

Zinner's Syndrome: Case Report of a Rare Maldevelopment in the Male Genitourinary Tract

Zinner Sendromu: Erkek Ürogenital Sistemde Nadir Görülen Bir Gelişim Bozukluğu Olgusu

© Célia Sousa¹, © Inês Portugal Teixeira¹, © Sofia Helena Ferreira², © Ana Teixeira³, © Sílvia Costa Dias¹

¹Saint John Hospital, Clinic of Radiology, Porto, Portugal

²Saint John Hospital, Clinic of Pediatrics, Porto, Portugal

³Centro Materno-infantil do Norte, Clinic of Pediatric Nephrology, Porto, Portugal

Abstract

Zinner's syndrome is a rare embryologic anomaly of the distal portion of the mesonephric duct comprising a triad of ipsilateral renal agenesis, seminal vesicle cyst and ejaculatory duct obstruction. We report ultrasound and magnetic resonance imaging findings of this rare developmental anomaly involving the mesonephric duct, in an asymptomatic 17-year-old boy who would probably remain undiagnosed until later age. The patient presented with no specific symptoms and was incidentally diagnosed on imaging exams.

Keywords: Zinner's syndrome, Seminal vesicles, Ejaculatory duct, Renal agenesis

Öz

Zinner sendromu; ipsilateral renal agenezi, seminal vezikül kisti ve ejakülatör kanal obstrüksiyon triadını içeren, mezonefrik kanalın distal kısmında nadir görülen, embriyolojik bir anomalidir. Muhtemelen daha sonraki yaşlara kadar tanı konulmadan kalacak olan 17 yaşındaki asemptomatik bir erkek çocukta, mezonefrik kanalı tutan bu nadir gelişimsel anomalinin ultrason ve manyetik rezonans görüntüleme bulgularını sunuyoruz. Spesifik bir semptomu bulunmayan hastaya, görüntülenme muayenelerinde tesadüfen tanı konulmuştur.

Anahtar Kelimeler: Zinner sendromu, Seminal vezikül, Ejakülatör kanal, Renal agenezi

Introduction

Zinner's syndrome is a rare embryologic anomaly of the distal portion of the mesonephric duct, responsible for the emergence of the ureteric bud, vas deferens, ejaculatory duct, seminal vesicle, hemitrigone, epididymis and paradidymis, during the 4th and the 13th gestational week. The common origin of the mesonephric duct and the ureteric bud, which will form the definitive adult kidney, is responsible for the associated maldevelopment of the genital and urinary tracts, including atresia of the ejaculatory duct, obstruction of the seminal vesicles with formation of cysts and ipsilateral renal agenesis or dysplasia (1,2,3,4). More rarely, the ureteric bud can arise in a more cranial position of the mesonephric duct, leading to a distal ureteric bud with anomalous drainage, for example into

the vas deferens, seminal vesicle, ejaculatory duct, bladder neck and urethra.

This syndrome is considered to be the male version of Mayer-Rokitansky-Kuster-Hauser syndrome (5). The diagnosis is usually made between the 2nd and the 5th decades of life, concomitant with the period of maximum sexual activity, when cysts start to become more apparent (1). Patients usually present with multiple and unspecific symptoms, and the diagnosis is often made by imaging studies. The clinical manifestations include voiding symptoms (dysuria, frequency, urgency), haematuria, perineal or scrotal pain, recurrent urinary tract infection, epididymitis, prostatitis, painful ejaculation, haemospermia, and infertility. Small cysts in the seminal vesicles, less than 5 cm in size, are usually asymptomatic and often detected incidentally (4,5,6,7). We report ultrasound (US) and magnetic resonance imaging

Correspondence: Célia Sousa MD, Saint John Hospital, Clinic of Radiology, Porto, Portugal

Phone: +351-225512100 **E-mail:** celia.sousa17@gmail.com **ORCID-ID:** orcid.org/0000-0001-6876-9906

Received: 23.10.2018 **Accepted:** 30.11.2018

Cite this article as: Sousa C, Teixeira IP, Ferreira SH, Teixeira A, Dias SC. Zinner's Syndrome: Case Report of a Rare Maldevelopment in the Male Genitourinary Tract. J Urol Surg 2019;6(1):65-67

©Copyright 2019 by the Association of Urological Surgery / Journal of Urological Surgery published by Galenos Publishing House.



(MRI) findings of this extremely rare developmental anomaly involving the mesonephric duct in a 17-year-old boy who would probably remain undiagnosed until later age. The patient presented with no symptoms related to this anomaly and was incidentally diagnosed on imaging exams.

Case Presentation

A 17-year-old boy with a medical history of Fabry disease and right renal agenesis was sent to our department for routine US evaluation. He was asymptomatic, with no abdominal or genitourinary complaints. External genitalia were normally developed and the vas deferens was palpable bilaterally. Blood analysis, renal function and urinalysis were also normal.

We performed US that revealed the presence of a multilocular cystic mass located in the right side of the pelvis inferolateral to the urinary bladder. The mass had internal low level echoes, probably due to hemorrhage or increased concentration of proteinaceous content. US also showed absence of kidney in the right renal fossa (Figure 1). With the US findings, the diagnosis of Zinner's syndrome was considered.

Pelvic MRI was subsequently performed to confirm that the cystic mass was arising from the seminal vesicle. MRI showed a multilocular cystic lesion replacing the right seminal vesicle and measuring 74x56 mm. The multiloculated seminal vesicle cyst presented hypersignal on T2 and T1-weighted images, which reflects hemorrhage or increased proteinaceous content. T1 post-contrast and subtraction images showed lack of contrast enhancement. Additionally, a tubular structure with a superior blind-ending was also apparent, coursing along the right iliac

vessels and draining into the right seminal vesicle, consistent with ectopic atretic ureter (Figure 2). On imaging findings, no clearly dilation of the vas deferens was found along its path.

We found no association in the literature between Zinner's syndrome and Fabry disease.

At present, the patient remains asymptomatic, on conservative management and with periodic clinical follow-up.

Oral informed patient consent for publication has been obtained.

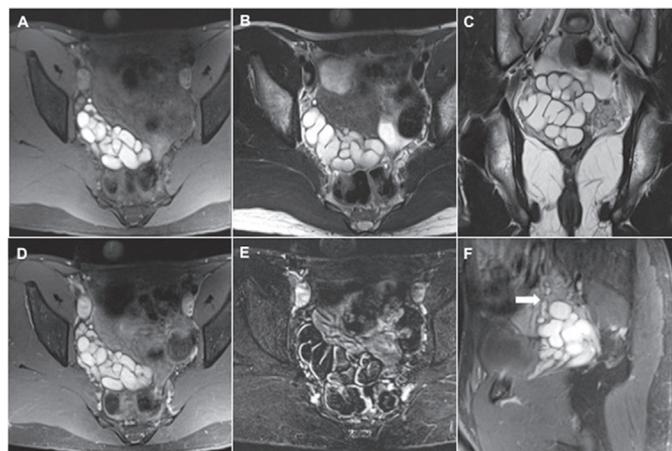


Figure 2. A) Axial T1W shows a hyperintense multiloculated right seminal vesicle cyst, reflecting probably increased concentration of proteinaceous fluid or eventually haemorrhage, B) axial and, C) coronal T2W magnetic resonance imaging demonstrate multiple cystic intercommunicating hyperintense structures. The lesion measures 74x56 mm, D) axial T1W magnetic resonance imaging post-contrast and, E) subtraction images show lack of contrast enhancement, F) parasagittal T1W magnetic resonance imaging showing the tubular remnant ureter (arrow) communicating with the right seminal vesicle cyst

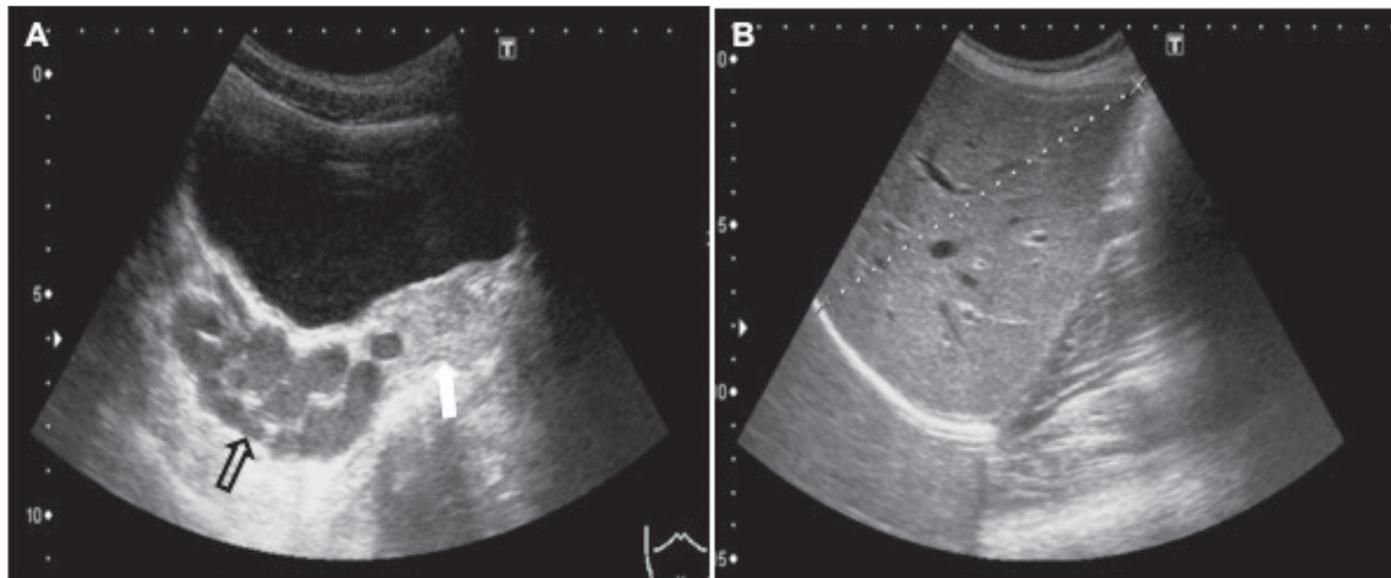


Figure 1. A) Pelvic ultrasound shows a multiloculated cystic structure (open arrow) in the right side of the pelvis inferolateral to the urinary bladder and adjacent to the prostate (white arrow). The mass has low level internal echoes, resulting from haemorrhage or increased proteinaceous content; B) absent right kidney in the renal fossa

Discussion

Zinner's syndrome is a rare malformation affecting the seminal vesicle and the upper urinary tract simultaneously. The diagnosis is usually made between the 2nd and the 5th decades of life due to enlargement of the seminal vesicle cysts (at least 5 cm to induce symptoms) (1,2,3,4). Most commonly, patients present with genitourinary symptoms and perineal pain that was not our case probably due to early diagnosis.

In the evaluation of patients with suspected mesonephric duct development failure, US can be very useful for the diagnosis, detecting an anechoic cystic mass in the pelvic fossa, but it can be more limited to study the rest of the pelvic smaller anatomic structures and their relationships. MRI offers anatomic details of the different pelvic structures, leading to a definitive diagnosis and is also of utmost importance for surgical planning. On MRI, seminal vesicle cysts are in a characteristic periprostatic and paramedian location, with bright signal on T2-weighted images, variable signal intensity on T1-weighted images, depending on the amount of protein or blood content, and no contrast enhancement. MRI may also reveal an ectopic ureter draining into the seminal vesicle, which is often difficult to identify with other imaging techniques.

Differential diagnosis involves several cystic diseases of other pelvic organs, including prostatic utricle cysts, ejaculatory duct cysts, prostatic cysts, diverticula of ampulla of vas deferens, ureteroceles, and abscess. Cyst location and other developmental abnormalities (renal agenesis or anomalies of the external genitalia) help to make the differential diagnosis (4,8).

Treatment of the seminal vesicle cysts is determined according to symptom existence. Symptomatic seminal vesicle cysts may require interventional treatment, which may be by minimally invasive surgery, or an approach through natural orifices (transrectal aspiration and transurethral resection), or open surgery. Minimally invasive approach with laparoscopic surgery and most recently with robotic-assisted approach has gained substantial acceptance and is the preferential method in most cases. Transrectal and transurethral approaches portend the risk of recurrence and open surgery is only reserved for recurrent or complex cases (9).

In conclusion, congenital anomalies of the male urogenital tract may not be considered by clinicians, especially when patients present with no precise symptoms. Familiarity with the imagiological findings of this anomaly is essential to make a

prompt diagnosis, with MRI proving the best soft tissue contrast. In this case, concomitant follow-up and screening, as well as genetic counseling during adolescence and adult life are also advisable.

Ethics

Informed Consent: Oral informed patient consent for publication has been obtained.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: C.S., S.H.F., A.T., Concept: C.S., I.P.T., S.C.D., Design: C.S., I.P.T., Data Collection or Processing: C.S., I.P.T., S.H.F., A.T., S.C.D., Analysis or Interpretation: C.S., S.C.D., A.T., Literature Search: C.S., I.P.T., Writing: C.S., S.C.D., A.T.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

References

1. Pereira BJ, Sousa L, Azinhais P, Conceição P, Borges R, Leão R, Brandão A, Temido P, Retroz E, Sobral F. Zinner's syndrome: an up-to-date review of the literature based on a clinical case. *Andrologia* 2009;41:322-330.
2. Livingston L, Larsen CR. Seminal vesicle cyst with ipsilateral renal agenesis. *AJR Am J Roentgenol* 2000;175:177-180.
3. King BF, Hattery RR, Lieber MM, Berquist TH, Williamson Jr B, Hartman GW. Congenital cystic disease of the seminal vesicle. *Radiology* 1991;178:207-211.
4. Fiaschetti V, Greco L, Giuricin V, De Vivo D, Di Caprera E, Di Trapano R, Castellani F, Floris R. Zinner syndrome diagnosed by magnetic resonance imaging and computed tomography: role of imaging to identify and evaluate the uncommon variation in development of the male genital tract. *Radiol Case Rep* 2017;12:54-58.
5. Mehra S, Ranjan R, Chandra Garga U. Zinner syndrome - a rare developmental anomaly of the mesonephric duct diagnosed on magnetic resonance imaging. *Radiol Case Rep* 2016;11:313-317.
6. Slaoui A, Regragui S, Lasri A, Karmouni T, El Khader K, Koutani A, Ibn Attya A. Zinner's syndrome: report of two cases and review of the literature. *Basic Clin Androl* 2016;26:10.
7. Rappe BJM, Meuleman EJH, Debruyne FMJ. Seminal vesicle cyst with ipsilateral renal agenesis. *Urol Int* 1993;50:54-56.
8. Sundar R, Sundar G. Zinner syndrome: an uncommon cause of painful ejaculation. *BMJ Case Rep* 2015:2015.
9. Kiremit MC, Acar O, Sag AA, Koseoglu E, Kilic M, Kordan Y, Balbay MD. Minimally invasive management of Zinner's syndrome with same-session robot-assisted seminal vesiculectomy and ipsilateral nephroureterectomy using a single geometry of trocars. *J Endourol Case Rep* 2018;4:186-189.