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Case Report

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

Shibani Mehra MD[*], *Rajeev Ranjan MD*, *Umesh Chandra Garga MD*

Department of Radiology, Dr RML Hospital, Postgraduate Institute for Medical Education and Research, Baba Kharak Singh Marg, New Delhi 110001, India

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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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* *Corresponding author.*
E-mail address: shibani.mehra@yahoo.in (S. Mehra).

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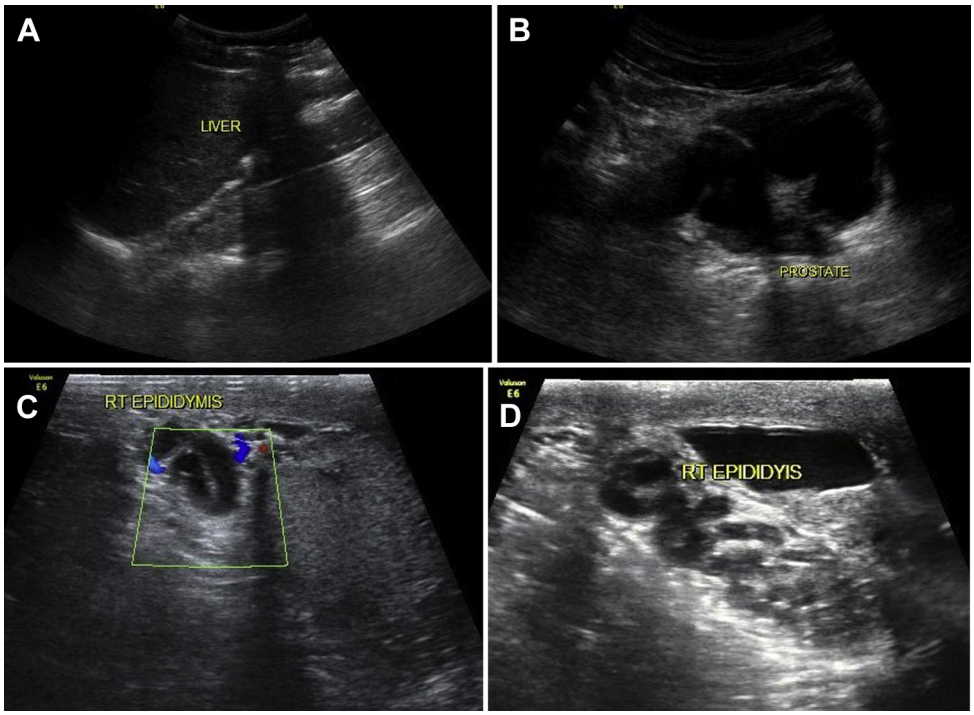

Fig. 1 – (A) Absent kidney in the right renal fossa. (B) Pelvic ultrasound shows cystic tubular structure posterior and to the right of the urinary bladder extending up to the midline. (C and D) Scrotal sonography image showing tubular dilatation in the head of epididymis with tortuous tubular nonvascular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

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