Case Report

Zinner syndrome mimicking bladder outlet obstruction managed with aspiration

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Abstract

Zinner syndrome is a rare cystic malformation of seminal vesicle which consists a triad of unilateral renal agenesis, ipsilateral seminal vesicle cyst, and ejaculatory duct obstruction. The usual presentation is between the third and fourth decades of life, with infertility being the most common complaint. Ultrasound, cystoscopy, and magnetic resonance tomography (MRI) can easily detect this condition. Treatment option varies according to the presenting symptoms of the patient. We present a case of a 19-year-old male with recurrent episodes of urinary tract infection (UTI) and poor urinary stream. On ultrasound examination, the patient was found to have absent right kidney with a cystic swelling noted to be arising from prostate or seminal vesicle region which was further confirmed on MRI examination. Cystoscopy reveals a bulge on the right side of the verumontanum abutting the neck of the bladder. An ultrasound-guided aspiration of the cyst was performed which relieved the symptoms of the patient. Cystic abnormalities of the seminal vesicle are very uncommon. Symptomatic cases may present as recurrent UTI, infertility, bladder outlet obstruction, and painful ejaculation. Surveillance may be the option in the absence of clinical manifestations. Interventions such as image-guided aspiration or surgical procedures are appropriate when conservative measures prove ineffective.

Keywords: Cyst, cystoscopy, magnetic resonance tomography, seminal vesicle, ultrasound, Zinner syndrome

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INTRODUCTION

There are various congenital abnormalities noted in the urogenital system ranging from innocuous condition such as duplication of the ureter to life-threatening condition such as bilateral renal agenesis.[1] The most common organ affected with malformations is the ureter in the urogenital tract. Congenital malformations of the seminal vesicle are very uncommon. Malformations of seminal vesicle range from complete absence of the seminal vesicle to solid and cystic enlargement.[2]

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Most of the malformations are cystic in nature due to collection of inspissated material in the seminal vesicle following obstruction of the ejaculatory duct. Malformation occurs due to insults between 4 and 13 weeks of gestation. Zinner syndrome is a cystic malformation of the Wolffian duct, a male counterpart of Mayer–Rokitansky–Küster–Hauser syndrome seen in women, seen in 0.005% of all the cases.[3,4] It consists a triad of ipsilateral seminal vesicle cyst, ejaculatory duct obstruction, and most commonly unilateral renal agenesis. The usual age of presentation is

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the third or fourth decade of life.[5] Most of the patients are asymptomatic which are detected incidentally. Other patients present with chief complaints of infertility, dysuria, urgency, prostatism, and painful ejaculation due to obstruction of the ejaculatory ducts and distension of the seminal vesicle.

CASE REPORT

A 19-year-old unmarried male patient presented to the outpatient department with complaints of recurrent episode of urinary tract infections (UTIs). The patient also complained of poor urinary stream which slightly improved on straining from the past 1 year. The patient was average built, and clinical parameters were normal. Abdominal examination and the external genitalia were normal, and Vas deferens was palpable bilaterally. On digital rectal examination, there was a soft, smooth, nontender bulge just above the prostrate. Laboratory investigations were in the normal range. Urine culture was sterile. Uroflometry done initially showed a maximum flow rate of 7 ml/min.

Computed tomography (CT) scan already done from outside revealed absent right kidney and well-defined peripherally enhancing cystic lesion with hyperdense contents arising likely from the seminal vesicle. Ultrasound examination of the abdomen showed absent right-sided kidney with mild degree of cystitis. Transrectal ultrasonography showed a cystic lesion of size 5 cm × 4 cm, with internal echoes arising from the right seminal vesicle abutting the prostrate and urinary bladder posteriorly. Both the testes were of normal size. MRI scan showed a well-defined paramidline cystic lesion of size 5.67 cm × 4.22 cm [Figure 1], with hyperintensity on T1- and T2-weighted images. The swelling was indenting the base of bladder superiorly [Figure 2] and the right side of the prostate and seminal vesicle inferiorly with inspissated secretions within [Figure 3].

Cystoscopy was performed which shows a smooth bulge present at the bladder neck from the right side, crossing the midline [Figure 4]. Serum prostate-specific antigen was normal. Semen analysis showed oligospermia with reduced motile spermatozoa.

After discussing the condition and future outcomes, the patient did not agree for surgical removal, henceforth, transrectal ultrasound-guided aspiration was done yielding 8 ml thick viscid content with no sperms on microscopic examination. The swelling decreased in size and maximum flow rate improved to 23 ml/min. The patient followed up for 4 months and is healthy with adequate urinary stream and flow rate maintained at 19 ml/min.

DISCUSSION

Congenital malformations of the urinary tract have been seen to be known to affect multiple organs. Congenital anomalies of the kidney and urinary tract describe the various conditions in the pediatric age group.[1] This includes ureteral duplications (1% of population),[6] horseshoe kidney (1 in 500 people),[7] obstructive renal dysplasia (3.7% of population),[8] and ADPK (1 in 1000 individuals).[9]

The mesonephric duct plays an important role in the development of the urogenital system. It is a paired organ which, in males, gives rise to hemitrigone, bladder neck, urethra, seminal vesicle, vas deferens, efferent ducts, epididymis, paradidymis, and appendix epididymis under the influence of testosterone and anti-Mullerian hormone.This development takes place in the first 3 months of pregnancy, and any insult seriously hampers the

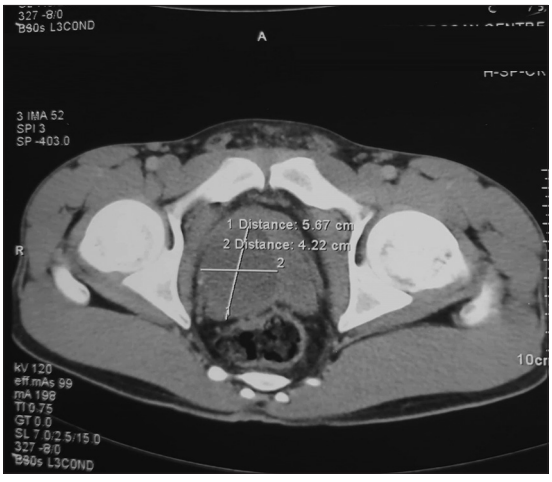

Figure 1: MRI scan (transverse section) showing seminal vesicle cyst compressing the urinary bladder

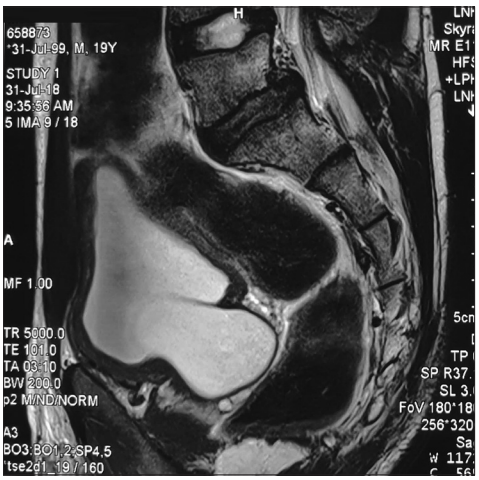

Figure 2: MRI scan (sagittal section) showing seminal vesicle cyst

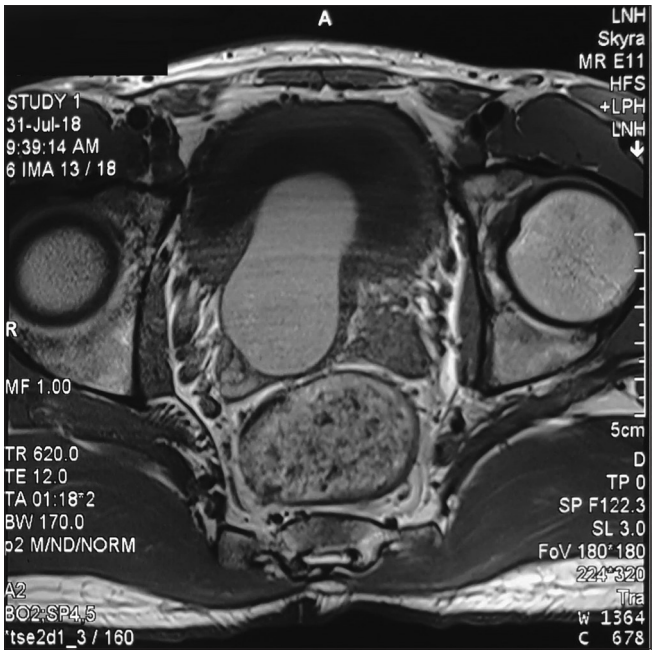

Figure 3: MRI scan (transverse section) showing seminal vesicle cyst

development of the kidney, ureter, seminal vesicle, and vas deferens. Cystic malformation of the seminal vesicle is very uncommon. It occurs due to impaired development of the distal part of mesonephric ducts causing obstruction of the ejaculatory ducts and dilatation of seminal vesicle. It is also associated with renal agenesis due to involvement of the ureteric bud.[1]

Zinner first described his findings in 1914 as a triad of unilateral renal agenesis, ipsilateral seminal vesicle cyst, and ejaculatory duct obstruction.[2] Zinner syndrome is a cystic malformation of the Wolffian duct, a male counterpart of Mayer–Rokitansky–Küster-Hauser syndrome seen in women. Casey *et al*. also reported a pentad including cystic dysplasia of rete testis, seminal vesicle cyst, ipsilateral renal agenesis, partial hemitrigonal development, and epididymal dilatation in a radiological study.[10] About 100 cases of Zinner syndrome have been reported so far.[2]

Most of the patients with cyst size <5 cm are asymptomatic.[11] Other patients manifest at the reproductive age group, with infertility as the most common presentation.[12] The various other manifestations reported till date include infertility, UTIs, painful ejaculations, epididymitis, prostatitis, and bladder outlet obstruction.[2,13,14]

Most of the cysts are discovered by ultrasonography and digital rectal examination. Clinical presentation depends on the size and location of the cyst; however, cyst reaching up to 12 cm has been reported.[15]

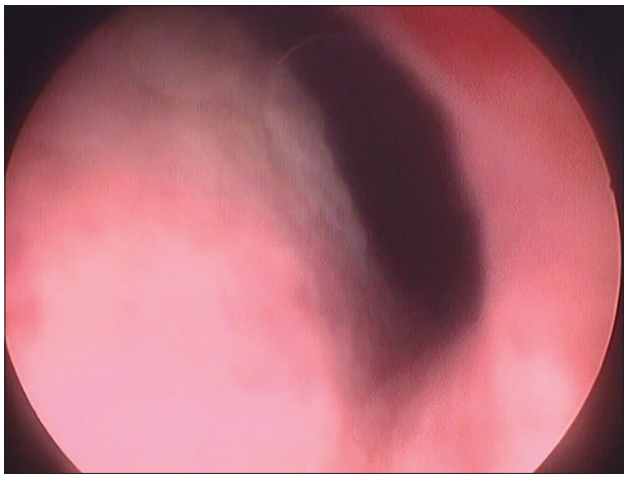

Figure 4: Cystoscopy view of seminal vesicle cyst bulging on the right side right side

The diagnostic workup of these patients includes transrectal ultrasonography, CT scanning, cystoscopy, and MR imaging. Intravenous urography can be used to see the cystic mass impinging on the bladder. MR scanning can best identify the renal agenesis and associated cystic mass.[16,17] Transrectal ultrasound can also determine the size and location of the cyst.[5] A seminal vesicle cyst may show anechoic contents or contain debris due to previous hemorrhage or infection but is limited by its small field of view. MRI is the best investigation due to greater tissue contrast resolution. It gives detailed pelvic anatomy and differentiation of cystic masses, therefore providing a definitive diagnosis. Seminal vesicle cysts appear low attenuation on T1-weighted images and high attenuation on T2-weighted images.[5]

All the patients undergoing treatment should undergo infertility workup before any treatment is offered. Surveillance may be the option in the absence of clinical manifestations. Intervention or surgical procedures are appropriate when conservative measures prove ineffective. There are various treatment options that include antibiotic therapy for infected inspissated collection and UTIs, percutaneous image-guided aspiration, sclerosant injection, transurethral resection of the ejaculatory ducts or surgery.[5,18-20] Recently, robot-assisted laparoscopic surgery is also reported for Zinner syndrome. There is no current preferred method of treatment due to paucity of the literature and limited number of cases reported. Minimally invasive options should be preferred to open surgery; however, size of the cyst and complaints may decide the course of the treatment.[19] The method of treatment depends on the symptoms of the patient. Only the symptomatic cases are treated. The percutaneous method of aspiration of cyst was reported by Kajita *et al*. which was

found to be effective in Zinner syndrome patient.[20] The same method was adopted in our case. Open surgery was found to effective by Van den Ouden *et al*. with complete resolution of symptoms. The cure rate of 80% was found in transurethral unroofing of the cyst.[21] Laparoscopic surgery with transperitoneal approaches was also found to be having a successful outcome.[22] Major consideration of surgery is preserving the fertility of the patient. Due to proximity of the vital pelvic structure, more minimally invasive procedures are devised.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Rodriguez MM. Congenital anomalies of the kidney and the urinary tract (CAKUT). Fetal Pediatr Pathol 2014;33:293-320.
2. Kenney PJ, Leeson MD. Congenital anomalies of the seminal vesicles: Spectrum of computed tomographic findings. Radiology 1983;149:247-51.
3. McCallum T, Milunsky J, Munarriz R, Carson R, Sadeghi-Nejad H, Oates R, *et al*. Unilateral renal agenesis associated with congenital bilateral absence of the vas deferens: Phenotypic findings and genetic considerations. Hum Reprod 2001;16:282-8.
4. Sheih CP, Hung CS, Wei CF, Lin CY. Cystic dilatations within the pelvis in patients with ipsilateral renal agenesis or dysplasia. J Urol 1990;144:324-7.
5. Kardoust Parizi M, Shakhssalim N. Management of Zinner's syndrome associated with contralateral seminal vesicle hypoplasia: A case report. Case Rep Urol 2013;2013:494215.
6. Siomou E, Papadopoulou F, Kollios KD, Photopoulos A, Evagelidou E, Androulakakis P, *et al*. Duplex collecting system diagnosed during the first 6 years of life after a first urinary tract infection: A study of 63 children. J Urol 2006;175:678-81.
7. Kirkpatrick JJ, Leslie SW. Horseshoe kidney. In: StatPearls. Treasure Island (FL): StatPearls Publishing; 2018. Available from: https://www.ncbi.nlm.nih.gov/books/NBK431105/. [Last updated on 2018 Mar 15].
8. Chen RY, Chang H. Renal dysplasia. Arch Pathol Lab Med 2015;139:547-51.
9. Willey CJ, Blais JD, Hall AK, Krasa HB, Makin AJ, Czerwiec FS. Prevalence of autosomal dominant polycystic kidney disease in the European Union. Nephrol Dial Transplant 2017;32:1356-63.
10. Casey RG, Stunell H, Buckley O, Flynn R, Torreggiani WC. A unique radiological pentad of mesonephric duct abnormalities in a young man presenting with testicular swelling. Br J Radiol 2008;81:e93-6.
11. Shah S, Patel R, Sinha R, Harris M. Zinner syndrome: An unusual cause of bladder outflow obstruction. BJR Case Rep 2017;3:20160094.
12. Aghaways I, Ahmed SM. Endourologic intervention for management of infertility in a man with Zinner syndrome resulting in a natural pregnancy. J Endourol Case Rep 2016;2:71-3.
13. Beeby DI. Seminal vesicle cyst associated with ipsilateral renal agenesis: Case report and review of literature. J Urol 1974;112:120-2.
14. Levisay GL, Holder J, Weigel JW. Ureteral ectopia associated with seminal vesicle cyst and ipsilateral renal agenesis. Radiology 1975;114:575-6.
15. Logigan H, Manea C, Crişan N, Pop C, Bungărdean C, Hiriscau I, *et al*. Zinner syndrome: A case report and literature review. Rev Românæ Urol 2014;13:44-7.
16. 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;11:313-7.
17. 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.
18. Yurdakul T, Gokce G, Kilic O, Piskin MM. Transurethral resection of ejaculatory ducts in the treatment of complete ejaculatory duct obstruction. Int Urol Nephrol 2008;40:369-72.
19. Kord E, Zisman A, Darawsha AE, Dally N, Noh PH, Neheman A, *et al*. Minimally invasive approach for treatment of seminal vesicle cyst associated with ipsilateral renal agenesis. Urol Int 2017;99:338-42.
20. Kajita Y, Shimizu Y, Takao N, Iwaki H, Yamauchi T. Seminal vesicle cyst with ipsilateral renal agenesis: A case report. Hinyokika Kiyo 2001;47:203-6.
21. van den Ouden D, Blom JH, Bangma C, de Spiegeleer AH. Diagnosis and management of seminal vesicle cysts associated with ipsilateral renal agenesis: A pooled analysis of 52 cases. Eur Urol 1998;33:433-40.
22. Seo IY, Kim HS, Rim JS. Congenital seminal vesicle cyst associated with ipsilateral renal agenesis. Yonsei Med J 2009;50:560-3.