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ZINNER SYNDROME

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Abstract

Developmental malformations of the urogenital tract are rare, and most of them are cystic anomalies. In the embryogenesis, exactly during the first trimester of gestation developmental arrest affecting mesonephric (Wolffian) duct results in unilateral renal agenesis.

Ipsilateral seminal vesicle cyst also affecting the caudal end of Mullerian duct produces ipsilateral ejaculatory duct obstruction. Most of the patients with this malformation are asymptomatic until the 2nd or 3rd decade of life. Initially, majority of patients have nonspecific symptoms such as perineal discomfort, urinary urgency, prostatism, painful ejaculation and dysuria.

We report an uncommon case of a 24-year-old patient presented with symptoms of lower urinary tract irritation. Radiologic imaging modalities as ultrasonography, contrast-enhanced computed tomography and magnetic resonance imaging are all helpful in diagnosis of this extremely rare developmental anomaly.

***Keywords*:** MRI, seminal vesicles, ejaculatory duct, CT

Introduction

Zinner syndrome, known as the triad of renal agenesis, cysts in the ipsilateral seminal vesicle and ejaculatory duct obstruction, was described by A. Zinner in 1914 [1].

Congenital anomalies of the seminal vesicles are infrequent; most of them have cystic malformations and some are associated with malformations of the upper urinary system.

The common embryological origin of the urinary and genital tracts (mesonephros or Wolf ducts and the ureteric bud) can produce, in the presence of an anomaly, an alteration in the development of both systems [2]. It is often diagnosed in the second and third decades of life and can lead to serious complications, particularly infertility [3].

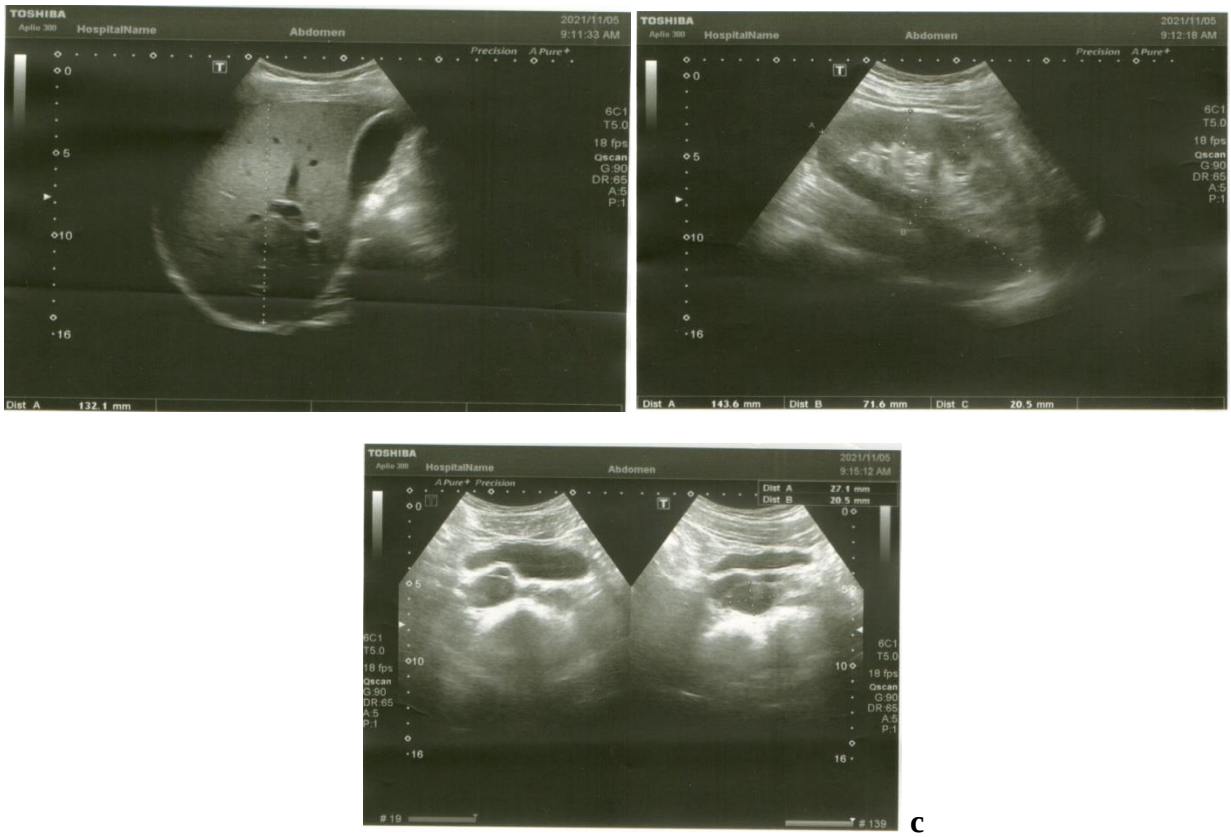
Magnetic resonance imaging (MRI) has a prominent place in the diagnostic arsenal and remains the examination of choice to make the diagnosis [4].

Case report

We report an unusual case of a 24-year-old male who presented with history of irritative lower urinary tract symptoms, intermittent hematuria and perineal discomfort. Physical examination revealed no palpable pelvic mass and external genital organs were normally developed. The patient had no endocrine or systemic disease.

Ultrasonography of abdominal organs detected an empty right renal fossa (Figure 1A), and compensatory hypertrophy of the left kidney (Figure 1b). The other organs in the abdomen were normal. Ultrasonography of pelvis revealed a right retrovesical cystic mass (27x20 mm) in the projection of the right seminal vesicle (Figure 2).

a **b**

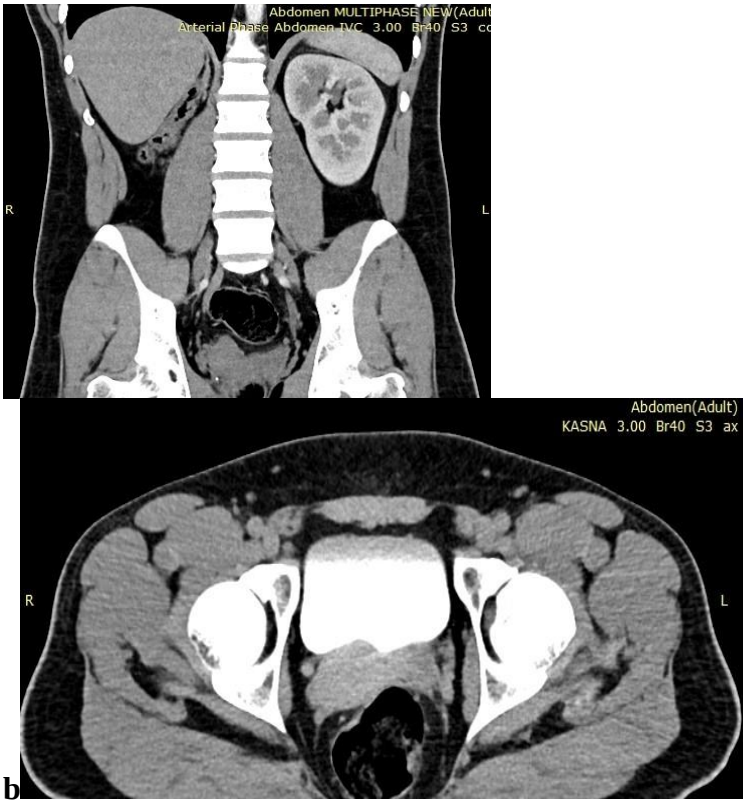


c

Figure 1. (a) Absent kidney in the right renal fossa. (**b**) Abdominal ultrasound showed compensatory hypertrophy of the left kidney (14x7 cm). (**c**) Cystic lesion right of the urinary bladder extending up to the midline**.**

Abdominal and pelvis contrast-enhanced computed tomography showed right renal agenesia. At the same side in the periprostatic region, inferolateral to the urinary bladder, a fluid-attenuation non-enhancing mass was seen (Figure 2a and 2b). There was no contrast concentration on the delayed scans by the cystic mass that ruled out the opinion for ectopic kidney.

a



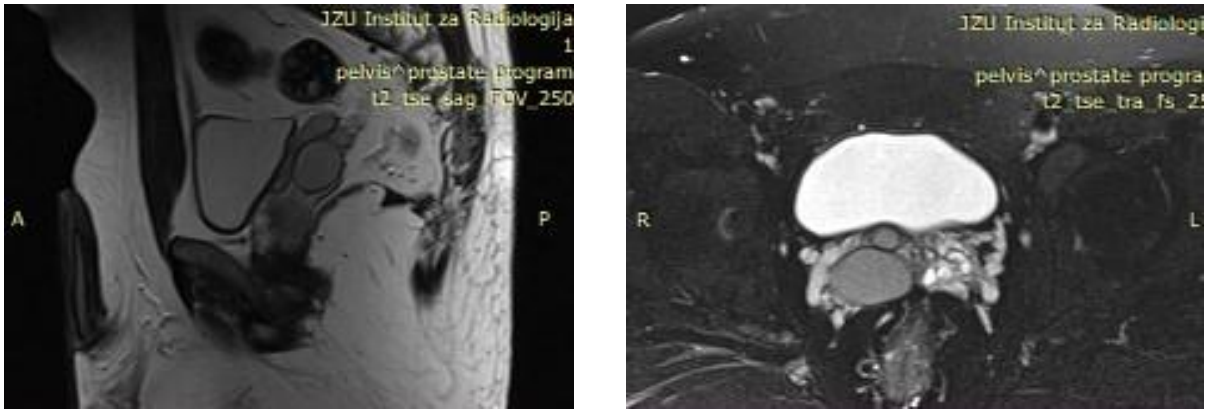
b

Figure 2. Contrast enhanced computed tomography (CECT) of abdomen and pelvis. (**a**) Coronal contrast enhanced scan revealed empty right renal fossa and compensatory hypertrophy of the left kidney that showed normal CT features. (**b**) Axial image showed large well-marginated non-enhancing mass in the periprosthetic region between the bladder and rectum.

Pelvic magnetic resonance imaging (MRI) was performed with basal sequences, which demonstrated multiple cystic to tubular structures in the right seminal vesicle that appeared hyperintense on T2-weighted images and hypointense on T1-weighted images as well as a fluid level suggesting proteinaceous content. On the sequence with diffusion (DWI) there was no restriction and in T2-weighted images with gadolinium injection there was no enhancing mass.

a **b**

Figure 3. (a) Axial T1W MR image: Cystic hypointense structures lying close together in the region of right seminal vesicle, indenting the right lateral bladder wall and the bladder base. (**b**) Sagittal T1W MR image: the same cystic tubular dilated structures visible behind the bladder in sagittal reconstruction.



a **b**

Figure 4. Pelvic MRI. (a) Sagittal T2 FOV showed large cystic structure retrovesical in the region of the right seminal vesicle. (b) Axial T2 fat saturation post-gadolinium: cystic proteinaceous content structure lying close together in the region of the right seminal vesicle.

Imaging findings lead to the diagnosis of Zinner syndrome. Being aware of the impact of this syndrome on the fertility status, the patient was asked to undergo a semen analysis which showed normal values (sperm count was 70.000.000/ml with a semen volume of 3,6 mL). Blood routine examination and urine laboratory tests including urine culture, were also normal.

Discussion

Zinner syndrome is one of the rarest congenital abnormalities of the urogenital system, which is usually present and diagnosed during the 2nd to the 4th decade of life [5].

The incidence is 1 in 3,000 to 1 in 4,000 newborns. Patients with Zinner syndrome do not present usually with any problems, but sometimes they have decreased urine output, increased urinary urgency, perineum pain, or epididymitis with urine infection. Infertility should be ruled out in male patients.

The conservative treatment is a choice in asymptomatic patients. Infertility and its pathogenesis are unclear, but it is more likely due to the associated ejaculatory duct obstruction. Examination of seminal fluid should be systematic in the case of Zinner syndrome; results can show a low ejaculatory volume, azoospermia, alkaline pH, low concentration of carnitine and fructose in the seminal plasma, and high citrate level [6].

Imaging remains the key method to diagnosis. CT scan is superior to ultrasonography, and reported findings are cystic pelvic mass arising from an enlarged seminal vesicle associated with ipsilateral renal agenesia, but it may be insufficient to confirm the diagnosis.

MRI is considered to be superior to CT scan in the analysis of these conditions [4].

With its invasiveness, multiple pulse sequences, multiplanar capability, and high soft-tissue resolution, it is considered as "gold standard" for diagnosis and clinical follow-up.

Conclusion

The combination of ipsilateral renal agenesia and seminal vesicle cyst is one of the rarest urogenital tract anomalies. Some imaging techniques like ultrasonography, computed tomography and MRI of the abdomen and pelvis will detect the rare urogenital tract anomalies.

Ultrasonography is a noninvasive examination and could provide valuable information. CT scan could be sufficient in establishing the diagnosis if the origin of the pelvic mass is defined. MRI is the method of choice for establishing a precise diagnosis and successful therapeutic management.

The conservative treatment is the principal plan in asymptomatic patients with regular follow-ups. Surgical interventions are required in symptomatic patients, and they include surgical aspiration of cysts through perineal or laparoscopic approach or percutaneous cyst drainage. Our patient was diagnosed with Zinner syndrome incidentally by CT scan. The patient was asymptomatic and was managed with regular follow-ups.

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