BMC Urology

CASE REPORT

Open Access

A rare variant of zinner syndrome with ejaculatory duct cyst: case report and challenges in diagnosis and management

Guixing Tang[1], Yanhua Feng[1], Zhaohui Wang[1], Yu Yang[2], Yupin Liu[3], Zunguang Bai[1*] and Jun Pan[1*]

Abstract

Background Zinner syndrome (ZS) is a congenital malformation characterized by a triad of mesonephric (Wolffian) duct dysplasia, first identified by Zinner in 1914. The classical presentation of ZS includes unilateral renal hypoplasia or dysplasia, ipsilateral seminal vesicle cysts, and obstruction of the ejaculatory duct. This case presents a rare variant of the syndrome, where an ejaculatory duct cyst is observed instead of the typical ipsilateral seminal vesicle cyst. The ejaculatory duct cyst affected the vas deferens bilaterally, leading to bilateral atrophy or erosion of the seminal vesicle glands, and resulted in the absence of seminal fluid, ultimately causing azoospermia and infertility.

Case presentation Prior to surgery, the patient experienced a sensation of incomplete defecation, accompanied by mild anal distension. Two semen analyses revealed azoospermia, and magnetic resonance imaging/magnetic resonance urography indicated the absence of the left renal component and suggested the presence of a seminal vesicle cyst. It was hypothesized that the azoospermia resulted from compression of the contralateral ejaculatory duct by the seminal vesicle cyst on the affected side. Consequently, a decision was made to proceed with laparoscopic resection.During the surgical procedure, no seminal vesicle cyst was identified; however, an ejaculatory duct cyst was discovered, wherein the bilateral vasa deferentia converged without any alternative outlet. The cyst was subsequently resected. Postoperatively, the patient's clinical symptoms resolved, although the issue of infertility remained unaddressed.

Conclusion This case describes a rare Zinner syndrome variant where an ejaculatory duct cyst replaces the seminal vesicle cyst, leading to seminal vesicle atrophy, azoospermia, and infertility. An unreported variant was discovered during surgery, underscoring the importance of preoperative imaging. Laparoscopic removal alleviated symptoms but not infertility, indicating that assisted reproduction might be necessary for ZS-related azoospermia. This case expands knowledge of ZS variants and their impact on fertility.

Keywords Case report, Zinner syndrome, Ejaculatory duct cyst, Azoospermia

*Correspondence:
Zunguang Bai
zunguangbai@163.com
Jun Pan
Johnpcn@126.com

[1]Department of Urology, The Second Affiliated Hospital of Guangzhou University of Chinese Medicine, Guangzhou, China
[2]Department of Pathology, The Second Affiliated Hospital of Guangzhou University of Chinese Medicine, Guangzhou, China
[3]Department of Imaging, The Second Affiliated Hospital of Guangzhou University of Chinese Medicine, Guangzhou, China

Background

Zinner syndrome (ZS), initially characterized by Zinner in 1914, is a congenital anomaly involving a triad of mesonephric (Wolffian) duct dysplasia. This syndrome is defined by the presence of unilateral renal hypoplasia or dysplasia, ipsilateral seminal vesicle cysts, and ejaculatory duct obstruction [1]. During embryonic development, specifically between the 5th and 13th weeks, any factor that induces dysplasia of the distal middle renal canal can result in the absence of the associated ureter, renal dysplasia, and atresia of the ejaculatory canal [2].In this instance, the bilateral vasa deferentia were implicated alongside congenital unilateral absence of the kidney and ureter. The cyst identified was an ejaculatory duct cyst rather than a seminal vesicle cyst. Both seminal vesicle glands exhibited atrophy or erosion. This diagnosis was corroborated by intraoperative examination and postoperative pathological analysis.

Case presentation

A 26-year-old male patient presented with abdominal pain attributed to small calculi located in the right lower ureter. A color Doppler ultrasound examination of the urinary system revealed ZS, indicating the presence of a posterior cystic mass in the prostate and the absence of the left kidney. The patient reported a regular sexual history spanning five years, during which he employed the coitus interruptus method for contraception. Over the past year, there has been a gradual decrease in ejaculatory volume, with the semen appearing clear. In the preceding month, only a minimal amount of transparent fluid was emitted during sexual intercourse.Ejaculation occurred without evident pain or discomfort, and no blood was detected in the seminal fluid. The patient reported no urinary discomfort; however, he experienced a sensation of incomplete defecation accompanied by mild anal distension. His medical records indicated no history of cryptorchidism, delayed pubertal development, or scrotal-perineal discomfort. He denied any family history of related conditions or previous surgical interventions. Bilateral testicular development was normal, although there was mild expansion of the bilateral epididymis and vasa deferentia. A digital rectal examination identified a cystic mass, with no tenderness detected in the prostate region. Two semen analyses confirmed absolute azoospermia, with semen volume ranging from 0.1 to 0.2 ml. The concentrations of neutral alpha-glucosidase, citric acid, fructose, and zinc were measured at 0.06 mU/time, 8.42 μmol/time, 0.05 g/L, and 4.59 mmol/L, respectively. A scrotal ultrasound examination indicated a left testicular volume of 10.1 mL and a right testicular volume of 10.4 mL. Peripheral blood karyotype analysis demonstrated a 46,XY chromosomal pattern. The sex hormone profile was as follows: estradiol (E2) at 126 pmol/L, luteinizing hormone (LH) at 6.14 IU/L, testosterone at 17.7 nmol/L, prolactin at 586 mIU/L, and follicle-stimulating hormone (FSH) at 6.44 IU/L. Abdominal magnetic resonance imaging and magnetic resonance urography (MRI/MRU) revealed the absence of the left kidney and left ureter (Fig. 1A). Additionally, a round cystic lesion was observed in the posterior aspect of the prostate gland and seminal vesicle.The lesion exhibited a uniform signal, with no enhancement observed on the contrast-enhanced scan, and its boundary appeared well-defined. The lesion measured 5.9×6.2×5.0 cm (Fig. 1B). However, the demarcation between the leading edge of the lesion and the seminal vesicle was indistinct, with notable compression of the proximal ejaculatory duct (Fig. 1C). Clinically, the diagnosis suggested Zinner syndrome (ZS) with azoospermia. The azoospermia was attributed to the compression of the seminal vesicle cyst on an otherwise healthy ejaculatory duct. Nonetheless, intraoperative findings did not corroborate our preoperative assessment.

Following comprehensive communication with the patient and his family, they opted for surgical intervention. The surgical procedures undertaken included endoscopic examination via seminal vesicle endoscopy and cystoscopy, retrograde catheterization of the right ureter, and laparoscopic cyst resection. Cystoscopy revealed a smooth urethral mucosal surface with no significant hyperplasia of the mass, which was visible on both sides of the verumontanum submucosa, exhibiting suspicious black punctate pigmentation (Fig. 1D). Notably, no bilateral ejaculatory duct openings were observed upon increased water pressure.The bladder neck was positioned slightly higher, and there was a noticeable elevation in the trigone region of the bladder. The right ureteral orifice was clearly visible; however, the left ureteral orifice could not be identified (Fig. 1E). The mucous membrane of the bladder wall appeared smooth. A right external ureteral stent was placed during the procedure to prevent inadvertent injury while excising the cyst. Laparoscopic examination revealed a raised cystic mass located inferior to the bladder, with both vasa deferentia appearing normal, although the right vas deferens was slightly dilated.The cyst's surface was meticulously dissected from its base, ensuring the preservation of the bilateral vascular and neural structures. The dimensions of the cyst were 5×5×6 cm, with the bilateral vasa deferentia converging into the cyst from the left and right anterolateral aspects (Fig. 1F). The cyst's base appeared to originate from the ampullae of the bilateral vasa deferentia. Palpation revealed that the left seminal vesicle was significantly atrophied (Fig. 1G), while the right seminal vesicle was not accessible. The right lateral wall of the cyst exhibited a focal patch-like structure resembling seminal vesicle tissue, which had fused with the cystic

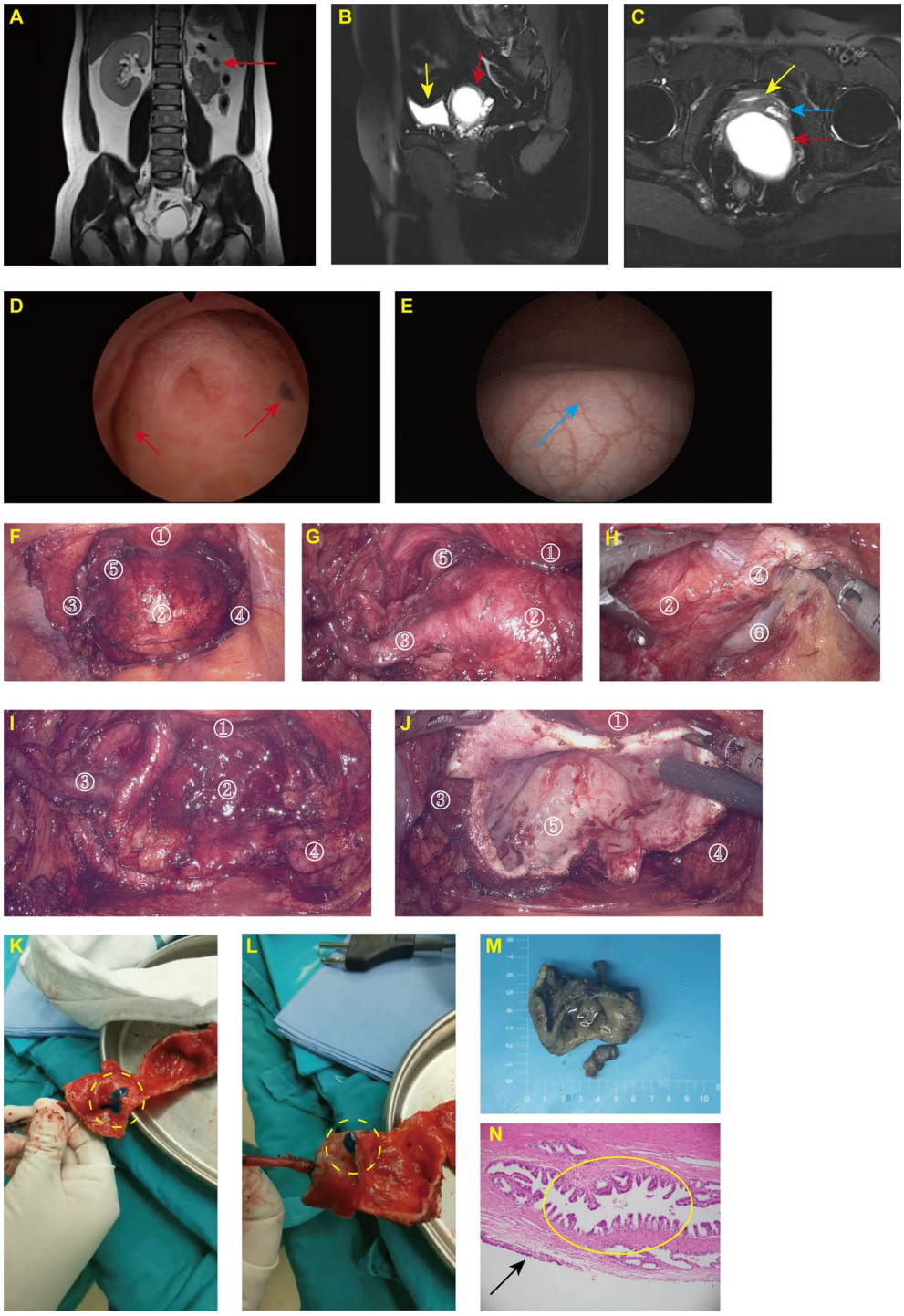

Fig. 1 **A**: Coronal T2 MRI: The left kidney and left ureter were absent (red arrow); **B**: sagittal view T2 MRI: the round cystic lesion (red arrow); the bladder (yellow arrow); **C**: axial T2 MRI: the round cystic lesion (red arrow); the bladder (yellow arrow); the left seminal vesicle can only be seen at this level and it had diminished in size (blue arrow); **D**: The melanin deposits on both sides of the verumonis (red arrow); **E**: The left ureteral orifice could not be explored (blue arrow); **F-H**: ①The bladder, ②cyst, ③the right vas deferens,④ the left vas deferens, ⑤the left shrunken seminal vesicle, ⑥which was similar to the seminal vesicle structure on the right side wall of the cyst; **I-J**: ①The bladder, ②the cyst was empty,③ the left vas deferens, ④the right vas deferens, ⑤the cyst was opened with no visible exit; **K-L**: Methylene blue was injected into both sides of the proximal vas deferens and methylene blue was seen draining from the inner surface of the cyst (yellow circle); **M-N**: Postoperative pathology. **A**: Gross specimen; **B**: cyst wall (black arrow). A small amount of atrophied seminal vesicle tissue was observed in the focal area around the cyst wall (red circle)

wall, rendering separation unfeasible (Fig. 1H). Approximately 100 ml of yellowish-brown, cloudy, and viscous fluid was aspirated from the sac using a needle.

Subsequent routine semen and seminal plasma biochemical analyses revealed a substantial presence of non-viable spermatozoa, accompanied by elevated α-glucosidase enzyme activity (3,358.80 mU/time), a normal citric acid concentration, and a negative result for fructose.The cyst was dissected in a longitudinal manner, revealing a thicker cyst wall, while the base was not examined upon opening. On the right side of the cystic cavity, focal and patch-like structures were observed, which had fused with the cystic wall and were inseparable from it. The bilateral vasa deferentia converged into the cyst, which lacked any additional openings (Fig. 1I-J). The typical periampullary structure of the bilateral vasa deferentia was absent, consistent with the patient's bilateral ejaculatory duct obstruction.This cyst was not a seminal vesicle cyst resulting from unilateral obstruction of the ejaculatory duct, as is characteristic of Zinner syndrome (ZS). Instead, it was identified as an ejaculatory duct cyst formed by the fusion of the two ducts. The absence of seminal vesicle fluid in the cystic fluid impeded semen liquefaction and sperm activation. Consequently, natural conception was not feasible despite treatment efforts. Therefore, it was decided to resect the cyst and perform a left seminal vesiculotomy along with a bilateral vasectomy. Methylene blue was administered into the proximal vas deferens on both sides of the postoperative specimens, verifying that the orifices of both vas deferens were situated within the cysts, with no additional openings identified within the cysts (Fig. 1K-L). Consequently, the patient's bilateral ejaculatory ducts coalesce to form an ejaculatory duct cyst lacking an outlet, ultimately resulting in azoospermia.

Two tissue specimens were submitted for postoperative pathological examination (Fig. 1M-N). The first specimen comprised capsular wall tissue, where the majority of the inner capsule wall lacked epithelial lining; however, a localized region exhibited columnar epithelium coverage, alongside a minor presence of atrophic seminal vesicle tissue adjacent to the cyst wall. Notably, the absent right seminal vesicle was identified within the right lateral wall of the cyst. The second specimen consisted of free seminal vesicle tissue.Postoperative pathological analysis corroborated the intraoperative findings. The patient experienced an immediate restoration of erectile function following the surgical procedure and resumed his preoperative sexual activity level after one month of follow-up. Over a six-month follow-up period, the patient reported no symptoms of discomfort. His erectile function remained normal, and he maintained the same level of sexual activity and satisfaction as prior to the surgery, although he noted a reduced volume of clear fluid in his ejaculate.

Discussion and conclusions

This case study delineates three uncommon variations of Zinner syndrome: atrophy of the left seminal vesicle accompanied by the absence of the ipsilateral kidney and ureter, an underdeveloped right seminal vesicle adhered to the cyst wall, and fused ejaculatory ducts forming a singular cyst. While individual anomalies similar to these have been reported in the literature [3–5], the concurrent presence of these three variations has not been previously documented, indicating a potentially unique variant of Zinner syndrome.

The exact mechanism through which ZS leads to infertility is not yet fully understood [6]. Certain studies associate it with oligospermia or azoospermia [7], whereas others propose that obstruction of the ejaculatory duct may induce the production of anti-sperm antibodies or other inhibitory factors [8–11]. Although deferentovesiculography (DVG) was previously considered the diagnostic gold standard [12–13], its use has declined due to its invasive nature; instead, enhanced computed tomography (CT) and magnetic resonance imaging (MRI) are now more commonly employed [14, 15].

Various treatment modalities, including transurethral resection of the ejaculatory duct (TURED), have demonstrated variable outcomes. Some studies report enhancements in semen quality and increased pregnancy rates [16, 17], whereas others emphasize the occurrence of complications such as urinary and ejaculatory dysfunction [18]. Alternative interventions, such as open surgery, laparoscopic, and robotic techniques, have been investigated [4, 19, 20]; however, their efficacy in addressing infertility remains constrained [21]. Consequently, assisted reproductive technology (ART) is progressively regarded as the principal strategy for achieving fertility in these patients [22].

Although initially presumed to be a standard case of Zinner syndrome, the findings indicate an atypical developmental variant. The observed history of decreasing semen volume and the fusion of the right seminal vesicle with the cyst suggest the potential for acquired inflammatory changes. In patients with complete obstructive azoospermia, diagnostic vasography (DVG) continues to be instrumental in precisely identifying obstructions and informing treatment strategies, especially in scenarios where surgical intervention may not rectify infertility.

Patient perspective and follow-up

The patient articulated that prior to surgery, his primary objective was the excision of the cyst to address his infertility concerns. He did not anticipate receiving a diagnosis of atypical Zinner syndrome but has since reconciled

with it. During subsequent follow-up, he noted the resolution of his preoperative symptoms, which included the sensation of incomplete defecation and mild anal distension, following the surgical intervention. Despite this symptomatic improvement, he acknowledged the potential persistence of infertility and proactively engaged in assisted reproductive techniques. These efforts proved successful, culminating in a pregnancy with his partner, which brought them significant relief and joy.

Abbreviations

CT	Computed tomography
DVG	Deferento-vesiculography
MRI	Magnetic resonance imaging
MRU	Magnetic resonance urography
TURED	Transurethral resection of the ejaculatory duct
ZS	Zinner syndrome

Acknowledgements
Thanks to Pathology and Radiology for providing accurate information.

Author contributions
GT, YF, ZW, and ZB conceived the report; GT, YF and ZB operated on the patient; YY provided and interpreted pathological sections; YL provided the image data; ZB and JP made substantial contributions to conception and design of the report and agreed to be accountable for all aspects of the work. All authors read and approved the final manuscript. Each author contributed important intellectual content during manuscript drafting or revision.

Funding
This work was supported by grants from Traditional Chinese Medicine Bureau of Guangdong Province (NO.20222079).

Data availability
Data is provided within the manuscript.

Declarations

Ethics approval and consent to participate
This case report was reviewed and approved by the Ethics Committee of Guangdong Provincial Hospital of Chinese Medicine. (No.G2022-04).

Consent for publication
The written informed consent to publish this information has been obtained from the study participant.

Competing interests
The authors declare no competing interests.

Received: 15 July 2024 / Accepted: 19 November 2024
Published online: 04 December 2024

References

1. Cascini V, Renzo DD, Guerriero V et al. Zinner Syndrome in Pediatric Age: issues in the diagnosis and treatment of a rare malformation Complex[J]. Frontiers in Pediatrics. 2019;7. https://doi.org/10.3389/fped.2019.00129
2. Schukfeh N, Kuebler JF, Schirg E, et al. Dysplastic kidney and not renal agenesis is the commonly associated anomaly in infants with seminal vesicle cyst[J]. BJU Int. 2010;103(6):816–9. https://doi.org/10.1111/j.1464-410X.2008.08072.x.
3. Cito G, Gemma L, Giachini C et al. Sperm retrieval by conventional testicular sperm extraction for assisted reproduction in patients with Zinner syndrome[J].Clinical and Experimental Reproductive Medicine. 2021;48(1):85–90. https://doi.org/10.5653/cerm.2020.03769.
4. Razdan S, Kryvenko ON, et al. Robotic-assisted laparoscopic vesiculectomy in a patient with atypical Zinner syndrome presenting with large cyst involving bilateral seminal vesicles and vasa deferentia[J]. Urol Case Rep. 2018;18:79–81. https://doi.org/10.1016/j.eucr.2018.03.014.
5. Van Blom dOD, Bangma JHM. C, Diagnosis and management of seminal vesicle cysts associated with ipsilateral renal agenesis: a pooled analysis of 52 cases.[J]. European Urology. 1998;33(5):433–40. https://doi.org/10.1159/000019632
6. Manousakas T, Kyriakou G, Serafenitinides E, et al. Partial vesiculectomy in an infertile man with seminal vesicle cyst, ipsilateral renal agenesis, and cryptorchidism. Urology. 2002;59(602):24–6. https://doi.org/10.1016/S0090-4295(01)01675-2.
7. Ferna´ndez FJC, Cardoso JVG, Rubio RM, et al. Quiste Gigante De vesı´cula seminal asociado con agenesia renal ipsilateral. Actas Urol Esp. 2002;26:218–23. https://doi.org/10.1016/S0210-4806(02)72761-7.
8. Hendry WF, Parslow JM, Parkinson MC et al. Unilateral testicular obstruction: orchidectomy or reconstruction? Hum Reprod 1994,9:463–70. https://doi.org/10.1093/oxfordjournals.humrep.a137985.
9. Hendry WF. Disorders of ejaculation:congenital,acquired and functional[J]. Br J Urol. 1998;82:331–41. https://doi.org/10.1046/j.1464-410X.1998.00758.x.
10. Pereira BJ, Sousa L, Azinhais P et al. Zinner's syndrome:an up-to-date review of the literature based on a clinical case[J]. Andrologia. 2010;41(5):322–30. https://doi.org/10.1111/j.1439-0272.2009.00939.x
11. Pace G, Galatioto GP, Guala` L, et al. Ejaculatory duct obstruction caused by a right giant seminal vesicle with an ipsilateral upper urinary tract agenesia: an embryologic malformation. Fertil Steril. 2008;89:390–4. https://doi.org/10.1016/j.fertnstert.2007.03.009.
12. Purohit RS, Wu DS, Shinohara K et al. A prospective comparison of 3 diagnostic methods to evaluate Ejaculatory Duct Obstruction[J]. J Urol. 2004;171(1):232–6. https://doi.org/10.1097/01.ju.0000101909.70651.d1
13. Furtado A. Three Cases of Cystic Seminal Vesicle Associated with unilateral renal Agenesis[J]. Br J Urol. 1973;45. https://doi.org/10.1111/j.1464-410X.1973.tb06817.x
14. Narlawar RS, Hanchate V, Raut A, et al. Renal agenesis and seminal vesicle cyst. J Ultrasound Med. 2003;22:225–8. https://doi.org/10.7863/jum.2003.22.2.225.
15. Jiang XS, Wang HJ, Lin JH et al. Zinner's syndrome: clinical features and imaging diagnosis[J]. Asian J Androl. 2018;20:316–7. https://doi.org/10.4103/1008-682X.210295
16. Netto NR, Esteves SC, Neves PA. Transurethral resection of partially obstructed ejaculatory ducts: seminal parameters and pregnancy outcomes according to the etiology of obstruction. J Urol. 1998;159:2048–53. https://doi.org/10.1097/00005392-199806000-00086
17. Aghaways I, Ahmed SM. Endourologic Intervention for Management of Infertility in a man with Zinner Syndrome resulting in a natural pregnancy. [J] J Endourology Case Rep. 2016;2(1):71–3. https://doi.org/10.1089/cren.2016.0010
18. Edward EC, Anoop MM, Leonard HB, et al. Laparoscopic management of congenital seminal vesicle cysts associated with ipsilateral renal agenesis. J Urol. 2002;167:1263–7. https://doi.org/10.1016/S0022-5347(05)65278-0.
19. Han P, Dong Q, Shi M, et al. Seminal vesicle cyst and ipsilateral renal agenesis: laparoscopic approach. Arch Androl. 2007;53:285–8. https://doi.org/10.1080/01485010701569882.
20. Cito G, Sforza S, Gemma L et al. Infertility case presentation in Zinner syndrome: can a long̊asting seminal tract obstruction cause secretory testicular injury?[J]. Andrologia. 2019;51(6). https://doi.org/10.1111/and.13436
21. Hofmann A, Vauth F, Roesch WH. Zinner syndrome and infertilitya literature review based on a clinical case[J]. Int J Impot Res. 2021;33:191–5. https://doi.org/10.1038/s41443-021-00476-x
22. Manousakas T, Kyriakou G, Serafenitinides E et al. Partial vesiculectomy in an infertile man with seminal vesicle cyst, ipsilateral renal agenesis, and cryptorchidism.Urology. 2002;59:60224–26.https://doi.org/10.1016/S0090-4295(01)01675–2.

Publisher's note