Radiology Case Reports 20 (2025) 1721–1725

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Zinner syndrome unveiled: Ectopic ureter and seminal vesicle cyst leading to urinary dysfunction: A case report ☆,☆☆

Zaid Sawaftah[a], *Khaled Sawafta*[a], *Moath Hattab*[a], *Adel Abu Al Rub*[a], *Omar Sawafta*[a], *Jehad Khamaysa*[b], *Humam Emad Rajha*[c], *Jana Dibas*[a,*], *Muath Daraghmeh*[d], *Yazan Dibas*[e]

[a] *Department of Medicine, An Najah National University, Nablus, Palestine*
[b] *Department of Radiology, Tubas Turkish Governmental Hospital, Tubas, Palestine*
[c] *College of Medicine, QU Health, Qatar University, Doha, Qatar*
[d] *Department of Radiology, Patient's Friends Society, Nablus, Palestine*
[e] *Department of Radiology, Makassed hospital, Jerusalem, Palestine*

ARTICLE INFO

Article history:
Received 28 October 2024
Revised 25 November 2024
Accepted 6 December 2024

Keywords:
Renal agenesis
Ejaculatory duct obstruction
Zinner
Seminal vesicle cyst
Tamsulosin

ABSTRACT

Zinner syndrome (ZS) is a rare congenital urological condition characterized by a triad of ipsilateral seminal vesicle cysts, unilateral renal agenesis, and ejaculatory duct obstruction, first described in 1914. This case report details the presentation and management of a 27-year-old male diagnosed with ZS following a 2-month history of urinary frequency, hesitancy, dysuria, and painful ejaculation. Physical examination revealed a left lower abdominal mass, and imaging confirmed the classic findings of ZS, including unilateral renal agenesis, an enlarged seminal vesicle cyst, and an ectopic ureter. Conservative treatment with tamsulosin initially improved symptoms, but due to the cyst's large size and the risk of complications, laparoscopic excision was recommended for definitive management. ZS is often diagnosed late due to nonspecific symptoms, with an average diagnosis age of 29.35 years. MRI is the gold standard for diagnosis, revealing seminal vesicle cysts, renal agenesis, and ejaculatory duct obstruction. Conservative management is reserved for asymptomatic cases, while symptomatic patients benefit from surgical intervention. This case emphasizes the importance of timely imaging, particularly in patients with nonspecific lower urinary tract symptoms, and highlights the role of tamsulosin as an effective interim therapy before definitive surgical treatment. The case underscores the need for clear diagnostic criteria

☆ Acknowledgments: We express our deep gratitude to the medical ward staff for their invaluable support in completing this report. No specific grant from funding agencies was received for this work.
☆☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.
* Corresponding author.
E-mail address: janadebas7@gmail.com (J. Dibas).
https://doi.org/10.1016/j.radcr.2024.12.017

and treatment pathways to improve outcomes in this rare condition, which can lead to infertility if not managed appropriately.

Introduction

Zinner syndrome (ZS) is an extremely rare congenital urological condition characterized by ipsilateral seminal vesicle cysts, unilateral renal agenesis, and ejaculatory duct obstruction. First described by Zinner in 1914, this anomaly arises from abnormal development of the mesonephric (Wolffian) duct during early embryogenesis, specifically between the 4th and 13th weeks of gestation [1]. The incidence of ZS is approximately 0.046%, with infertility documented in up to 45% of cases [2].

Diagnosis is often delayed due to nonspecific symptoms, including lower urinary tract symptoms (LUTS) and infertility, typically manifesting in the second or third decade of life. The average age of diagnosis is around 29.35 years [2], and some of ZS clinical manifestations include prostatitis, painful ejaculation, and perineal discomfort, with severity often correlating with cyst size [3].

The nonspecific presentation complicates treatment selection. Conservative management is recommended for asymptomatic individuals, while invasive treatments are reserved for symptomatic patients who fail conservative measures [4].

Case presentation

A 27-year-old male presented to the urology clinic with a 2-month history of increased urinary frequency, hesitancy, mild dysuria, occasional painful ejaculation, and a sensation of rectal fullness, along with intermittent lower abdominal discomfort. His medical history was notable for having only 1 kidney. He denied any history of urinary tract infections, hematuria, or problems with ejaculation. Given his lack of sexual activity, his fertility status was undetermined. Physical examination revealed normal vital signs, mild tenderness in the left flank, and a palpable mass in the left pelvic region, along with mild testicular tenderness but no visible swelling or erythema.

Considering the urinary symptoms and palpable mass, further investigations were initiated. Urinalysis and semen analysis were normal. Abdominal ultrasound showed a cystic mass in the left pelvis, suggestive of seminal vesicle enlargement. An MRI with contrast revealed marked dilatation of the left seminal vesicle with high-signal content, indicating fluid accumulation (Fig. 1A). Additionally, a dilated tubular structure identified as an ectopic ureter was traced from the left lumbar to the pelvic region (Fig. 1B), draining into the enlarged multicystic seminal vesicle (Figs. 1C-F). A schematic illustration of the anatomy of this case is present in (Fig. 2).

The patient was initially treated with the alpha-blocker tamsulosin, which improved urinary hesitancy and flow, along with analgesics for pain relief. After 2 weeks, he reported significant improvement in urinary frequency and discomfort. However, due to the size of the seminal vesicle cyst and fear potential complications, such as infections and infertility, surgical intervention was deemed necessary. The patient was referred to a specialized urology center for laparoscopic excision of the cyst.

Discussion

Congenital anomalies of the kidney and urinary tract (CAKUT) have a prevalence of 4-30/10,000 live births [5]. Among these, Zinner syndrome (ZS) is an extremely rare triad of urinary and reproductive tract malformations, characterized by ipsilateral seminal vesicle cysts, unilateral renal agenesis, and ejaculatory duct obstruction. Since its first description by Dr. Zinner in 1914, approximately 214 patients have been documented from 1999 to 2020 [1,2]. We identified around 50 additional cases, 16 of which were from Taiwan, bringing the estimated total in the literature to approximately 270-290 [6].

In ZS, the ureteric bud fails to connect with the metanephros due to incomplete migration, disrupting the differentiation of the metanephric blastema and resulting in renal agenesis and ejaculatory duct atresia [7]. Mutations in the metanephric blastema or disruptions in retinoic acid signaling may further inhibit the growth of the ureteric bud, contributing to these anomalies [4]. Despite these developments, gonadal maturation continues, leading to cystic enlargement of the seminal vesicle.

ZS typically presents in the second to fourth decades of life, often coinciding with reproductive age. Symptoms arise when seminal vesicle cysts exceed 5 cm, causing mass effects on surrounding structures or urinary tract complications [5]. Common symptoms include dysuria (37%), urinary frequency (33%), perineal pain (29%), and epididymitis (27%) [2]. Imaging studies, particularly MRI, are the gold standard for diagnosis, revealing seminal vesicle cysts, unilateral renal agenesis, and ejaculatory duct obstruction [8].

Diagnosing asymptomatic ZS is particularly challenging, making imaging analysis critical for accurate diagnosis and surgical planning. Transrectal ultrasound (TRUS) is the preferred technique for assessing seminal vesicle cysts in adults, while transabdominal ultrasound is often first-line in adolescents [9]. Characteristic findings include cystic masses in the retrovesical space and the absence of the ipsilateral kidney. TRUS provides detailed information regarding cyst characteristics, aiding in interventions if necessary [9].

Computed tomography (CT) scans are also utilized but have limitations compared to MRI in delineating cyst origins due to lower contrast resolution. MRI allows clearer visualization of pelvic anatomy, with simple seminal vesicle cysts show-

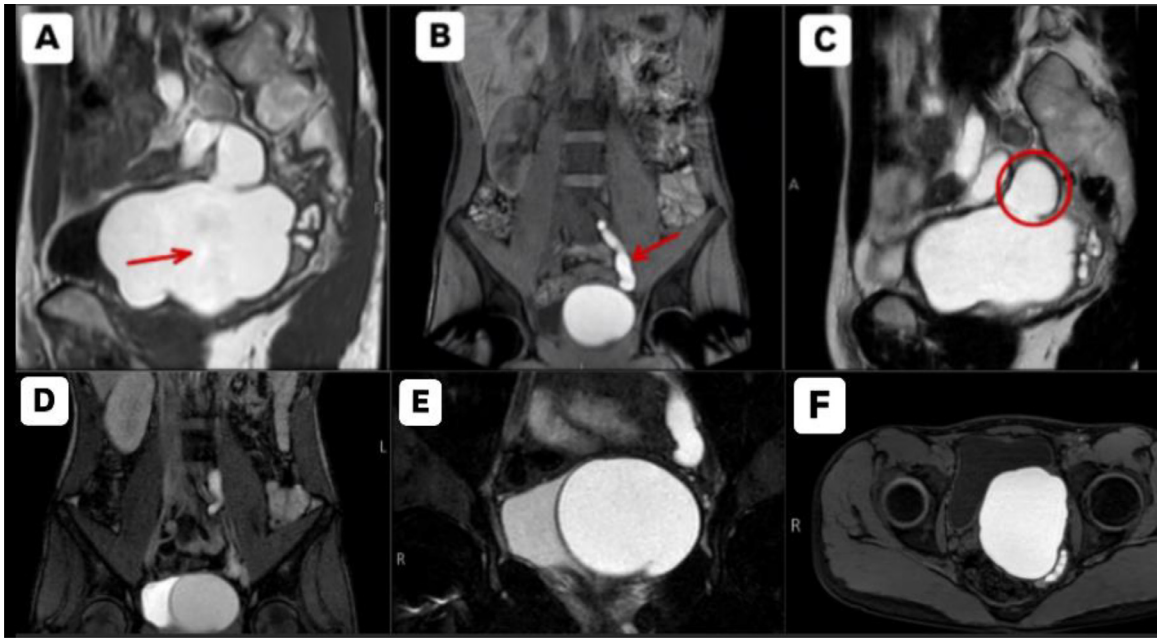

Fig. 1 – This figure illustrates the MRI findings. The labelled structures correspond to specific finding location. (A) MRI T1-weighted sagittal section demonstrating significant dilatation of the left seminal vesicle, which shows high signal content, suggesting the presence of hemorrhagic fluid or a high proteinaceous component. (B) A tubular structure in the left lumbar and pelvic region, identified as the dilated ureter. (C) MRI T2-weighted sagittal section revealing that the dilated ureter empties ectopically into the left seminal vesicle, which is markedly enlarged. Also (D-F) showing a large cystic lesion.

Table 1 – Classification of seminal vesicle cysts in Zinner syndrome.

Wall of the lesions	Types	Size	Density	Content	Management
Thin wall and homogeneous	Type I	type I and II cysts are smaller and have no clinical symptoms, while lesions larger than 5 cm stay symptomatic	CT value <20 HU	Liquid such as water	Asymptomatic not requiring any treatment. Symptomatic or diameter of lesion >5 cm can be conservative treatment, aspiration, laparoscopic, or robot assisted surgery
	Type II		CT value >30-40 HU	Proteinaceous, haematic or purulent	
Thickened and enhanced wall	Type III		Heterogenous in density	Purulent with inflammatory wall or abscess -formation	laparoscopic, or robot assisted surgery
	Type IV			Malignant transformation	Open Surgical resection, with or without Chemoradiotherapy

Types I though IV correspond to seminal vesicle cyst severity.
Type I and II have thin walls, are less severe and often asymptomatic.
Type III and IV have thicker walls, more severe, and usually require intervention.

ing specific imaging characteristics [8]. Differentiating seminal vesicle cysts from other cystic lesions, such as Müllerian duct cysts or Cowper duct cysts, is critical for diagnosis. This distinction relies on the cyst's location relative to the bladder neck [10].

The classification of seminal vesicle cysts in Zinner syndrome is crucial for clinical decision-making and is based on their imaging characteristics. The classification includes type I (simple cyst), type II (complex cyst), type III (cyst with concurrent infections or abscesses), and type IV (cyst with malignant transformation), and is described in Table 1 [11]. Accurate identification of these types on imaging assists in determining the most appropriate treatment strategy.

In ZS, ejaculatory duct obstruction can lead to infertility, with approximately 45% of affected men experiencing such outcome [2]. Although typically impacting only 1 ejaculatory duct, documented cases of azoospermia suggest more complex underlying mechanisms [12].

Treatment options for ZS include surgery, aspiration, and observation. Surgical intervention is generally recommended for symptomatic patients, while transurethral resection of the ejaculatory duct is preferred when infertility is the primary concern. Asymptomatic cysts can often be managed with regular follow-up [2].

This case aligns with the median age of diagnosis for ZS, as the patient presented with the classic triad of an ipsilateral

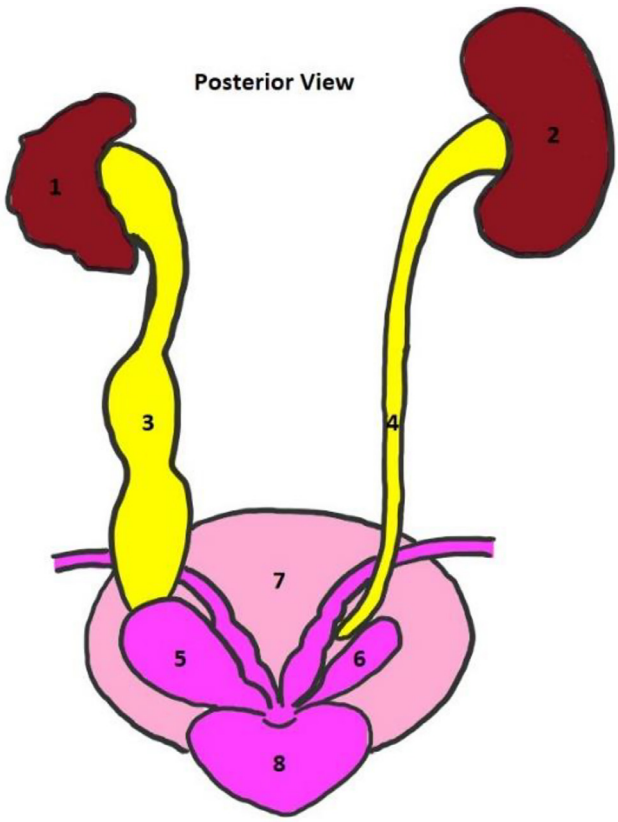

Fig. 2 – Schematic illustration depicting the anatomical features of the case. Panel 1 highlights the presence of a dysplastic left kidney, illustrating its abnormal development and hypoplasia, characteristic of renal agenesis. Panel 3 shows the enlarged and tortuous left ureter, which aberrantly inserts into the seminal vesicle rather than the bladder. Panel 5 presents the cystic enlargement of the left seminal vesicle, a direct consequence of the ectopic ureter draining into this structure, resulting in fluid accumulation and subsequent dilation.

seminal vesicle cyst, renal agenesis, and ectopic ureter. Notably, the left-sided occurrence is less common. Initial treatment with Tamsulosin improved symptoms, but laparoscopic excision was necessary due to the cyst's size and potential complications.

This case underscores the importance of high-resolution imaging for accurate diagnosis, particularly in patients with nonspecific symptoms. The balance between conservative and surgical management is essential to prevent complications, including infertility, in patients with ZS [2,13].

Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

Patient consent

Written informed consent was obtained from the patients for their anonymized information to be published in this article.

REFERENCES

[1] Sada F, Cekaj E, Saraci B, Shazi O, Al-Madani A, Jahanian S, et al. Challenging clinical presentation of Zinner syndrome. Radiol Case Rep 2023;18(1):256–9. doi:10.1016/j.radcr.2022.10.006.

[2] 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 (in eng). Ann Palliat Med 2021;10(2):2271–82. doi:10.21037/apm-20-1997.

[3] Dai R, Jiang F, Fan J, He D, Li L, Wu K. Clinicopathological and genetic features of Zinner's syndrome: two case reports and review of the literature," (in English). Front Urol, Case Rep 2023;3:2–5. doi:10.3389/fruro.2023.1257368.
[4] Kumar S, G KI, Khalil-Khan A, Arul Pitchai ADP, Sathiamoorthy R, Raju E. "Zinner syndrome," (in eng). Cureus 2022;14(11):e31308. doi:10.7759/cureus.31308.
[5] UKKA. Congenital anomalies of the kidneys and urinary tracts. The UK Kidney Association; London, United Kingdom: 2024. Available at: https://ukkidney.org/rare-renal/clinician/congenital-anomalies-kidneys-and-urinary-tracts#:~:text=Congenital%20anomalies%20of%20the%20kidneys%20and%20urinary%20tract%20(CAKUT)%20occur,of%20all%20anomalies%20diagnosed%20antenatally. Accessed January 10, 2024.
[6] Lin CC, Sheu JC, Tsai PS, Lee MD, Lin TH, Tsai JD. Zinner syndrome in children: clinical presentation, imaging findings, diagnosis, and outcome," (in eng). Pediatr Nephrol 2022;37(12):3075–84. doi:10.1007/s00467-022-05516-2.
[7] Cascini V, Di Renzo D, Guerriero V, Lauriti G, Chiesa PLelli. Zinner syndrome in pediatric age: issues in the diagnosis and treatment of a rare malformation complex," (in eng). Front Pediatr 2019;7:129. doi:10.3389/fped.2019.00129.
[8] Shah S, Patel R, Sinha R, Harris M. Zinner syndrome: an unusual cause of bladder outflow obstruction," (in eng). BJR Case Rep 2017;3(2):20160094. doi:10.1259/bjrcr.20160094.
[9] Hofmann A, Vauth F, Roesch WH. Correction: Zinner syndrome and infertility—a literature review based on a clinical case. Int J Impotence Res 2022;34(5):2–5. doi:10.1038/s41443-021-00476-x.
[10] Beyene E, Tadele E, Negassa M. Zinner syndrome: a first case report from Ethiopia. Radiol Case Rep 2023;18(1):86–90. doi:10.1016/j.radcr.2022.09.074.
[11] 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," (in eng). Andrologia 2020;52(1):e13397. doi:10.1111/and.13397.
[12] Avellino GJ, Lipshultz LI, Sigman M, Hwang K. Transurethral resection of the ejaculatory ducts: etiology of obstruction and surgical treatment options," (in eng). Fertil Steril 2019;111(3):427–43. doi:10.1016/j.fertnstert.2019.01.001.
[13] Kelly NP, Fuentes-Bonachera A, Shields WP, Cullen IM, Daly PJ. Long-term surveillance and laparoscopic management of Zinner syndrome. Case Rep Urol 2021;2021(1):6626511. doi:10.1155/2021/6626511.