Case Report

Zinner Syndrome: A Case Report of Rare Urogenital Anomaly

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Abstract

Zinner Syndrome is a rare urogenital tract anomaly with unilateral renal agenesis, ipsilateral seminal vesicle cyst, and ipsilateral ejaculatory duct obstruction. The syndrome occurs due to an insult to the development of the paramesonephric duct during embryogenesis. Patients may present late due to dysejaculation as a result of obstruction of the ejaculatory duct or commonly remain asymptomatic. Here, we present such a case presenting with dysejaculation which was diagnosed with Zinner syndrome on ultrasound.

Keywords: Dysejaculation, ejaculatory duct obstruction, renal agenesis, Zinner syndrome

Introduction

Ejaculatory pain also known as dysejaculation is a fairly common but poorly understood medical issue. This condition is more common in men with chronic prostatitis and chronic pelvic pain syndrome. The causes vary from the disorder of the ejaculatory pathway and prostate, sexually transmitted infection to vague psychological causes and the condition is often idiopathic.[1] Obstruction of the ejaculatory duct, congenital or acquired (stones, infection) can lead to dysejaculation.[2] The congenital obstruction of the ejaculatory duct can be associated with other urogenital development disorders. Here, we present a case of such association of ejaculatory duct obstruction and congenital absence of ipsilateral kidney-which is also known as Zinner syndrome.

Case Report

A 27-year-old young male patient was referred to the Radiology department for Trans-rectal ultrasonography (USG). He complained of ejaculatory pain, dysuria, and perineal discomfort for few weeks. His examination findings were unremarkable except for tenderness on the per-rectal examination. A semen culture demonstrated the growth of *Staphylococcus aureus*. A clinical diagnosis of chronic prostatitis was made and Trans-Rectal Ultrasound (TRUS) examination was ordered. TRUS examination was performed with probe (EC 9-4 in Acuson 9 × 3 Elite Siemens ultrasound system). The examination revealed tubular dilatation of the right ejaculatory duct with narrowing at the terminal part [Figure 1]. Cystic dilatation of the right seminal vesicle was also noted [Figure 2]. We performed abdominal USG to consider other associated urogenital anomalies. On USG, agenesis of the right kidney was also noted [Figure 3]. We had a doubt on Zinner syndrome. A Magnetic resonance examination was performed to confirm the diagnosis. Magnetic resonance imaging demonstrated cystic lesions in the seminal vesicle with tubular dilatation of the ejaculatory duct with the transition at the distal end near the urethral opening [Figure 4]. The absence of the right kidney was also documented [Figure 3]. We made a diagnosis of Zinner syndrome with secondary infection based on our findings. Differential diagnoses were acquired obstruction of ejaculatory duct and associated renal agenesis.

The patient was put on antibiotics for 2 weeks and transurethral resection of verumontanum with drainage of seminal vesicle cyst was performed.

Discussion

The ejaculatory duct, seminal vesicles, and vas difference all are developed from the paramesonephric duct. The distal mesonephric duct also forms the ureteric bud, which

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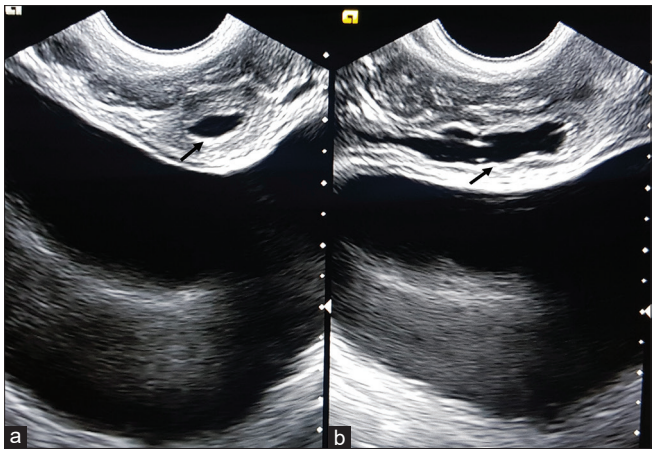

Figure 1: Trans-rectal ultrasound image demonstrating tubular dilatation of the right ejaculatory duct with narrowing at the terminal part (Black arrow)

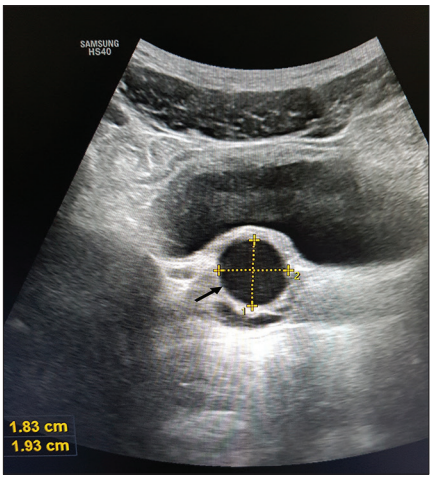

Figure 2: Ultrasound image demonstrating cystic lesion in seminal vesicle (Black arrow)

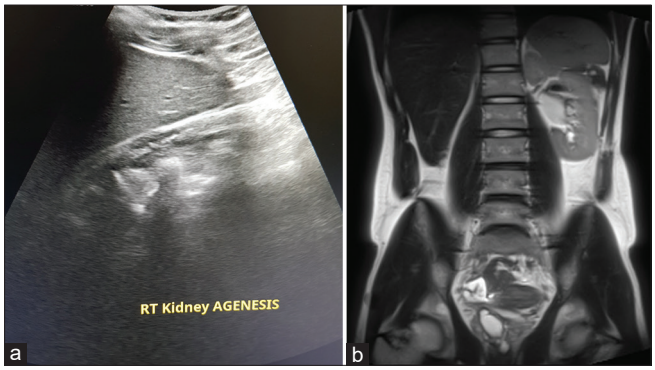

Figure 3: (a) Ultrasound image demonstrating absent right kidney in the right renal area. (b) Magnetic resonance imaging coronal image of the abdomen demonstrating absent right kidney

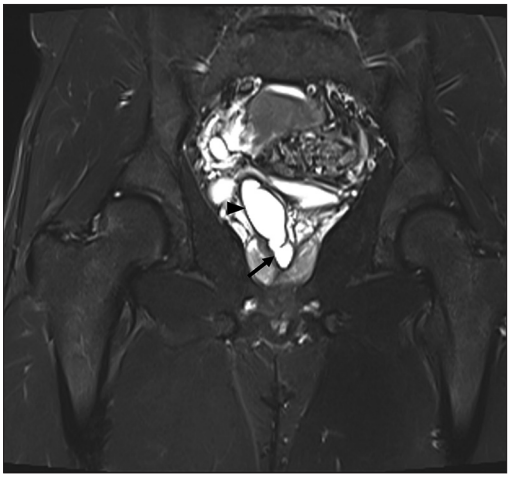

Figure 4: Magnetic resonance imaging Turbo inversion recovery magnitude sequence, coronal section showing cystic lesions in the seminal vesicle (Black arrowhead) with tubular dilatation of the ejaculatory duct (Black arrow) with the transition at the distal end near the urethral opening

induces the metanephric blastema to form kidneys. Thus, an insult to the distal paramesonephric duct during early embryogenesis will lead to an abnormality of kidneys and also cause obstruction of the ejaculatory duct leading to Zinner syndrome.[3-5] Zinner Syndrome is a rare urogenital tract anomaly with unilateral renal agenesis, ipsilateral seminal vesicle cyst, and ipsilateral ejaculatory duct obstruction. It is the male counterpart of Mayer–Rokitansky–Küster–Hauser syndrome seen in women.

Traditionally vasography was used as the investigation of choice to evaluate ejaculatory duct obstruction; however, due to its invasive nature and development of high resolution, TRUS and endo-rectal MRI are rarely used in current day practice.[3] TRUS findings in ejaculatory duct obstruction can be dilated ejaculatory duct, midline cystic lesions, hyperechoic lesions (calcifications), seminal vesicle cyst. Also dilated seminal vesicles may sometimes be present in normal fertile men, making the diagnosis of ejaculatory duct obstruction difficult. Midline cyst causing compression of ejaculatory duct may occasionally be present. These include utricular cyst, Mullerian duct cysts, and Wolffian duct cyst (containing sperm). It is extremely difficult to differentiate between these cysts on imaging. These cysts sometimes are difficult to differentiate from ejaculatory duct cysts on USG, however ejaculatory duct cysts tend to be slightly paramedian in location. Furthermore, the treatment of all these cysts is the same, i.e., transurethral resection of ejaculatory duct (TURED).[4,6,7]

In our case, ejaculatory duct was dilated with cystic dilatation of ipsilateral seminal vesicle making the diagnosis of ejaculatory duct obstruction. Furthermore, no midline cystic lesions causing compression of the ejaculatory duct could be seen.

MRI is the best investigation due to its multiplanner ability and excellent soft-tissue resolution. It gives detailed pelvic anatomy and differentiation of cystic masses providing a definitive diagnosis. The differential diagnosis is based on the location of the cyst, its relation with genitourinary abnormality

and its content. As with any cystic lesion in the body, seminal vesicle cysts appear low attenuation on T1-weighted images and high attenuation on T2-weighted images. The presence of high protein content or previous hemorrhage in the cyst leads to high signal on T1 and lower signal intensity on T2-weighted images. The presence of convoluted tail communicating with seminal vesicle and high signal on T1 images favor seminal vesicle cyst.[7,8]

Management includes follow up for asymptomatic cases while the symptomatic patients are treated with antibiotics for infected cyst, image-guided percutaneous aspiration, sclerosant injection, TURED or surgery. Our patient was treated with antibiotic and transurethral resection of verumontanum with drainage of seminal vesicle cyst. His first follow-up after surgery showed a significant reduction in the size of seminal vesicle cystic lesion.

Conclusion

The triad of ipsilateral renal agenesis and seminal vesicle cyst with ejaculatory duct obstruction is one of the rarest urogenital tract anomalies. Imaging techniques like USG abdomen, pelvis, and TRUS are useful to diagnose the abnormalities. Added imaging modalities such as CT scan and MRI provide additional benefits in the aid of the diagnosis. Basically, the management of choice in this syndrome is conservative with periodic follow-up of the patient. Surgical intervention is reserved only in the case of symptomatic patients. Knowledge of urogenital embryology helps to seek for and identify rare syndromes with multiple aberrations.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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