CASE REPORT
Pediatric Urology

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A Young Boy with Renal Agenesis and Ectopic Seminal Vesicle: Zinner Syndrome

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Abstract

Zinner syndrome is a rare disorder, and it is associated with unilateral renal agenesis, ipsilateral ejaculatory duct atresia (obstruction), and cystic dilatation of the ipsilateral seminal vesicle. Here, we present an eighteen-year-old male with Zinner syndrome who presented to a urology clinic with perineal/pelvic pain and dysuria. We also emphasize the computed tomography and magnetic resonance imaging findings of Zinner syndrome.

Keywords: Imaging, congenital malformations, renal agenesis

Introduction

Zinner syndrome is a rare disease that is seen in males. It occurs due to a developmental abnormality of the distal part of the Wolffian duct. Zinner first described the Zinner syndrome in 1914 as unilateral renal agenesis, ipsilateral ejaculatory duct obstruction, and ipsilateral seminal vesicle cyst (1-3). It is also accepted as the male equivalent of Müllerian agenesis (Mayer-Rokitansky syndrome) seen in females. Zinner syndrome is associated with unilateral renal agenesis, ipsilateral ejaculatory duct atresia (obstruction), and cystic dilatation of the ipsilateral seminal vesicle (3). The pathogenesis of Zinner syndrome is related to the mesonephric (Wolffian) duct (4). The mesonephric (Wolffian) duct develops into the prostatic part of the urethra, ductus deferens, seminal vesicle, and epididymis owing to testosterone and anti-mullerian hormone (5). An abnormality of development of the distal part of the Wolffian duct can because by atresia (obstruction) of the ejaculatory duct, leading to the dilatation and cysts of the ipsilateral seminal vesicle. Furthermore, the concurrent ureteral budding abnormalities can lead to renal agenesis or dysplasia. These developmental pathologies can result in azoospermia and oligospermia, which cause primer infertility in males (3-5). Also, seminal vesicle cysts and cystic dilatation of the ipsilateral seminal vesicle can have mass effects and pelvic perineal pain (4,5). Diagnosis of Zinner syndrome can be made by ultrasonography or computed tomography (CT) scan; but magnetic resonance imaging (MRI) gives a better resolution of the local anatomy. Treatment is usually conservative, but surgery may be required in selected cases.

Case Report

An eighteen-year-old boy presented to the urologic clinic with pelvic pain and dysuria. Physical examination and laboratory test results, including complete biochemical profile, C-reactive protein, blood count, and urinalysis results, were within normal limits. A urine specimen for culture was obtained. An abdominal ultrasound showed right renal agenesis and a 31x40x52 mm heterogeneous cystic lesion in the right lower quadrant. Unenhanced abdominal CT showed the right renal agenesis and ipsilateral pelvic lesion with well-defined borders (Figure 1). The following pelvic MRI revealed that the ipsilateral pelvic lesion was a cyst with proteinaceous/hemorrhagic content and was compatible with an ectopic and dilated right seminal vesicle (Figure 2). Zinner syndrome should be considered first in the differential diagnosis, characterized by unilateral renal agenesis, ipsilateral ejaculatory duct atresia (obstruction), and cystic

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dilatation of the ipsilateral seminal vesicle (1). After diagnosis, sperm analysis results were within normal limits, except for mild hematospermia. Laparoscopic surgical resection of the right seminal vesicle was performed due to hematospermia and clinical symptoms in this case. The patient was discharged without complications on the second day after the operation. The one-year follow-up of the patient was uneventful and her clinical complaints wholly regressed. Informed consent was obtained from the patient.

Discussion

Zinner syndrome is an uncommon anomaly, and its frequency is not known precisely. According to a study on ultrasonography, the incidence of seminal vesicle cysts in patients with ipsilateral renal dysplasia or aplasia (agenesis) is 0.0046% (6). The first-line examination of the assessment of Wolffian duct anomaly is the ultrasound owing to its non-invasive, radiation-free, and easily applicable nature. On ultrasound, seminal cysts are seen as a cystic pelvic mass that can demonstrate thick or irregular wall and mural calcifications. The internal echoes depend on hemorrhage or infection (1,3). Excretory urography and kidney scintigraphy can indicate ipsilateral kidney agenesis or dysgenesis. Pelvic MRI is an appropriate tool for the detailed examination of pelvic anomalies. It is a gold standard modality for evaluating pelvic abnormalities due to magnificent soft-tissue resolution and multiplanar visualization without radiation. When an absence of urogenital structures is detected on imaging, it should be cautiously examined for possible other Wolffian duct anomalies such as renal agenesis and seminal vesicle cyst (3). While figuring out the etiology of abdominal pain caused by the pressure effects of the seminal vesicle cyst over the adjacent structures, the Zinner syndrome can be diagnosed as in this study (4,5). However, the patients can admit primering infertility caused by azoospermia and oligospermia (6).

Cystic lesions of the seminal vesicle may imitate other pelvic lesions. That's why an accurate diagnosis is required. The differentiation of the cystic pelvic lesions is mainly based on the position as median, paramedian, or lateral (2). Also, abdominal pain causes, such as acute appendicitis, should be excluded. Ultrasonography or CT should be used for differential diagnosis if needed (5,6). Early diagnosis is principal to prevent complications such as infertility in patients with Zinner syndrome and

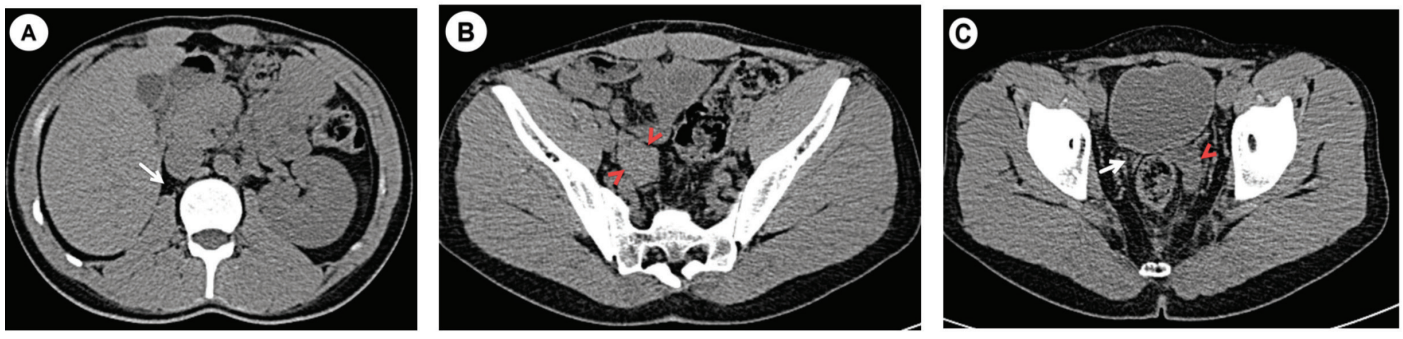

Figure 1. a) Axial unenhanced computed tomography (CT) image at the level of the upper abdomen shows right renal agenesis (arrow). **b)** Axial CT image at the level of the lower abdomen shows well-defined pelvic lesion (arrowheads). **c)** Axial CT image at the level of prostate demonstrates the normal left seminal vesicle (arrowhead) and the absence of the right seminal vesicle (arrow)

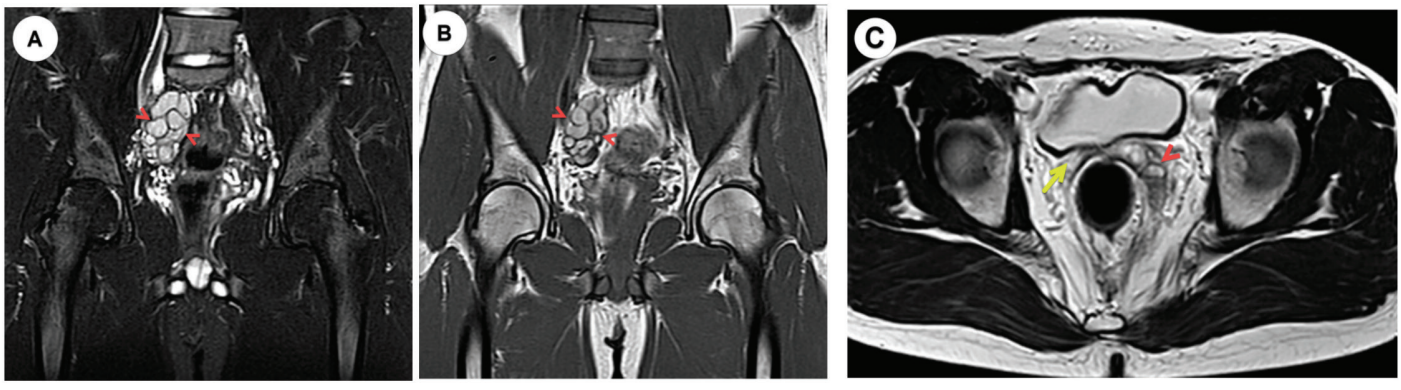

Figure 2. a) T2-weighted and **b)** T1-weighted coronal magnetic resonance (MR) images of the pelvis show the hyperintense pelvic lesion on both sequences is consistent with a proteinaceous/hemorrhagic cyst (arrowheads). **c)** T1-weighted axial MR image at the level of prostate demonstrates the normal left seminal vesicle (arrowhead) and the absence of the right seminal vesicle (arrow). The right pelvic mass is consistent with ectopic and a dilated right seminal vesicle, according to pelvic MR imaging findings

avoid nephrotoxic drugs. surgical excision may be considered depending on the size and location of the seminal vesicle cysts and clinical symptoms. Treatment is often conservative if the patient is asymptomatic (1,5). Moreover, since renal agenesis will occur in a patient with Zinner syndrome, a single kidney should be protected by measures such as avoiding nephrotoxic drugs and nephrotoxic contrast agents.

Conclusion

In conclusion, Zinner syndrome is an unusual anomaly, which includes unilateral kidney agenesis, ipsilateral ejaculatory duct obstruction, and seminal vesicle cyst. Moreover, seminal vesicle ectopia and cystic dilatation may also be seen in patients with Zinner syndrome, as in this case. Zinner syndrome should be kept in mind in the differential diagnosis of young patients with abdominal pain, infertility, and urinary symptoms.

Ethics

Informed Consent: Informed consent was obtained from the patient.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: S.Ç., Concept: A.Y.Y., F.U., Design: A.Y.Y., F.U., Data Collection or Processing: A.Y.Y., F.U., S.Ç., Analysis or Interpretation: F.U., Literature Search: A.Y.Y., F.U., Writing: A.Y.Y., F.U.

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