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Case Report

A rare case of Zinner syndrome: Triad of unilateral renal agenesis, ipsilateral seminal vesicle cyst and ejaculatory duct obstruction ☆,☆☆,★

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ABSTRACT

A rare triad of Wolffian duct anomaly known as Zinner syndrome includes unilateral renal agenesis with ipsilateral seminal vesicle cyst and ejaculatory duct obstruction. It is often diagnosed in third and fourth decades of life. Patient presents with dysuria, perineal pain, infertility and painful ejaculation. The aim of this case report is to show the importance of the radiological imaging on diagnosis of Zinner syndrome. MRI being the modality of the choice for the confirmation of the diagnosis is vital in further management of the syndrome.

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Introduction

Zinner syndrome is a rare congenital abnormality often diagnosed in third and fourth decades of life.[1] It is the combination of three abnormalities of Wolffian duct including seminal vesicle cyst, ipsilateral renal agenesis and ejaculatory duct obstruction. It is named after A. Zinner who was the first to describe this syndrome in 1914 A.D.[2] Here we report a case of 42 years old male presenting with non-specific genitourinary symptoms, who was later diagnosed with this rare condition of Zinner syndrome with the help of radiological imaging investigations.

Case

A 42 years old adult male presented to the emergency department with complaint of difficulty in passing urine for last few hours. The patient also complained having similar episodes previously with increased frequency of micturation and dysuria, however there was no history of dribbling or poor urinary stream. He had been married for 5 years and had a daughter. He had no known endocrine or systemic diseases. On presentation he was afebrile and his heart rate, blood pressure and oxygen saturation were within normal limits. On physical examination abdomen was mildly distended over hypogastrium due to the distended urinary bladder.

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* Patient Consent: A written, informed consent was obtained from the patient for his information about case history including the radiological diagnostic images would be used for publication in an open access medical journal under anonymity.

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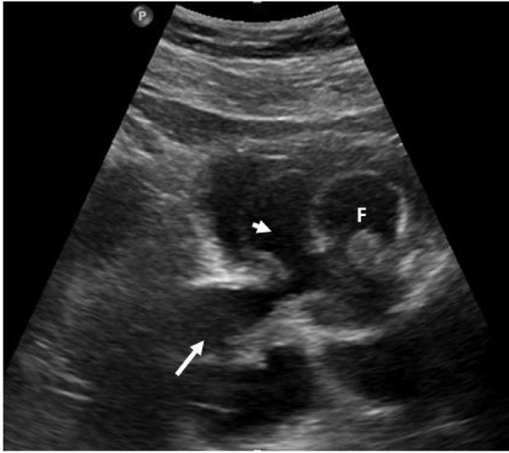


Fig. 1 – Sonography of pelvis showing right-sided retrovesical cystic lesion (long arrow) and right ureterocele (short arrow). F denotes Foley catheter bulb within the urinary bladder

Rests of the examination findings were unremarkable. External genitalia were well developed. Routine hematological and biochemical investigations were within normal limits. Urinalysis was normal.

For further evaluation of the cause of dysuria, patient was then referred to the department of Radiology and Imaging. An abdomen and pelvic ultrasound was performed which revealed empty right renal fossa with hypertrophied left kidney. A well-defined anechoic structure was noted in right retrovesical region which was initially assumed to be a right-sided hydronephrotic ectopic pelvic kidney. Separate well defined anechoic structure was also noted in the urinary bladder lumen arising from right vesicoureteric junction suggesting a ureterocele (Fig. 1).

CT (computed tomography) urogram of the patient was done. The findings revealed a well-defined fluid attenuating hypodense lesion measuring approximately 3×3 cm of size in right pelvic region at periprostatic area. No post contrast enhancement was appreciated and no vascular supply could be traced up to the lesion. No contrast excretion from this lesion was seen; hence ruling out the possibility of ectopic pelvic kidney. As right seminal vesicle was not identified separately from this lesion, provisional diagnosis of seminal vesicle cyst was made. Right sided ureterocele was also identified (Fig. 2). Only distal segment of right ureter was visualized which was dilated with blind ending proximal segment. The left kidney was hypertrophied measuring 14.5×4 cm and showed prompt excretion of the contrast. Left ureter and left seminal vesicle were morphologically normal.

To confirm the origin of this cystic lesion, MRI (magnetic resonance imaging) of abdomen and pelvis was performed which revealed a well-defined cystic lesion following homogeneous fluid signal intensity in all sequences measuring approximately 3.5×3 cm at the site of right seminal vesicle. There was tubular cystic dilatation of distal portion of right vas deferens. Findings identified in CT urogram of right ureterocele with dilated distal ureter and blind ending proximal segment was corroborative in MRI images (Fig. 3).

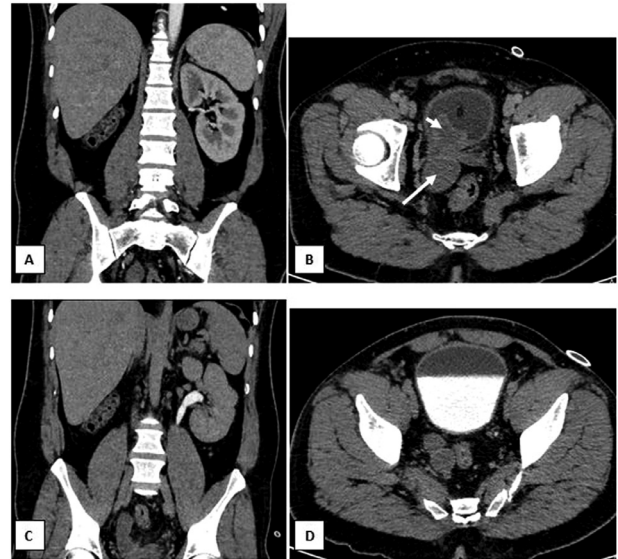


Fig. 2 – (A) Coronal reformatted CT image showing hypertrophied left kidney and absent right kidney. (B) Axial CT scan showing right-sided seminal vesicle cyst (long arrow) and right ureterocele (short arrow). (C, D) Coronal reformatted and axial CT images in delayed phase showing normal excretion of contrast by left kidney with accumulation of contrast in urinary bladder

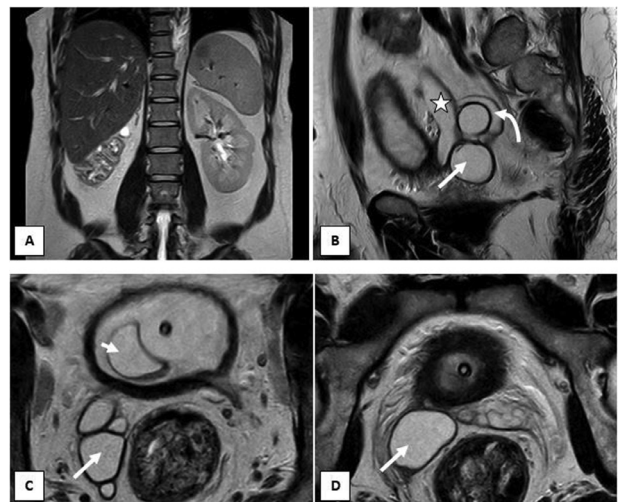


Fig. 3 – (A) Coronal T2 weighted MR image showing hypertrophied left kidney and absent right kidney. (B) Para-sagittal and (C, D) axial T2 weighted MR images showing right-sided seminal vesicle cyst (long arrow), right ureterocele (short arrow), enlarged vas deferens (curved arrow) and dilated distal right ureter (asterisk). Also note the normal vesicoureteric junction and seminal vesicle in left side

Thus, inclusive findings of USG, CT urogram and MRI of right renal agenesis with ipsilateral seminal vesicle cyst, tubular dilatation of ipsilateral vas deferens and right ureterocele with blind ending proximal ureter lead to the diagnosis of Zinner syndrome which is a mesonephric duct anomaly.

Discussion

Zinner syndrome is a Wolffian duct abnormality having triad of findings of unilateral renal agenesis, ipsilateral seminal vesicle cyst and ejaculatory duct obstruction.[2] It is considered to be the male counterpart of Mullerian duct anomaly which is characterized by vaginal agenesis and genitourinary abnormalities also known as Mayer-Rokitansky-Küster-Hauser syndrome.[3] In approximately 27% the ureteric bud remnant is noted. The ureter of same side to the renal agenesis may be absent, incomplete or have abnormal course.[4] The patient with Zinner syndrome presents with symptoms of difficulty in micturation, urinary obstruction, increase in frequency and urgency, painful ejaculation and epididymitis.[5] however it can be silent with no symptoms or patient may present with non-specific pain in perineal region or infertility. Seminal vesicle cyst smaller than 5 cm in size is usually asymptomatic, but large cyst can cause obstruction of urinary bladder and colon. Cysts larger than 12 cm are considered as giant cyst.[6] Semen analysis of the patient with Zinner syndrome shows decrease semen volume, oligozoospermia and azoospermia due to ejaculatory duct obstruction leading to infertility seen in approximately 45%.[2] Small sized testis and ureterocele of the same side have also been reported.[7] On digital rectal examination there may be presence of well-defined cystic lesion palpable adjacent to the seminal vesicle however it can remain undetected on physical examination.[8]

Different radiological imaging methods including ultrasonography, CT scan and MRI play important roles to evaluate such cystic lesions.[1] Ultrasonography being the initial tool of investigation reveals a well-defined anechoic cystic lesion in the pelvis with empty ipsilateral renal fossa.[9] CT scan of abdomen and pelvis is considered more superior than ultrasonography. On CT scan periprostatic fluid attenuating cystic lesion in posterior aspect of urinary bladder along with ipsilateral renal agenesis is reported, however this may not be sufficient to make the diagnosis.[10] MRI is the modality of choice for the accurate anatomical demonstration of the male genital tract and hence confirming the periprostatic cystic lesion whether originating from seminal vesicle or not.[11] On MRI the seminal vesicle cyst appears hypointense on T1 and hyperintense on T2 weighted images; high protein cyst being exception which would have high signal on T1 weighted and low signal on T2 weighted images. A convoluted tail communicating with the cystic lesion and seminal vesical will confirm the origin of the cyst.[1] Seminal vesicle cyst needs to be differentiated from ejaculatory duct cyst, ureterocele and urinary bladder diverticulum.[12] MRI plays an important role for distinguishing these possibilities. For symptomatic seminal vesicle cyst, the choice of treatment is aspiration via surgical or laparoscopic approach which will relieve the obstruction of ejaculatory duct and hence restore the fertility.[13]

Conclusion

Though being a rare condition, Zinner syndrome is encountered in adult males presenting with non-specific symptoms such as dysuria, infertility, perineal pain and painful ejaculation. Radiological imaging plays an important role in diagnosing this triad of unilateral renal agenesis ipsilateral seminal vesical cyst, and ejaculatory duct obstruction. Though ultrasonography and CT scan can be helpful for suggesting this diagnosis, MRI scanning remains the choice of modality for accurate anatomical delineation of male genital system and thus confirms the diagnosis.

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