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Case Report

Zinner syndrome—a rare developmental anomaly of the mesonephric duct diagnosed on magnetic resonance imaging

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ABSTRACT

Developmental anomalies of the urogenital tract are rare but often encountered. Zinner's syndrome is a rare congenital abnormality of mesonephric (Wolffian) duct consisting of unilateral renal agenesis, ipsilateral seminal vesicle cyst, and ipsilateral ejaculatory duct obstruction due to developmental arrest in early embryogenesis affecting the caudal end of Mullerian duct and only approximately a 100 cases have been reported so far. Radiologic modalities such as intravenous pyelography, ultrasonography, vasovesiculography, contrast enhanced computed tomography, and magnetic resonance imaging are all helpful in diagnosis of this unusual entity. We present here an extremely rare developmental anomaly involving the Mullerian ducts, which would remain undiagnosed but for radiologic imaging. The patient presented with symptoms of lower urinary tract irritation.

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Case report

We report an unusual case of a 25-year-old adult male who presented with history of dysuria, passage of fiber-like structures in urine, and frequency of micturition since 4 years. There was history of infertility as he had been married for 3 years, but his spouse had not conceived in spite of multiple attempts to conceive. The patient had no endocrine or systemic disease. The physical examination revealed normally developed external genitalia. Vas deferens was palpable bilaterally. The blood work and biochemistry was normal. A routine urine examination showed 40% sperms in

the urine sample. Subsequent to this, semen examination was done, and the sperm count was reported to be 11 million with a semen volume of 1.9 mL (normal reference range of sperm count is >20 million sperms and 2–6 mL ejaculate volume).

A previous ultrasound abdomen reported grossly hydronephrotic pelvic kidney with dilated tortuous right ureter. The patient was referred to the radiology department for further evaluation. A sonogram of abdomen showed the absence of kidney in the right renal fossa with compensatory hypertrophy of the normally situated left kidney and a cystic mass in the right side of pelvis in the periprostatic region (Figs. 1A and B). A provisional diagnosis of hydronephrotic ectopic right kidney

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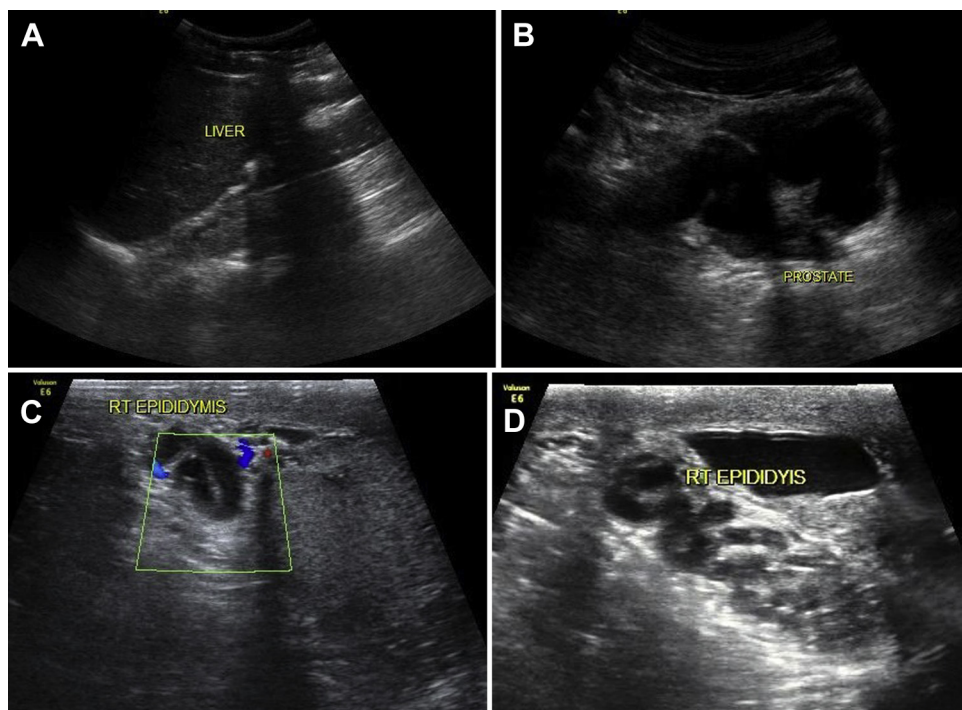


Fig. 1 – (A) Absent kidney in the right renal fossa. (B) Pelvic ultrasound shows cystic tubular structure posterior and to the right of the urinary bladder extending up to the midline. (C and D) Scrotal sonography image showing tubular dilatation in the head of epididymis with tortuous tubular nonvascular structures seen in the right spermatic cord.

was made based on these sonography findings. On scrotal sonography, dilated tubular structures were demonstrated in the tail of right epididymis and in the right spermatic cord (Figs. 1C and D). A contrast-enhanced abdominopelvic computed tomography (CT) was performed. A fluid attenuation nonenhancing mass was seen in the right side of pelvis inferolateral to the urinary bladder in the periprostatic region (Fig. 2A). On delayed scans, there was no excretion of contrast by the cystic mass, and the possibility of an ectopic kidney was ruled out (Fig. 2B). A dilated tubular structure was seen extending up from the mass into the right lower abdomen, ending abruptly between the common iliac vessels in the midline (Figs. 2C and D). The possibility of an atretic ureter was considered and a magnetic resonance imaging (MRI) pelvis was subsequently performed to ascertain whether the pelvic mass was arising from the seminal vesicles and to confirm whether the abruptly ending tubular structure was an atretic ureter. MR demonstrated multiple cystic-to-tubular structures in the right seminal vesicle appearing hyperintense on T2-weighted images and hypointense on T1W image, with similar tubular dilatation in the left seminal vesicle also (Figs. 3A and B). A dilated tortuous tubular structure was seen coursing along the right iliac vessels toward the midline terminating abruptly at the level of aortic bifurcation (Figs. 4A and B). It had no connection with the bladder and because of its course upward into the abdomen from the pelvis along the iliac vessels, it was diagnosed to be an ectopic atretic ureter (Fig. 4C). The absent right kidney was confirmed on CT images (Fig. 5).

The sonography and CT findings were reviewed, and the absence of right kidney with compensatory hypertrophy of the normally situated left kidney, dilatation within the right

epididymis and vas deferens seen on sonography, and MR findings of grossly dilated bilateral ejaculatory ducts with seminal vesicle cysts, along with the visualization of atretic right ureter on CT and MRI lead to the diagnosis of Zinner's syndrome, which is a developmental anomaly of the mesonephric duct. Intraoperative laparoscopic findings confirmed our diagnosis, and transurethral derroofing of the seminal vesicle cysts was performed.

Discussion

Zinner's syndrome, first described in 1914 by Zinner, is a rare congenital malformation of the seminal vesicles and ipsilateral upper urinary tract [1,2]. The patients are usually diagnosed in the 2nd-3rd decade of life and present with symptoms of dysuria (37%), frequency (33%), perineal pain (29%), and epididymitis (27%) [3]. It is considered to be the male counterpart of Mayer-Rokitansky-Kuster-Hausers syndrome seen in females [4]. An insult occurring before the 7th gestation week causes maldevelopment of the distal part of the mesonephric duct producing atresia of both ejaculatory duct and the ureteric bud [5].

The close embryologic relationship between the genital and urinary tracts explains the developmental aberrations leading to this anomaly. The Mesonephric or Wolffian duct, which forms the male reproductive system as well as the ureteric bud, is a paired structure. The orifice of the distal mesonephric duct and the ureteric bud separates between 6th-8th gestational week, and the ureteric orifice migrates toward the metanephric blastema, whereas the distal part of

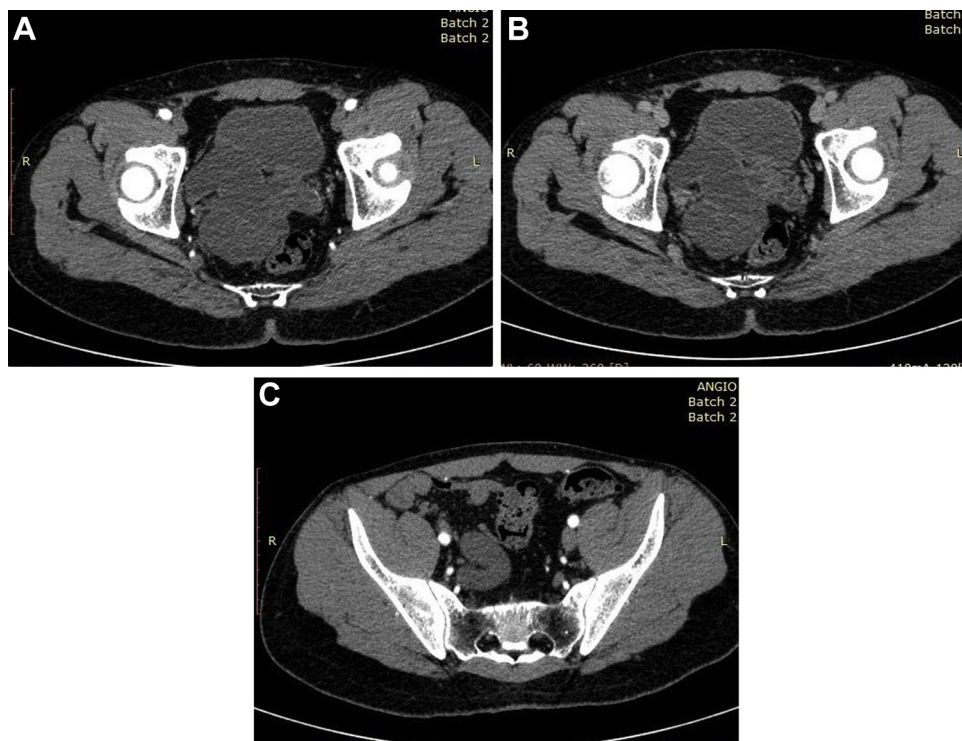


Fig. 2 – (A) Contrast enhanced computed tomography (CECT) pelvis: axial image showing lobulated large well-margined nonenhancing mass in the periprostatic region between the bladder and rectum. The mass is indenting the posterolateral bladder wall and extends across the midline to the left. **(B)** CECT pelvis-delayed images: the pelvic mass shows no contrast enhancement or excretion. **(C, D)** Axial CECT images: a tubular structure seen slightly to the right of the false pelvis, adjacent to the right external iliac and internal iliac arteries, ending abruptly at the level of the common iliac arteries where it appears round and cystic.

mesonephric duct under the influence of testosterone and anti-Müllerian hormone, forms the hemitrigone, the bladder neck, the urethra up to the external sphincter, the seminal vesicle, vas deferens, ejaculatory ducts, epididymis, paradidymis, and appendix of the epididymis [6]. During the 4th to 6th week of gestation, the metanephric blastema secretes growth factors, which induce the growth of the ureteric bud toward it. The ureteric bud too secretes growth factors and

proliferates, fusing with the metanephric blastema, and in turn inducing the blastema to become the primitive kidney on undergoing mesenchymal to epithelial transition [7].

Disturbance in any of these inductive events during this period of embryogenesis such as mutation of metanephric blastema, or disruption of retinoic acid signaling, causes inhibition of ureteric bud growth with failure of fusion of the ureteric bud with the metanephric blastema and renal agenesis or renal

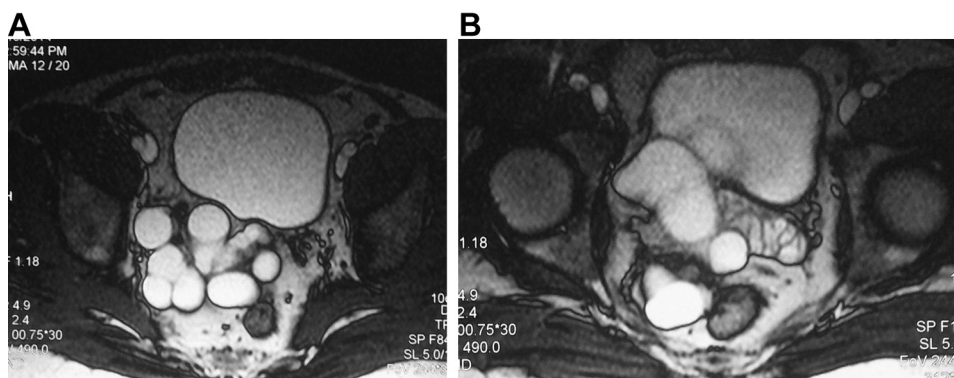


Fig. 3 – (A) Axial T2W MR image. Multiple cystic hyperintense structures lying close together, in the region of the right seminal vesicle, indenting the right lateral bladder wall and the bladder base, extending inferiorly into the spermatic cord. **(B)** Pelvic MR T2 axial image: similar cystic tubular dilated structures visible in the left seminal vesicle. MR, magnetic resonance.

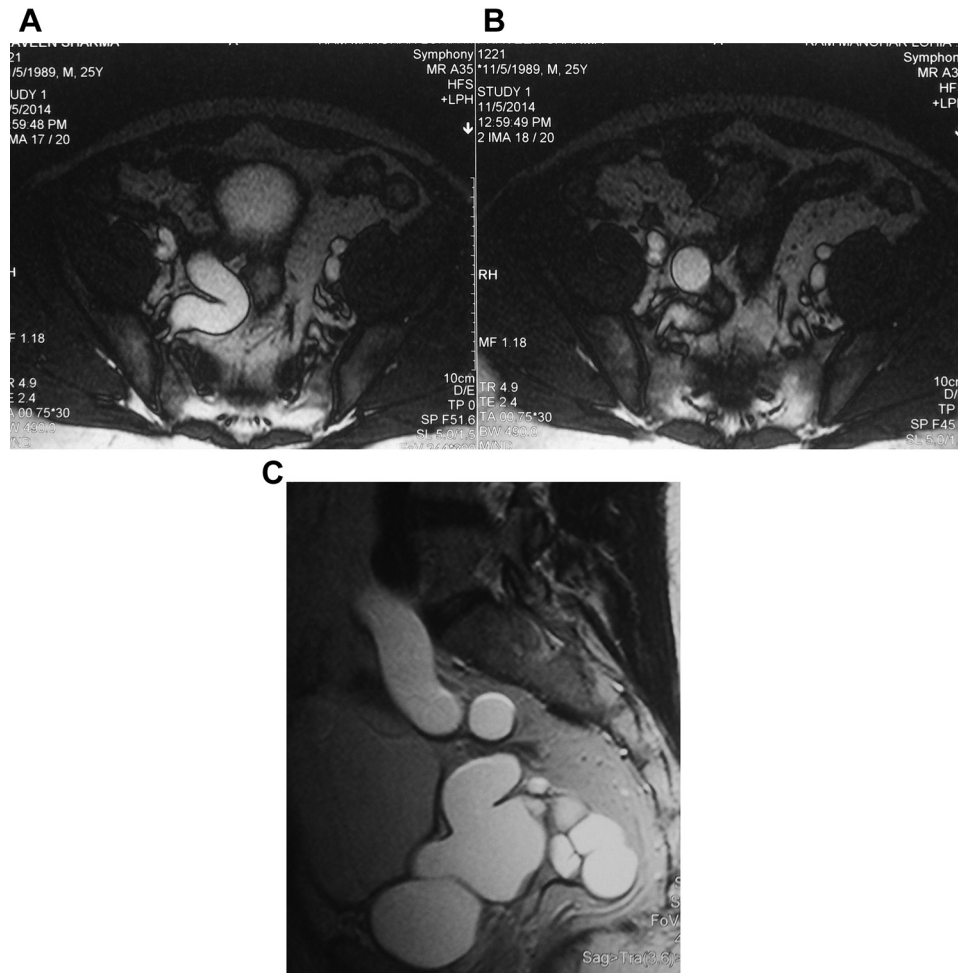


Fig. 4 – (A, B) T2W MR: a tortuous tubular structure seen posterolaterally in the pelvis adjacent to the internal iliac vessels, on the right side in this axial T2 MR image. The structure assumes a round shape at a slightly superior level. (C) Sagittal T2W MR: image shows the tubular cystic structure extending upward from the false pelvis to the level of aortic bifurcation where it is seen to end abruptly anterior to the vessels. MR, magnetic resonance.

hypoplasia. Simultaneous failure of the ureteric bud to separate from the lower part of mesonephric duct leads to atresia of ejaculatory ducts and obstruction of the seminal vesicles with accumulation of secretions resulting in cystic dilatation [8].



Fig. 5 – Axial computed tomography: image shows single left kidney with absent right kidney.

Zinner's syndrome is thus characterized by the triad of ipsilateral ejaculatory duct obstruction, seminal vesicle cysts, and renal agenesis.

Dysuria, frequency of micturition, perineal pain, and painful ejaculation due to pressure of the seminal vesicle cysts are the presenting complaints [9]. Later oligozoospermia and even azoospermia develops, leading to infertility. The condition comes to the notice of the clinician either in young adults in the second to fourth decade when patients present with infertility.

Imaging enables accurate diagnosis of this rare anomaly of the genitourinary tract. On sonography, ipsilateral kidney is absent, and the obstructed ejaculatory ducts are seen as anechoic structures in the pelvis, though hemorrhage or infection can produce internal low level echoes [10]. CT is better than sonography, and CT findings are of a retro-vesicular periprostatic cystic mass along with ipsilateral renal agenesis [11].

MRI is the modality of choice for precise delineation of the anatomy of the male genital tract, for demonstrating the

seminal vesicles and evaluating anomalies of the mesonephric duct and to differentiate the seminal vesicle cysts from other cystic pelvic masses [12]. MR is excellent for delineation of prostate and seminal vesicles and confirming that the cystic periprostatic structures are indeed within the seminal vesicles [13]. The characteristic periprostatic and paramedian location enables the accurate identification of the seminal vesicle cysts on MR, which appear hypointense or hyperintense on T1W depending on the presence of amount of protein content and hyperintense on T2W images [14]. A convoluted tail-like connection of the cystic tubules to the seminal vesicle also pins the seminal vesicles as the site of origin. Cysts are rarely more than 5 cm in size, although giant cysts larger than 12 cm have been described and these can cause bowel and bladder obstruction [9]. MR is superior to CT as it demonstrates the anatomic relations in the pelvis better and can identify the ectopic ureteric orifices, which are missed on CT. Vasovesiculography is another confirmatory imaging technique where an iodinated contrast material is injected into the vas deferens ducts and serves to demonstrate seminal vesicle patency. Reflux of contrast from the tortuous dilated seminal vesicles into the ipsilateral atretic ureter is seen in Zinner's syndrome because of ejaculatory duct obstruction.

Dilated seminiferous tubules need to be differentiated from cysts of prostatic duct and prostatic utricle. The latter have a median location. Vesical diverticula are cystic and paramedian, but accompanied with normal seminal vesicles. Ectopic ureterocele are laterally located tubular structures in the pelvis, but their course identifies them at the lower end of a ureter [14]. Ectopic hydronephrotic pelvic kidney appears as cystic pelvic mass, which demonstrates some contrast excretion and an aberrant artery entering it.

Aspiration of cyst fluid transperineally and identification of spermatozoa in it is also confirmatory for Zinner's syndrome.

Surgical aspiration of the seminal vesicle cysts through perineal or laparoscopic approach or percutaneous cyst drainage or a transurethral cyst deroofing is considered therapeutic and by relieving the ejaculatory duct obstruction provides excellent cure and symptomatic relief (100% and 75%, respectively) [12]. Infertility results from ejaculatory duct obstruction and when obstruction is released, fertility is restored.

Conclusion

Developmental anomalies of the urogenital system are often not considered by clinicians when patients present with vague symptoms pertaining to the urinary tract. Imaging alone has the ability to provide an accurate diagnosis and is quintessential for detecting Mullerian duct anomalies, with MRI proved best for delineation of anatomy of male genital tract.

Learning points

1. MRI is the best imaging modality for diagnosing genital tract developmental anomalies in males and females as the anatomic delineation is precise. Imaging with MR

enables confirmation of diagnosis by identifying cystic dilatation of seminiferous tubules in the characteristic periprostatic location. MRI is better for delineating the anatomic relations in the pelvis and has a superior soft tissue delineation and multiplanar capability.

2. Imaging is essential for confirming the triad of findings associated in Zinner's syndrome. Ejaculatory duct obstruction leads to the characteristic symptoms of dysuria and causes infertility in this rare developmental anomaly of the male genitor-urinary system.
3. An understanding of the anatomy and embryology of genitourinary tract is essential to make an accurate diagnosis of developmental anomalies of the genitourinary tract.

REFERENCES

- [1] Pereira BJ, Sousa L, Azhinais P, Conceicao P, Borges R, Leao R, et al. Zinner's syndrome: an up-to-date review of literature based on clinical case. *Andrologia* 2009;41(5):322–30.
- [2] Zinner A. Ein fall von intravesikaler Samenblasenzyste. *Wien Med Wochenschr* 1914;64:605.
- [3] King BF, Hattery RR, Lieber MM, Berquist TH, Williamson Jr B, Hartman GW. Congenital cystic disease of the seminal vesicle. *Radiology* 1991;178:207–11.
- [4] Gianna P, Giuseppe PG. Mayer-Rokitansky-Küster-Hauser syndrome and the Zinner syndrome, female and male malformation of reproductive system: are two separate entities? *J Chinese Clin Med* 2007;2:11.
- [5] Livingston L, Larson CR. Seminal vesicle cyst with ipsilateral renal agenesis. *AJR Am J Roentgenol* 2000;175:177–80.
- [6] Denes FT, Montanellato NI, Lopes RN, Barbosa Filho CM, Cabral AD. Seminal vesicle cyst and ipsilateral renal agenesis. *Urology* 1986;28:313–5.
- [7] Hannema SE, Hughes IA. Regulation of Wolffian duct development. *Horm Res* 2007;67:142–51.
- [8] McCallum TJ, Milunsky JM, Munarriz R, Carson R, Sadeghi-Nejad H, Oates RD. Unilateral renal agenesis associated with congenital bilateral absence of the vas deferens: phenotypic findings and genetic considerations. *Hum Reprod* 2001;16:282–8.
- [9] Ghonge NP, Aggarwal B, Sahu AK. Zinner syndrome: a unique triad of mesonephric duct abnormalities as an unusual cause of urinary symptoms in late adolescence. *Indian J Urol* 2010;26(3):444–7.
- [10] Trigaux JP, Van Beers B, Delchambre F. Male genital tract malformations associated with ipsilateral renal agenesis: sonographic findings. *J Clin Ultrasound* 1991;19:3–10.
- [11] Kenney PJ, Leeson MD. Congenital anomalies of the seminal vesicles: spectrum of computed tomographic findings. *Radiology* 1983;149:247–51.
- [12] Arora SS, Breiman RS, Webb EM, Westphalen AC, Yeh BM, Coakley FV. CT and MRI of congenital anomalies of seminal vesicles. *AJR Am J Roentgenol* 2007;189:130–5.
- [13] Murphy JO, Power RE, Akhtar M, Torreggiani WC, McDermott TE, Thornhill JA. Magnetic resonance imaging in the diagnosis of seminal vesicle cysts and associated anomalies. *J Urol* 2003;170:2386.
- [14] Van den Ouden D, Blom JHM, Bangma C, de Spiegeleer AHV. Diagnosis and management of seminal vesicle cysts associated with renal agenesis: a pooled analysis of 53 cases. *European Urology* 1998;33(5):433–40.