wjpmr, 2025, 11(7), 443-446

WORLD JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH
www.wjpmr.com

SJIF Impact Factor: 6.842

Case Report
ISSN (O): 2455-3301
ISSN (P): 3051-2557

ZINNER SYNDROM: CASE REPORT

***Nachid A., Safwate R., Moumen O., Kbirou A., Moataz A., Dakir M., Debbagh A. and Aboutaieb R.**

Department of Urology, CHU Ibn Rochd Casablanca.

***Corresponding Author: Nachid A.**
Department of Urology, CHU Ibn Rochd Casablanca.

Article Received on 13/05/2025 **Article Revised on 07/06/2025** **Article Accepted on 28/06/2025**

ABSTRACT
Zinner syndrome is a rare congenital anomaly resulting from Wolffian duct maldevelopment and is characterized by the triad of ipsilateral renal agenesis, seminal vesicle cyst, and ejaculatory duct obstruction. It often remains asymptomatic and is incidentally discovered, but when symptomatic, it can present with pelvic discomfort, urinary symptoms, or infertility. We report the case of a 39-year-old male, father of three, who presented with intermittent hemospermia. Imaging confirmed the diagnosis of Zinner syndrome, revealing a right seminal vesicle cyst associated with ipsilateral renal agenesis.

KEYWORDS: zinner, hemospermia, agenesis, seminal cyst.

INTRODUCTION

Zinner syndrome is a rare congenital anomaly of the distal Wolffian duct (mesonephros) in males[1], occurring during the first trimester of gestation.[2] It is characterized by a clinical triad[1]: renal hypoplasia[1] or agenesis, most commonly ipsilateral[3], a seminal vesicle cyst, and ipsilateral ejaculatory duct obstruction.[1]

First described by Zinner in 1914, with fewer than 200 cases reported[2], this syndrome may be symptomatic or discovered incidentally during clinical or radiological examinations.[1]

Symptomatic patients present with nonspecific signs between the second and fourth decades, including irritative and obstructive urinary symptoms, perineo-scrotal discomfort, post-ejaculatory pain, and infertility.[1,4] The presence of these symptoms is associated with seminal vesicle cysts measuring 5 cm or more, reported in one in three to five patients.[5,6] Due to its nonspecific presentation, Zinner syndrome can go undiagnosed.[7]

Depending on the symptoms, treatment may be conservative or surgical.[8]

In this article, we present a rare case of a 39 years old patient whose initial presentation was hemospermia and right scrotal pain.

CASE REPORT

A 39 years old father of 3, with no particular pathological history, presented with intermittent hemospermia, with the notion of low-abundance hematuria and pain in the right scortum, with no notion of erectile dysfunction, urethral discharge or other associated signs.

The clinical examination was unremarkable, with a BMI of 21.5. Rectal examination revealed a firm, painless prostate, with a well-defined, fairly firm mass adjacent to the right lobe that was tender to palpation.

The cytobacteriological analysis of the urine was sterile except for hematuria, while the spermogram revealed only hemospermia with otherwise normal parameters.

the patient subsequently benefited from a uro-scanner (figure1) with a complementary ultrasound (figure 2) objectifying right renal dysplasia with the presence of a cystic formation in the right seminiferous vesicle.

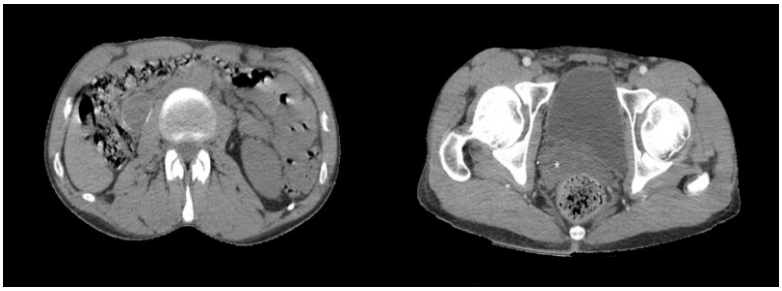

Figure 1: Uro CT Scan: Renal Dysplasia And Seminal Vesicle Cyst (Star).

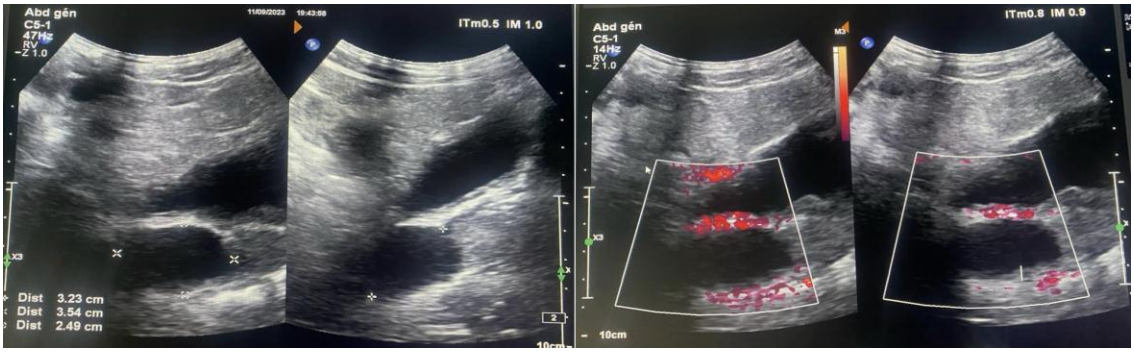

Figure 2: ultrasound scan showing seminal cyst.

Complementary MRI in T2 sequence in two planes, STIR and Diffusion was performed showing an intermediate signal in T2, hypersignal in diffusion with high ADC.

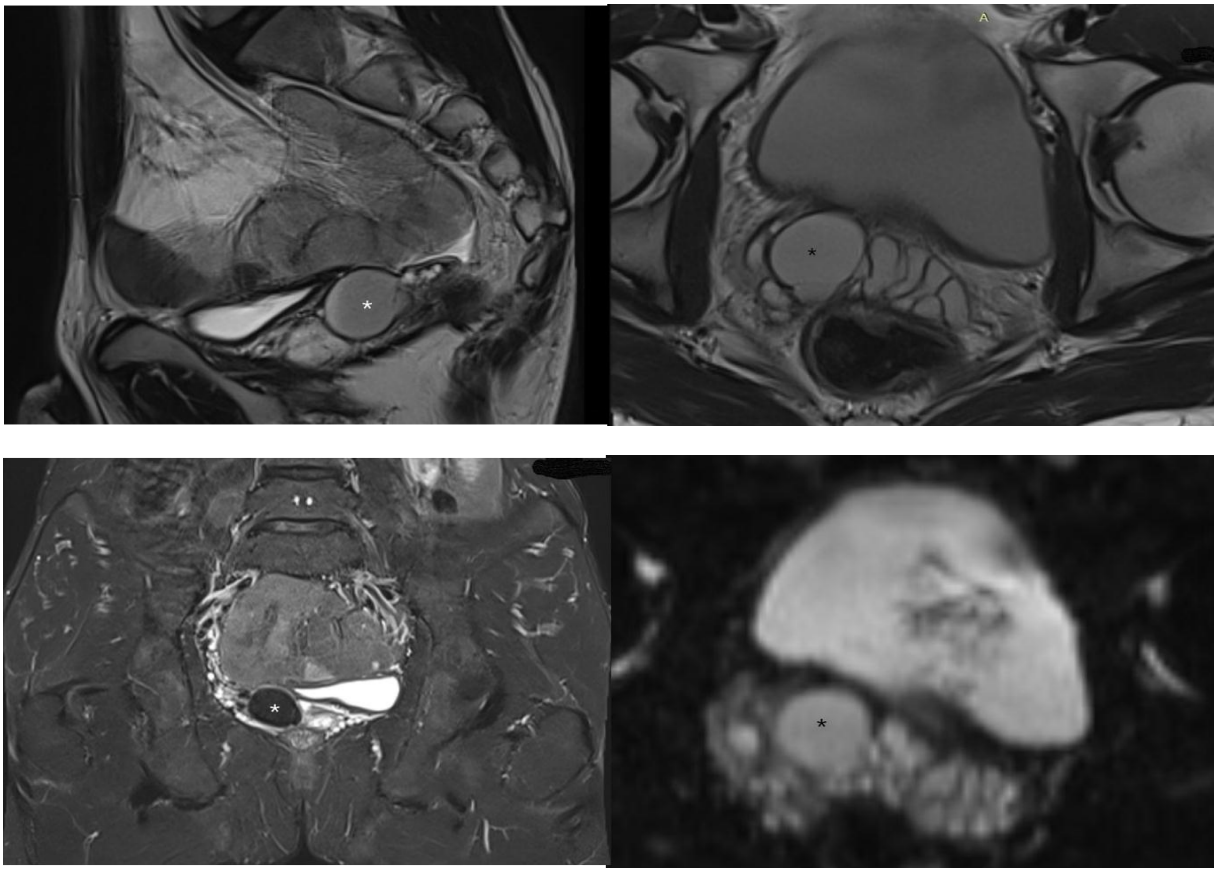

Figure 3: right seminal cyst (star) in intermediate T2 signal and diffusion hypersignal with high ADC.

DISCUSSION

The association of seminal vesicle cysts with ipsilateral renal agenesis was first identified by Smith in 1872 and later described by Zinner in 1914.[3] Zinner syndrome is considered the male equivalent of Mayer-Rokitansky-Küster-Hauser syndrome in females.[9-11]

This rare syndrome results from a developmental failure of the distal mesonephric (Wolffian) duct between the 4th and 13th weeks of gestation.[12] It is characterized by a clinical triad: seminal vesicle cyst, ipsilateral renal agenesis (most commonly), and ejaculatory duct obstruction.[13] Other urological anomalies such as ureterocele, hypospadias, or testicular and epididymal malformations may also be present.[14,15], with a low risk of malignant transformation.[9,14,16]

Failure of Wolffian duct development and the absence of the ureteral bud result in renal agenesis and ejaculatory duct atresia. The seminal vesicle cyst forms due to inadequate drainage despite normal gonadal development.[2]

Seminal vesicle cysts may be congenital or acquired[17], with bilateral cysts typically being acquired. Acquired cases are discovered during prostate surgery or after multiple episodes of chronic prostatitis.[18]

Symptoms usually appear between the second and fourth decades, coinciding with increased sexual activity.[14,15] The absence of symptoms makes this syndrome frequently misdiagnosed.[19-21]

Small cysts (<5 cm) are often asymptomatic and detected only during physical examination, either by abdominal palpation or digital rectal examination, where they may mimic a bulging prostate mass.[22,23]

Clinically, Zinner syndrome presents with a wide range of symptoms, primarily urinary issues such as dysuria, increased urinary frequency, urgency, hematuria, perineo-scrotal pain, painful ejaculation and defecation, and hematospermia (24). In a study by Van den Ouden et al. involving 52 patients, the most common symptoms were dysuria (37%), increased frequency (33%), perineal pain (29%), epididymitis (27%), post-ejaculatory pain (21%), and scrotal pain (13%). Infertility was found in 9 patients (17%).[6] In our case, the patient reported hemospermia and low hematuria, the sperm analysis revealed only hemospermia.

Lower urinary tract infections are observed in cases of giant cysts, which can reach up to 12 cm in size, causing urethral compression.[20,25]

Infertility, caused by ejaculatory duct compression, must be thoroughly investigated in suspected Zinner syndrome cases. It typically results in a low ejaculate volume (1 mL), azoospermia, an alkaline pH, and seminal fluid that is low in carnitine and fructose but rich in citrate.[2] Hormonal testing (FSH, LH, testosterone), urine analysis, urine culture, and cyst fluid analysis may also be considered.[26]

Differential diagnoses include ejaculatory duct cysts (typically more centrally located and rarely associated with renal agenesis), prostatic cysts, ureterocele, urethral diverticulum, abscess, utricular cysts, and hydronephrosis of a pelvic kidney.[14–16,19,27]

Abdominal and transrectal ultrasound are useful imaging techniques for detecting this syndrome. CT scans can reveal renal agenesis and seminal vesicle cysts but are surpassed by MRI, which is the gold standard for diagnosis. MRI typically shows a hyperintense T2 and hypointense T1 cyst, with a hyperintense T2 signal indicating the presence of blood or protein-rich fluid inside the cyst.[2,19,20,28]

Treatment depends on symptom severity. Conservative management includes antibiotics, transurethral cyst aspiration, with or without instillation therapy.[7] Invasive treatments, such as laparoscopic or robotic vesiculectomy, are reserved for highly symptomatic giant cysts.[3]

CONCLUSION

Zinner syndrome is a rare congenital anomaly resulting from a developmental failure of the distal Wolffian duct, leading to a clinical triad of seminal vesicle cyst, ipsilateral renal agenesis, and ejaculatory duct obstruction. Symptoms, often appearing in the second to fourth decades of life, range from urinary disturbances to infertility, though small cysts may remain asymptomatic. Diagnosis relies on imaging, with MRI being the gold standard for detecting seminal vesicle cysts and associated anomalies. Treatment varies based on symptom severity, with conservative management for mild cases and surgical intervention for symptomatic or giant cysts. In our case, given the mild symptoms, the patient opted for regular follow-up in a private healthcare facility.

REFERENCES

1. Tan Z, Li B, Zhang L, Han P, Huang H, Taylor A, et al. Classifying seminal vesicle cysts in the diagnosis and treatment of Zinner syndrome: A report of six cases and review of available literature. Andrologia [Internet]. févr 2020 [cité 17 oct 2020]; 52(1). Disponible sur: https: //onlinelibrary.wiley.com/doi/abs/10.1111/and.13397
2. Zinner A. Ein Fall von Intravesikaler Samen blasen cyste. Wien Med Wochenschr, 1914; 64: 605–609.
3. Florim S, Oliveira V, Rocha D. Zinner syndrome presenting with intermittent scrotal pain in a young man. Radiol Case Rep, déc 2018; 13(6): 1224‑7.
4. Seminal Vesicle Cysts With Upper Urinary Tract Abnormalities: A Singlecenter Case Series of Pediatric Zinner Syndrome Kosuke Takemura, Atsuko Sato, Yosuke Morizawa, Ryohei Kufukihara, Shun Iwasa, and Hiroyuki Satoh.
5. Fiaschetti et al., 2017.
6. van den Ouden, Blom, Bangma, & Spiegeleer, 1998.
7. Williams RD, Sandlow JI. Surgery of the seminal vesicles. In: Walsh PC, Retik AB, Vaughan ED, Wein AJ, editors. Campbell's urology. 7. Philadelphia: Saunders, 1998; p. 3299–307.
8. Cito G, Sforza S, Gemma L, Cocci 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 [Internet]. déc 2019 [cité 17 oct 2020]; 51(11). Disponible sur: https: //onlinelibrary.wiley.com/doi/abs/10.1111/and.13436
9. Kanavaki A, Vidal I, Merlini L, Hanquinet S. Congenital seminal vesicle cyst and ipsilateral renal agenesis (Zinner Syndrome): a rare association and its evolution from early childhood to adolescence. Eur J Pediatr Surg Reports, 2015; 03(02): 098–102.
10. 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.
11. Symeonidis EN, et al.Incidental finding of Zinner syndrome in a Greek military recruit: a case report of a rare clinical entity. Mil. Med. Res, 2019; 6(1): 4.

12. Zinner A. Ein fall von intravesikaler samenblasenzyste. Weien Med Wschr, 1914; 64: 604–10.
13. Schukfeh N, Kuebler JF, Schirg E, Petersen C, Ure BM, Gl€uer S. Dysplastic kidney and not renal agenesis is the commonly associated anomaly in infants with seminal vesicle cyst. BJU Int, 2009; 103: 816–819.
14. Pereira BJ, et al.Zinner's syndrome: an up-to-date review of the literature based on a clinical case. Andrologia, Oct. 2009; 41(5): 322–30.
15. Tan Z, Li B, Zhang L, Han P, Huang H, Taylor A, et al.Classifying seminal vesicle cysts in the diagnosis and treatment of Zinner syndrome : A report of six cases and review of available literature. Andrologia, 2019; June: 1–10.
16. Livingston L, Larsen CR. Original Report. AJR Am J Roentgenol, 2008; July: 177–80.
17. King BF, Hattery RR, Lieber MM, Berquist TH, Williamson Jr B, Hartman GW. Congenital cystic disease of the seminal vesicle. Radiology, 1991; 178(1): 207–11.
18. Rappe BJM, Meuleman EJH, Debruyne FMJ. Seminal vesicle cyst with ipsilateral renal agenesis. Urol Int, 1993; 50(1): 54–6.
19. Fiaschetti V, Greco L, Giuricin V, De Vivo D, Di Caprera E, Di Trapano R, et al.Congenital Seminal Vesicle Cyst and Ipsilateral Renal Agenesis (Zinner Syndrome): A Rare Association and Its Evolution from Early Childhood to Adolescence. J. Pediatr Surg Case Rep, 2015; 16(12): 530–2.
20. Slaoui A, Regragui S, Lasri A, Karmouni T, El Khader K, Koutani A, et al. Zinner's syndrome: report of two cases and review of the literature. Basic Clin Androl, déc 2016; 26(1): 10.
21. Campora M, Olivero A, Toncini C, Spina B, Fulcheri E, Terrone C, et al. Zinner Syndrome: A Diagnostic Challenge. The Aid of Morphology, Embryology, and Immunohistochemistry. Urology, oct 2017; 108: e3‑5.
22. Heaney JA, Pfister RC, Meares Jr EM. Giant cyst of the seminal vesicle with renal agenesis. AJR Am J Roentgenol, 1987; 149(1): 139–40.
23. Kenney PJ, Leeson MD. Congenital anomalies of the seminal vesicles: spectrum of computed tomographic findings. Radiology, 1983; 149(1): 247–51.
24. Beeby DI. Seminal vesicle cyst associated with ipsilateral renal agenesis: case report and review of literature. J Urol, 1974; 112: 120–2.
25. Sundar R,Sundar G. Zinner syndrome: An uncommon cause of painful ejaculation. BMJ Case Rep, 2015: 1–5 2015.
26. Levisay GL, Holder J, Weigel JW. Ureteral ectopia associated with seminal vesicle cyst and ipsilateral renal agenesis. Radiology, 1975; 114(3): 575–6.
27. Farrelly JS, Cleary MA, Weiss RM, Morotti R, Stitelman DH. Acquired seminal vesicle cyst in a teenager with a urethral stricture of unknown etiology. J. Pediatr Surg Case Rep, 2016; 7: 8–12.
28. Pavan N, Bucci S, Mazzon G, Bertolotto M, Trombetta C, Liguori G. It's not always varicocele: a strange case of Zinner syndrome. Can Urol Assoc J., 2015; 9(7–8): 535.