Case Report

Challenging clinical presentation of Zinner syndrome

Fatos Sada, MD, PhD, Elton Cekaj, MD, Abdallah Al-Madani, MD, Sepideh Jahanian, MD, Shamsun Nahar, MBBS, Juna Musa, MD, MSc, Ketë Mamilla, MD, Dijon Musli, MD, Murtaza Ahadi, MBBS, Florim Leniqi, BSc, Tamanna Agarwal, MD, Fjolla Hyseni, MD, PhD candidate, Valon Vokshi, MD, Adam Benjamin Fink, MD, FNU Deepali, MBBS, Jeton Shatri, MD, PhD, Sadi Bexheti, MD, PhD

a Department of Anesthesiology and Reanimation, University Clinical Center of Kosovo, Faculty of Medicine University of Pristina, Pristina, Kosovo
b Department of Radiology, Regional Hospital, Durrës, Albania
c Department of Radiology, Mother Teresa University Hospital Center, Tirana, Albania
d Department of Radiology, Regional Hospital, Shkoder, Albania
e Internship Medical Student KHMC, Amman, Jordan
f Department of Anesthesiology, Mayo Clinic, Rochester, MN, USA
g Dhaka Medical College, Dhaka, Bangladesh
h Department of Endocrinology Diabetes and Nutrition, Mayo Clinic, Rochester, MN, USA
i Department of Anesthesiology and Critical Care, Mother Teresa University Hospital Center, Tirana, Albania
j Department of Anatomy, Faculty of Medicine, University of Pristina, str. Rruga e Ilindenit pa nr. 1200 Tetovë Republic of North Macedonia, Pristina, Kosovo
k Oncology Department, Bolan Medical Complex Hospital, Quetta, Pakistan
l Faculty of Medicine, University of Gjakova, Gjakova, Kosovo
m Faculty of Medicine in Hradec Králové, Charles University, Prague, Czech Republic
n NYU Langone Health, New York, NY, USA
o Department of Anesthesiology and Reanimation, University Clinical Center of Kosovo, Prishtina, Kosovo
p 1st Faculty of Medicine at Charles University, Prague, Czech Republic
q Department of Gastroenterology & Hepatology, Mayo Clinic, Rochester, MN, USA

ARTICLE INFO

Article history:
Received 30 August 2022
Revised 26 September 2022
Accepted 3 October 2022

ABSTRACT

Zinner syndrome is a rare congenital malformation of the mesonephric duct comprising of seminal vesicle cyst, ipsilateral renal agenesis, and ejaculatory duct obstruction. Clinical presentation varies with perineal pain, painful ejaculation, hematospermia and infertility common presenting complaints. Here, we present a case of Zinner syndrome in a 35-year-old male patient.
Introduction

Zinner syndrome is a rare urological triad of unilateral renal agenesis, ipsilateral seminal vesicle cyst and ejaculatory duct obstruction. It was first reported by Zinner in 1914 [1], hence the name. It is associated with the abnormal development of the mesonephric or Wolffian duct in the first trimester of gestation [2]. Incomplete migration of the ureteric bud which originates from the Wolffian duct, causes failure to connect with the metanephros, leading to renal agenesis. However, the seminal vesicles continue to develop with insufficient drainage, resulting in distention and cyst formation [3,4]. This syndrome is mostly asymptomatic until the third or fourth decade of life, and symptoms usually manifest with the beginning of sexual activity. Patients commonly present with unspecific symptoms such as problems in voiding (dysuria, urgency, frequency), perineal pain, possible hematuria, recurrent urinary tract infections, and painful ejaculation. Infertility is also frequent, caused by ejaculatory duct obstruction [3,4]. It is an uncommon condition with less than a few hundred cases reported in literature.

Fig. 1 – Computed tomography images (A) 3D CT: defect in the urinary bladder from right seminal vesicle cystoma and left unique kidney. (B) Coronal CT: notes the filling of the urinary bladder with contrast media and compression from the right seminal vesicle cystoma. (C) Axial CT: notes the filling of the urinary bladder with contrast media and compression from the right seminal vesicle cystoma. (D) Right cystoma prominence in the urinary bladder and a unique left urinary jet is revealed. Hypertrophied left kidney is noted.
A 35-year-old man is brought from prison to the clinic with complaints of increasing abdominal discomfort. Blood pressure is 125/85 mmHg, heart rate 78/min, and temperature 37°C. Physical examination was normal. The external genitalia appear normal. There is no dysuria or hematuria. Rectal examination shows normal anal sphincter tone but a slightly enlarged prostate. PSA levels were normal. Hormonal profile and blood tests showed insignificant changes. Urinalysis was within normal range.

An abdominal ultrasound was ordered and a right cystic prominence in the urinary bladder with unique left urinary jet is revealed. Furthermore, a hypertrophied left kidney is noted.

Considering the ultrasound findings, Zinner syndrome was suspected. Further imaging examinations were ordered (Fig. 1).

IV contrast CT in different modes revealed:

- 3D CT: defect in the urinary bladder due to a right seminal vesicle cystoma and hypertrophied left kidney
- Axial CT: notes filling of the urinary bladder with contrast media and compression from a right seminal vesicle cystoma
- Coronal CT: notes filling of the urinary bladder with contrast media and compression from a right seminal vesicle cystoma.
- The MRI findings confirmed well differentiated right seminal vesicle cystoma from the urinary bladder and a hypertrophied left kidney.

Considering the imaging results, a diagnosis of Zinner syndrome was made. Surgical treatment was suggested to the patient along with close follow-up.

Discussion

Zinner’s syndrome is a triad of unilateral renal agenesis alongside ipsilateral seminal vesicle cysts and ejaculatory duct obstruction. Since Zinner first described the syndrome in 1914, approximately 100 cases have been reported in medical literature [5]. The condition originates during embryogenesis
due to maldevelopment of the Wolffian duct, occurring between the 4th and 13th week of gestation [6]. Although it is hard to accurately determine the prevalence of the syndrome, an analysis of 280,000 children with unilateral renal agenesis conducted by Sheih et al and published in 1990, discovered six children with cystic dilations of the seminal vesicles using ultrasonography [7]. This would indicate a prevalence of 0.002142857%. MRI serves as the gold standard to make a definitive diagnosis due to its superior ability to delineate between soft tissue structures in the pelvis. Cystic dilations within the seminal vesicles are typically no larger than 5cm although 12cm dilations have been reported in the literature [5]. Patients most frequently present during their 2nd and 3rd decades of life [5], although about 50 pediatric cases reported in literature, with 12 of them occurring in the first year of life [8]. A systematic review analyzing 214 cases between 1999–2020 by Liu et al found that 80.8% of cases presented with clinical symptoms while the rest were discovered incidentally. The mean age of patients was 29.35 years. Furthermore, the study found that the most common clinical symptoms were dysuria (26.0%), urinary frequency (24.2%), perianal pain (20.2%), abdominal pain (14.5%), urinary urgency and incontinence (13.9% each) [9]. The issue of infertility is of particular importance as up to 45% of males with Zinner syndrome suffer from infertility due to ejaculatory duct obstruction. The exact pathophysiological mechanism is not fully understood as azoospermia would not be expected in the setting of a unilateral ejaculatory duct obstruction, which is part of the constitutional triad of Zinner’s syndrome. Several theories have been suggested to explain how unilateral obstruction results in azoospermia and oligospermia but to date the exact mechanism is poorly understood. The first theory is that unilateral obstruction triggers an autoimmune response that produces anti-sperm antibodies that subsequently destroy sperm in the contra lateral testis. The second is that long lasting obstruction triggers the formation of reactive oxygen species that induce apoptosis in germ cells [6,10]. Treatment is surgical and is offered only to symptomatic patients [6,11]. Both open and transurethral unroofing of the cysts have been done besides the more aggressive vesciculectomy [11]. Minimally invasive robotic surgery offers better overall visualization and lower injury rates [6]. For patients suffering from ejaculatory duct obstruction, trans-urethral resection of the ejaculatory duct is performed. Following surgical treatment, studies have found an improvement of semen parameters ranging from 63.0–83.0% in patients suffering from ejaculatory duct obstruction [6]. In conclusion, this case represents a less frequent yet known presentation of Zinner’s syndrome, wherein the primary symptom is abdominal pain. Once a diagnosis is made utilizing correct imaging techniques, all symptomatic patients should be evaluated for surgical treatment. Additionally, they should undergo semen evaluation as surgical intervention may not only provide symptomatic relief but also improves semen parameters thus decreasing infertility.

**Conclusion**

Zinner syndrome has a varied clinical presentation. Physicians and radiologists should be aware of the clinical and radiological presentation of the condition. Magnetic resonance imaging is the imaging modality of choice for diagnosing the condition. Management is aimed at pain relief with surgical intervention reserved for severe cases.

**Patient consent**

We obtained written, informed consent for publication from the patient.

**References**

[1] Zinner A. Ein Fall von intravesikaler Samenblasenzytose. Wien Med Wochenschr 1914;64(605):e-9.

[2] Florim S, Oliveira V, Rocha D. Zinner syndrome presenting with intermittent scrotal pain in a young man. Radiol Case Rep 2018;13(6):1224–7. doi: 10.1016/j.radcr.2018.08.012.

[3] Pereira BJ, Sousa L, Azinhais P, Conceição P, Borges R, Léão R, et al. Zinner’s syndrome: an up-to-date review of the literature based on a clinical case. Andrologia 2009;41:322–30. doi: 10.1111/j.1439-0272.2009.00939.x.

[4] Ghangne NF, Aggarwal B, Sahu AK. Zinner syndrome: a unique triad of mesonephric duct abnormalities as an unusual cause of urinary symptoms in late adolescence. Indian J Urol 2010;26(3):444–7. doi: 10.4103/0970-1591.70592.

[5] Mehra S, Ranjan R, Garga UC. Zinner syndrome—a rare developmental anomaly of the mesonephric duct diagnosed on magnetic resonance imaging. Radiol Case Rep 2016;11(4):313–17. doi: 10.1016/j.radcr.2016.04.002.

[6] Hofmann A, Vauth F, Roesch WH. Zinner syndrome and infertility— a literature review based on a clinical case. Int J Impot Res 2021;33:191–5. doi: 10.1038/s41443-020-00360-0.

[7] Sheih C-P, Hung CS, Wei CF, Lin CY. Cystic dilatations within the pelvis in patients with ipsilateral renal agenesis or dysplasia. J Urol 1990;144(2):324–7.

[8] Cascini V, Di Renzo D, Guerriero V, Lauriti G, Lelli Chiesa P. Zinner syndrome in pediatric age: issues in the diagnosis and treatment of a rare malformation complex. Front Pediatr 2019;7:129. doi: 10.3389/fped.2019.00129.

[9] Liu T, Li X, Huang L, Li H, Cai K, Jiang J, et al. Zinner syndrome: an updated pooled analysis based on 214 cases from 1999 to 2020: systematic review. Ann Palliat Med 2021;10(2):2271–82. doi: 10.21037/apm-20-1997.

[10] Cito G, Sforza S, Gemma L, Coccì A, Di Maida F, Dabizzi S, et al. Infertility case presentation in Zinner syndrome: can a long-lasting seminal tract obstruction cause secretory testicular injury? Andrologia 2019;51(11):e13436. doi:10.1111/and.13436.

[11] Cascini V, Di Renzo D, Guerriero V, Lauriti G, Lelli Chiesa P. Zinner syndrome in pediatric age: issues in the diagnosis and treatment of a rare malformation complex. Front Pediatr 2019;7:129. doi: 10.3389/fped.2019.00129.