Case Report

Zinner syndrome – a rare radiological diagnosis in a young male presenting with recurrent dysuria

Dinesh Chataut, Surendra Rayamajhi, Sundar Suwal, Ajit Thapa

Abstract

Zinner syndrome is a rare congenital abnormality of the mesonephric (Wolffian) duct consisting of unilateral renal agenesis, ipsilateral seminal vesicle cyst, and ipsilateral ejaculatory duct obstruction. The syndrome usually presents in the second or third decade of life mainly after the beginning of sexual activity and usually in the form of voiding manifestations. The main imaging modality is magnetic resonance imaging (MRI); however, usually suspected in transabdominal sonography, which may give initial clue to the diagnosis. Herein, we report a rare case of a 24 - year male with right renal agenesis with cystic pelvic mass diagnosed as Zinner syndrome via multimodality imaging.

Keywords: MRI, Seminal vesicle cyst, Young male, Zinner syndrome

Author affiliations:

Department of Radiology and Imaging, Tribhuvan University Teaching Hospital, Maharajgunj, Kathmandu, Nepal

Correspondence:

Dr. Sundar Suwal,
Department of Radiology and Imaging, Tribhuvan University Teaching Hospital, Maharajgunj, Kathmandu, Nepal.

Email: sundarsuwal@iom.edu.np

ORCID: https://orcid.org/0000-0001-8534-3413

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Introduction

The congenital urinary tract anomalies are caused by defect in the morphogenesis of the urinary system and may include obstructive as well as nonobstructive urinary tract dilatation. These anomalies may coexist with alterations in number, size or position of the kidneys and are more common in males than in the females (M:F = 2.5:1).[1,2]

We report a rare congenital urinary tract anomaly in young male involving right urinary tract with associated right renal agenesis.

Case Report

A 24-year young male patient, who is the father of two children, presented with vague complaints of dysuria and pain during ejaculation for the last two years. Urine analysis and CBC was normal and there was no history of fever.

Transabdominal sonography showed a cystic lesion in right side of pelvis protruding into the urinary bladder at the region of the right vesicoureteric junction (VUJ). It measured about 7.2x5.3 cm in size. The right kidney was not visualized in the right renal fossa and pelvis with mild compensatory hypertrophy of the left kidney (**Figure 1**). With no history of prior surgery, sonographic diagnosis of suspected ectopic right kidney with right ureterocele was made.

Further evaluation of the patient was done with the CT urography after the renal function test came normal. On CT urography, the absence of the right kidney with absent renal artery and vein was confirmed. Right retrovesical cystic lesion was seen protruding into the bladder at the region of the right VUJ. Right seminal vesicle was not separately visualized. Dilated & blind-ended right distal ureter was

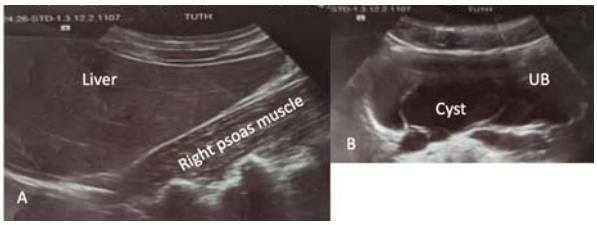

Figure 1. Transabdominal sonography images.
Empty right renal fossa (A); Large right paramedian pelvic cyst protruding into the bladder (B).

also noted extending from the L4 vertebral level down to the cystic structure in the pelvis (**Figure 2**). To confirm this, we performed the transrectal ultrasound (TRUS) which demonstrated dilated right ejaculatory duct, which was contiguous with the cystically dilated right seminal vesicle. Dilated right ureter was also communicating with dilated right seminal vesicle. Cystically dilated portion of right seminal vesicle was protruding into the bladder lumen. The cystic structure showed internal echoes as well, probably due to hemorrhage or infection (**Figure 3**).

Further, MR urography was done to support the findings of TRUS as MR Urography is considered the gold standard imaging tool. MRI confirmed the cystic structure as the dilated right seminal vesicle which was protruding into the urinary bladder at the right VUJ. The partially formed & dilated right distal ureter was also seen draining into the cystically dilated right seminal vesicle. Signal intensity of the dilated right seminal vesicle as well as dilated right ureter showed T1 mild hyperintensity and lower signal intensity as compared to bladder content in urography images.

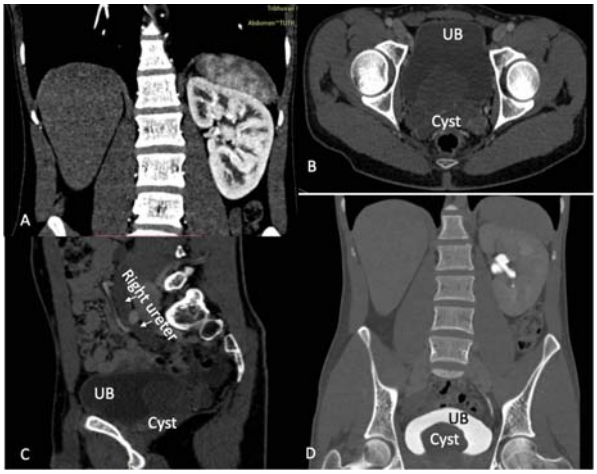

Figure 2. CT Urography images –
Empty right renal fossa with normal appearing left kidney (A); Slightly hyperdense (compared to the urinary bladder retrovesical cyst predominatly on right side, protruding into urinary bladder (B); Dilated right ureter opening into the cystic structure (C); Normally excreting left kidney with normally opacified left distal ureter, non-opacification of dilated right ureter and contrast in the urinary bladder and cystic structure appearing as filling defect within the urinary bladder (D).

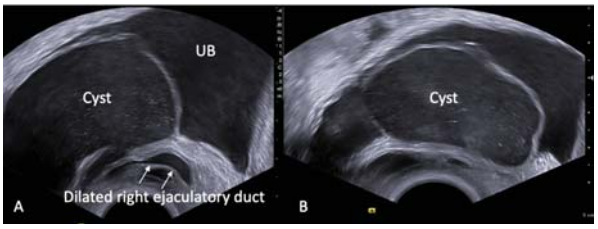

Figure 3. Transrectal sonography (TRUS) images showing dilated right ejaculatory duct communicating with dilated right seminal vesicle protruding into the urinary bladder (A); Rather than purely anechoic, there are innumerable punctate echogenic dots/ low level internal echoes seen within the cyst (B).

So, with all these imaging findings, the diagnosis of Zinner syndrome was given and the patient was counseled about the different management plans.

Discussion

Zinner syndrome is a rare congenital abnormality of the mesonephric (Wolffian) duct consisting of unilateral renal agenesis, ipsilateral seminal vesicle cyst, and ipsilateral ejaculatory duct obstruction.[3] Embryogenesis of the kidney, ureter, seminal vesicle and vas deferens can be altered if the insult occurs during the first trimester mainly between the 4th and 13th gestational week.[4] The ipsilateral ureter can be absent or incomplete or may have an abnormal course towards the seminal vesicle.[5,6] It is considered the male counterpart of Mayer-Rockitansky-Ku¨ster–Hauser syndrome.[7] Zinner described this syndrome for the first time in 1914, and about 200 cases have been reported in the literature.[8]

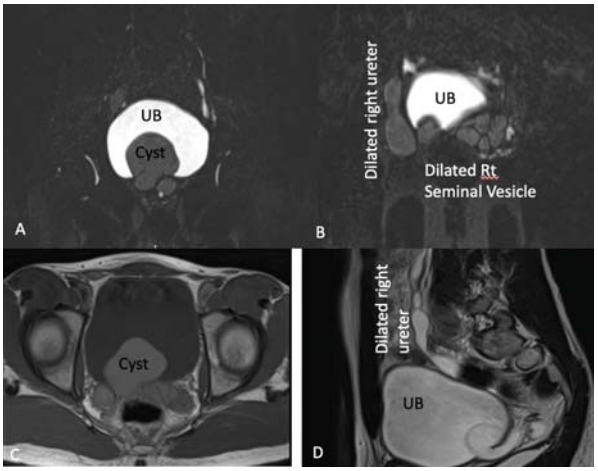

Figure 4. MR urography images demonstrating dilated right seminal vesicle with cystically dilated portion protruding into the bladder and the dilated right ureter (A,B); T1 weighted coronal image showing the dilated right seminal vesicle showing iso to mild high signal intensity content (C); T2 weighted sagittal images showing the dilated right ureter draining into the cystically dilated right seminal vesicle (D).

This syndrome is most often discovered incidentally. When symptomatic, usually presents in the second or third decade of life mainly after the beginning of sexual activity. The symptoms are mainly attributed to the cystic lesion of the seminal vesicle causing mass effect and often presents with voiding manifestations such as dysuria, prostatism, urgency, painful ejaculation, hematospermia, perineal pain/discomfort, and sometimes infertility.[9] The association of abnormalities is frequently found on the right side, with the right to the left ratio of 2:1.[10]

Magnetic resonance imaging (MRI) occupies a prominent place in the diagnostic arsenal and remains the examination of choice to make the diagnosis.[5] MRI can well delineate prostate and seminal vesicles, thus can confirm the location of the pelvic cyst within the seminal vesicle. MRI is superior to CT for diagnosis of the seminal vesicle cyst due to its high contrast resolution and more clear demonstration of anatomic relations in the pelvis.[11] However, renal agenesis and retrovesicle periprostatic cyst can be seen in CT as well. Another confirmatory imaging technique is vasovesiculography which will show reflux of contrast into the ipsilateral atretic ureter from dilated seminal vesicle due to ejaculatory duct obstruction.[3]

Transabdominal sonography is the initial investigation in most of the cases which will demonstrate the absent kidney in the renal fossa with ipsilateral cystic structure in the pelvis. Echotexture of the cyst may vary from anechoic cyst to low level internal echoes or hyperechoic contents within the cyst when hemorrhage or infection occurs.[12] TRUS may show dilated ejaculatory duct as well as communication of the periprostatic cyst with the dilated ejaculatory duct as seen in our case. Ultrasound guided transperineal aspiration or TRUS guided aspiration of the cyst fluid and identification of spermatozoa in it can also confirm the diagnosis.[3] Differential diagnosis of the seminal vesicle cyst include cysts of prostatic duct, prostatic utricle, vesical diverticula and ureterocele.[6]

Conclusion

Zinner syndrome is an uncommon cause of dysuria in men. This diagnosis should be suspected when young male presents with renal agenesis and ipsilateral pelvic cyst. CT may provide the diagnosis if it can precisely localize the origin of the cystic pelvic mass. However, MRI is considered the imaging modality of the choice which gives precise anatomical localization of the mass.

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