growth on culture.

Abdominal ultrasonography revealed left kidney agenesis and a left paravesical cyst—its location and appearance suggestive of a left ureterocoele—abutting on the urinary bladder wall (Figure 1).

Complementary transrectal ultrasonography revealed the left paravesical cyst to be contiguous with the left seminal vesicle (Figure 2). Computed tomography (CT) revealed left renal agenesis with a perivesicular, periprostatic cystic mass.

Magnetic resonance imaging (MRI) confirmed left renal agenesis with 2 distinct left seminal vesicle cysts, measuring 4.3 × 4.2 × 4.2 cm and 1.6 × 1.1 cm, respectively (Figure 3 and Figure 4). These features corresponded with Zinner syndrome.

As of the composition of this report, the patient remains asymptomatic after the initial antibiotic and analgesic treatment prescribed at the peripheral facility and the resolution of his dysuria at that time. The dysuria has not recurred, and

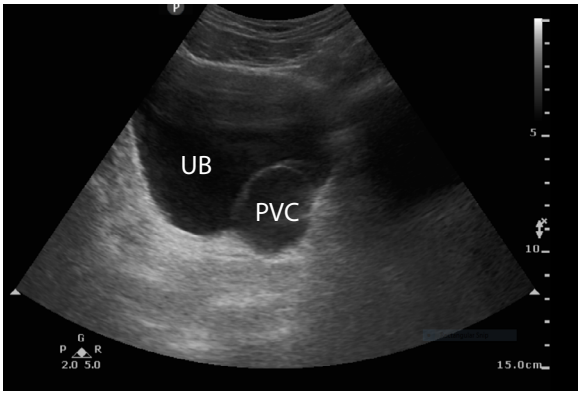

Figure 1. Transabdominal ultrasonogram showing a left perivesical cyst
PVC, perivesical cyst; UB, urinary bladder

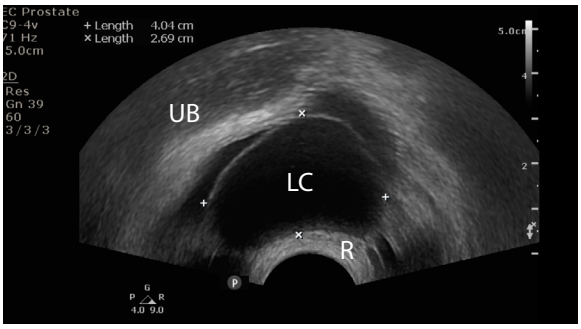

Figure 2. Transrectal ultrasonogram showing a left seminal vesicle cyst
LC, left seminal vesicle cyst; R, rectum; UB, urinary bladder

repeat evaluation 6 months after his initial presentation to our clinic revealed a normal urinalysis, as well as unchanged abdominal and pelvic sonography findings, with minimal post-void residual volume (30 mL). He remains on antihypertensive medication and on follow-up with the internal medicine clinic. Surgical management will be considered if the contexts of future abdominal or pelvic pain, with or without urinary or sexual dysfunction.

Discussion

The congenital malformations associated with Zinner syndrome occur in weeks 4 to 13 of embryogenesis and affect the distal portion of the mesonephric ducts, leading to seminal vesicle cyst formation. At the proximal end of the ureteral bud, the metanephros is not joined at the centre, leading to agenesis. Ureteral bud or mesonephric duct malformations cause Zinner syndrome.[6]-[8]

Patients commonly present between the ages of 11 and 40 years (mean age, 30 years). Symptom onset is attributable to an associated increase in cyst size and the onset of sexual activity. Small cysts (<5 cm) are usually asymptomatic.[3],[5],[9] Common manifestations include bladder obstruction and irritation symptoms, such as dysuria, frequency, haematuria, and prostatitis.[5],[10] Abdominal,

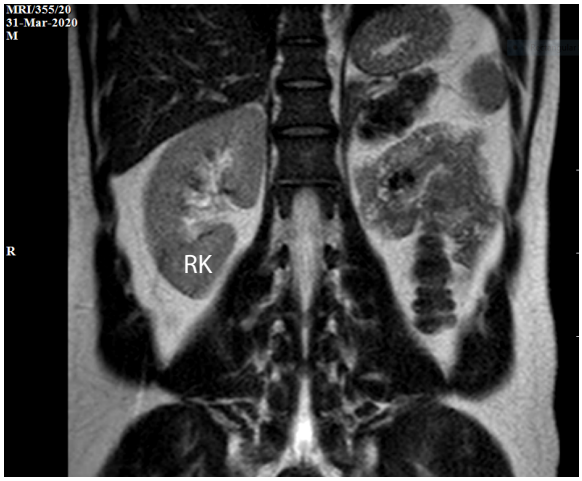

Figure 3. T2-weighted magnetic resonance image showing left renal agenesis
RK, right kidney

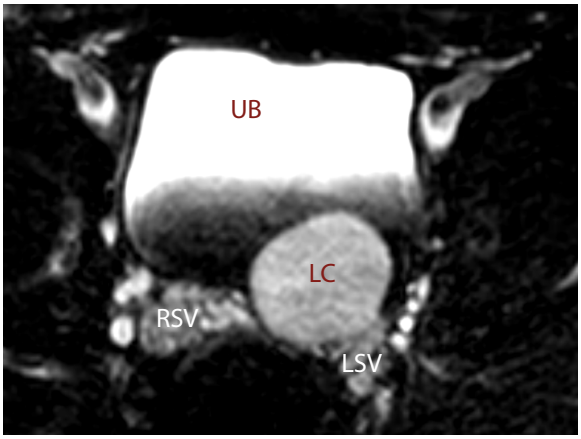

Figure 4. T2-weighted magnetic resonance image showing a left seminal vesicle cyst and a normal right seminal vesicle
LC, left seminal vesicle cyst; LSV, left seminal vesicle; RSV, right seminal vesicle; UB, urinary bladder

perineal, and pelvic pain are also common. Infertility was noted in 17% of patients in 2 case series, and ejaculatory duct obstruction can lead to multiple seminal abnormalities, such as oligospermia and azoospermia.[3],[5],[9],[11]

In a case series of 52 men, 79% of patients had a supraprostatic mass palpable by digital rectal exam.[5] Fewer cases (19%) can have normal examination findings, as noted in our patient. On cystoscopy, extravesical mass formation and hemitrigone have been observed.[3],[5]

Zinner syndrome is mainly diagnosed through radiological evaluation. Transrectal ultrasonography is recommended for the initial evaluation and is the most commonly used modality for diagnosing seminal vesicle cysts. An anechoic pelvic mass with a surrounding thick irregular wall is considered a positive finding.[3] CT usually reveals ipsilateral renal agenesis and retrovesicular periprostatic mass

formation. CT is superior to ultrasonography.[12] Transrectal ultrasonography and CT can be ideal for resource-constrained settings, especially if CT defines the origin of the pelvic mass.

MRI is the imaging modality of choice because it is excellent in delineating the male pelvic anatomy. T1-weighted signal hypointensity and T2-weighted signal hyperintensity characterize seminal vesicle cysts and facilitate a prompt diagnosis of Zinner syndrome.[3],[13]

Asymptomatic patients and those with mild symptoms are managed conservatively with regular follow-up.[2] Surgical management includes open exploration with resection of the seminal cysts (and ureteral remnant if it is present). This has excellent outcomes.[2],[3],[5] Laparoscopic and robotic surgery are the preferred minimally invasive treatment modalities because of their favourable safety profile and outcomes. These strategies allow for clear visualization of the challenging intrapelvic anatomy, and magnification facilitates seminal vesicle cyst dissection during laparoscopy.[14]-[17]

Conclusions

The triad of unilateral renal agenesis, ipsilateral seminal vesicle cysts, and ipsilateral ejaculatory duct obstruction is known as Zinner syndrome.

Zinner syndrome should be suspected in patients with unilateral renal agenesis who present with non-specific abdominal or pelvic pain, urinary irritation symptoms, and infertility.

MRI is the diagnostic radiological investigation of choice for Zinner syndrome.

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