Print ISSN: 2321-6379
Online ISSN: 2321-595X

Case Report

Zinner Syndrome – A Case Report

Nrudeep Chaitanya Kada[1], Kalaichezhian Mariappan[2]

[1]Post Graduate Resident, Department of Radio-diagnosis, Sree Balaji Medical College and Hospital, Chennai, Tamil Nadu, India, [2]Professor, Department of Radio-diagnosis, Sree Balaji Medical College and Hospital, Chennai, Tamil Nadu, India

Abstract

Zinner syndrome is a rare congenital malformation with the distal Wolffian duct anomaly in males. The triad of unilateral renal agenesis, ipsilateral seminal vesicle cyst, and ipsilateral ejaculatory duct obstruction characterizes it. Clinically, patients present with non-specific symptoms such as dysuria, ejaculatory disorders, and hypogastric or perineal pain. The diagnostic modalities include imaging techniques such as ultrasound scans, computed tomography, and magnetic resonance imaging (MRI). However, MRI is the standard gold technique for the confirmation of diagnosis. Herein, we present the case of a 14-year-old male patient who complained of vague lower abdominal pain and dysuria. Initially, an abdominal ultrasound revealed an absent right kidney and an ipsilateral seminal cyst. However, further MRI confirmed the diagnosis.

Keywords: Zinner syndrome, Renal agenesis, Dysuria, Ejaculatory disorders, Case report, Urogenital anomaly

INTRODUCTION

Zinner syndrome is a rare congenital condition characterized by a typical triad of seminal vesicle cyst, ipsilateral renal agenesis, and ejaculatory duct obstruction.[1] Conventionally, patients present in the second or third decade of life. Infertility is one of the most serious compliations.[2] Symptoms tend to manifest with the beginning of sexual activity due to the accumulation of seminal fluid in the seminal vesicles.[3] Clinically, patients present with nonspecific symptoms such as epididymitis, perineal discomfort, abdominal pain, frequent dysuria, infertility, and seminal vesical abscess so that the diagnosis may be delayed or missed until the beginning of sexual activity.[4] Zinner syndrome can sometimes be associated with other tumours or anomalies. Various radiological investigations, including ultrasonography (USG), computed tomography (CT), and magnetic resonance imaging (MRI), help in aiding the diagnosis, but MRI is the investigation of choice for complete imaging of the anatomy. Most cases need surgical intervention, but a few can be treated conservatively.

Access this article online

www.ijss-sn.com

Month of Submission : 12-2022
Month of Peer Review : 01-2023
Month of Acceptance : 01-2023
Month of Publishing : 02-2023

CASE REPORT

A 14-year-old male patient complained of vague lower abdominal pain and dysuria. On general examination, the vitals were found to be stable, and no significant abnormality was found on the systemic examination. The appearance of external genitalia was normal. Blood picture and urine examination were within normal limits. The patient was referred to the radiology department for further management. The abdominal ultrasound revealed an absent right kidney and an ipsilateral seminal cyst. For further clarification, an MRI of the whole abdomen was preferred over a CT scan to avoid exposure to excessive radiation to the paediatric patient. MRI abdomen revealed an absent right kidney (Figure 1) and a cystic lesion at the right posterolateral aspect of the urinary bladder. A fluid debris level was noted within the cystic lesion, which was suggestive of the seminal vesical cyst (Figure 2). There was compression on the adjacent urinary bladder. The right seminal vesicle was over-distended. The right ureter was moderately dilated with the upper blind end (Figure 3). The right ejaculatory duct was found to be obstructed by a seminal cyst from above. The right ureter appeared to drain into the right ejaculatory duct (Figure 4).

DISCUSSION

Zinner syndrome is a rare congenital malformation with the distal Wolffian duct anomaly in males, characterized by

Corresponding Author: Dr. Nrudeep Chaitanya Kada, Post Graduate Resident, Department of Radio-diagnosis, Sree Balaji Medical College and Hospital, Chennai, Tamil Nadu, India.

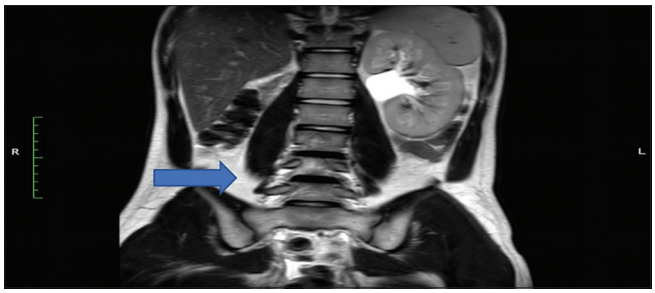

Figure 1: T2-weighted coronal section image showing absent right kidney

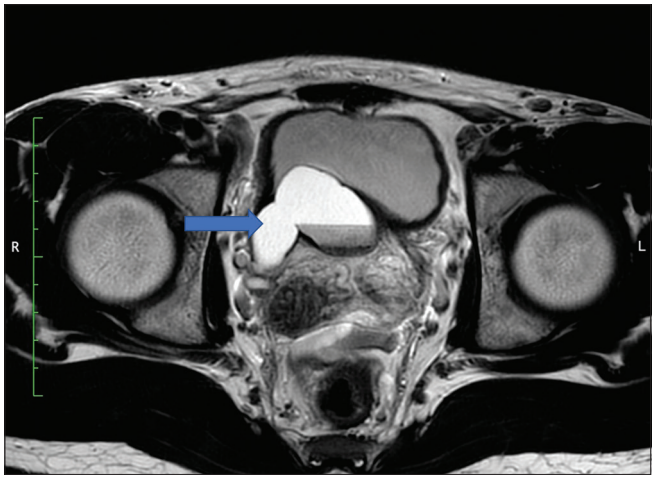

Figure 2: T2-weighted axial section image showing a lobulated cystic lesion at a right posterolateral aspect of the urinary bladder. The cystic lesion shows a fluid debris level, suggesting a seminal vesicle cyst. The cyst is causing compression on the adjacent urinary bladder

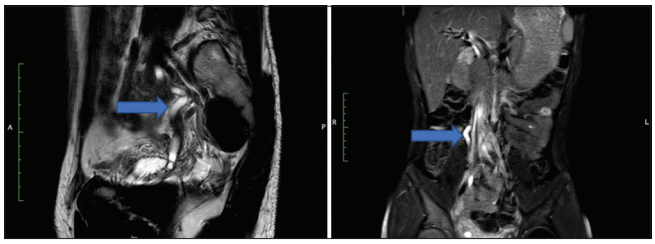

Figure 3: T2-weighted sagittal and a coronal image showing a moderately dilated right ureter with the lumen's upper blind-ending and hyperintense signals

a triad of seminal vesicle cyst, ipsilateral renal agenesis, and ejaculatory duct obstruction, which was first discovered in 1914.[4] Patients present with painful urination, increase urge and frequency to urinate, inflammation of the prostate gland, perineal or scrotal pain, epididymitis, and painful ejaculation. The diagnosis of this syndrome is delayed or missed, because symptoms appear with the beginning of a sexual activity.

Imaging plays an important role in the diagnosis of Zinner syndrome. Different imaging modalities include abdominal ultrasonography, abdominal CT scan, and MRI scan. Cystic

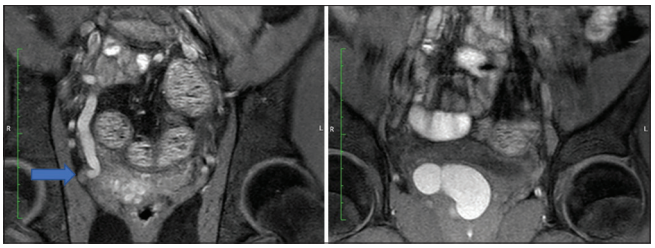

Figure 4: T1-weighted coronal image showing dilated right ureter with ectopic insertion into the right seminal vesicle duct/ ejaculatory duct

lesions are seen on the seminal vesicles. The size and location of the lesions can be estimated with the help of abdominal ultrasonography. The ultrasound also reveals the integrity of the bladder and the lack of the ipsilateral kidney.[5]

Analyses of the cyst contents and authentication of the diagnosis are done with the help of MRI. MRI is considered to be the gold standard in confirming the diagnosis. Cystic lesions are normally hypointense on T1 and hyperintense on T2. Blockage of the ejaculatory ducts can be authenticated, and a complete analysis of the glands can be carried out.[5]

Our case was of a 14-year-old male patient who complained of vague lower abdominal pain and dysuria. Blood and urine examinations were within normal limits. However, the abdominal ultrasound revealed an absent right kidney and an ipsilateral seminal cyst. MRI revealed an absent right kidney, seminal vesicle cyst, and right ejaculatory duct obstruction, confirming the Zinner syndrome diagnosis.

Almofareh *et al.*,[6] in their study, reported Zinner syndrome in a 35-year-old male patient who presented with primary infertility for 10 years, in whom ultrasound revealed an absent right kidney, CT confirmed right renal agenesis and right seminal vesicle cyst. Ibrahimi *et al.*[7] reported the case of a 33-year-old patient who presented with complaints of recurrent dysuria and ejaculatory disorders for the past 5 years in whom imaging studies revealed an empty left renal fossa and a left seminal vesicle which was compatible with the diagnosis of Zinner syndrome. Similarly, in the studies by Militaru *et al.*,[8] Demaeyer *et al.*,[9] Almuhanna *et al.*,[10] Gurung *et al.*,[11] Gorantla *et al.*,[12] Jaganathan *et al.*,[13] and Djidda *et al.*,[5] MRI confirmed Zinner syndrome.

CONCLUSION

With advances in radiology, the diagnosis of Zinner syndrome can be made before the 2nd and 3rd decades. Although uncommon, any doubt should be thoroughly investigated with initial screening by ultrasonography and

confirmation with MRI. The early detection would help the patient relieve the symptoms and prevent complications that may arise due to the same. Disease progression may be arrested and may also improve quality of life. Male patients presenting with uncommon symptoms such as dysuria and infertility without any identifiable cause may be a ground to investigate Zinner syndrome.

REFERENCES

1. Florim S, Oliveira V, Rocha D. Zinner syndrome presenting with intermittent scrotal pain in a young man. Radiol Case Rep 2018;13:1224-7.
2. AlArifi M, Al-Gahwary M, Gomha M. The association of renal agenesis and ipsilateral seminal vesicle cyst: Zinner syndrome case report. Case Rep Urol 2019;2019:1242149.
3. Pereira BJ, Sousa L, Azinhais P, Conceição P, Borges R, Leão R, *et al.* Zinner's syndrome: An up-to-date review of the literature based on a clinical case. Andrologia 2009;41:322-30.
4. Zinner A. A Confusing Diagnosis in the Face of Chronic Disabling Perineoscrotal Pain in the Young Subject, about a Case. Wien Med Wochenschr 1914;64:605-9.
5. Abakar D, Badi FE, Sabiri M, El Manjra S, Lezar S, Essodegui F. Zinner syndrome. Eur J Case Rep Intern Med 2021;8:002628.
6. Almofareh AM, Aljuaid KM, Almirabi SA, Hakami YM, Alwan AM. Zinner syndrome: A case report of a rare etiology of infertility. Cureus 2021;13:e17386.
7. Ibrahimi A, Hosni A, Ziani I, Laamrani FZ, Sayegh HE, Jroundi L, *et al.* Zinner's Syndrome: A rare diagnosis of dysuria based on imaging. Case Rep Urol 2020;2020:8826664.
8. Militaru V, Mihaly ZA, Ilea C, Coman M, Stanciu M, Crisan, N, *et al.* Zinner syndrome-A case report. Med Pharm Rep 2021;94 Suppl No 1:S47-50.
9. Demaeyer L, Holz S, Pamart D, Taylor S, Naudin M. Robotic management of painful Zinner syndrome, case report and review of the literature. Int J Surg Case Rep 2020;73:61-4.
10. Almuhanna AM, Alsuhaibani S, Almesned R, Almatar A, Alali H. Zinner's syndrome: A case report of a rare maldevelopment in the male genitourinary tract. Urol Case Rep 2021;39:101839.
11. Gurung B, Panta OB, Dhakal V, Ghimire RK. Zinner syndrome: A case report of rare urogenital anomaly. J Med Ultrasound 2021;30:59-61.
12. Gorantla R, Allu S, Rao A. Triad of unilateral renal dysgenesis with ipsilateral seminal vesical and ejaculatory duct obstruction: An uncommon urogenital congenital anomaly, Zinner syndrome-a case report. Indian J Radiol Imaging 2021;31:707-9.
13. Jaganathan D, Kathirvelu G, Nagarajan S, Ganesan UN. Rare cause of painful ejaculation-Zinner's syndrome: A case report. Am J Sonogr 2020;3:1.

How to cite this article: Kada NC, Mariappan K. Zinner Syndrome – A Case Report. Int J Sci Stud 2023;10(11):8-10.

Source of Support: Nil, **Conflicts of Interest:** None declared.