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Sonographic Characteristics of Zinner Syndrome; An Interesting Case Report

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Abstract

Background: Zinner syndrome is a rare congenital defect, typically manifests in young males with a variety of symptoms. The image findings are clearly recognizable, and recent therapies have been well-documented. Cystic lesions of the seminal vesicle can appear in huge numbers of patients. Due to the fact that the seminal vesicles and ureteral buds arise from the Wolffian duct, associated ipsilateral renal agenesis is a characteristic detected in many cases of ZS. For management, the ultrasound offers a clear view of the kidneys, and the rest of the pelvic structures. Case presentation: We present a case of a 25 years old non-married male with a seminal vesicle lesion. The patient first complained of recurrent urinary tract infection and dysuria. After being assessed clinically and radiologically using several modalities, the patient ultimately decided to live with the condition without receiving any immediate treatment, but he will continue his follow up schedule. **Conclusions:** US imaging has the potential role to deliver an accurate diagnosis for this case and confirmed CT scan.

Keywords: Zinner syndrome, Renal agenesis, Seminal vesicle cyst, infertility Ultrasonography.

Introduction:

Zinner syndrome is a rare congenital defect, typically manifests in young males with a variety of symptoms. The image findings are clearly recognizable, and recent therapies have been well-documented.[1]

ZS is characterized by ipsilateral renal agenesis, ejaculatory duct obstruction (EDO), and cyst ic seminal vesicles. ZS typically doesn't show any symptoms until sexual activity starts. EDO causes a buildup of seminal fluid, which then results in larger seminal vesicles.[2]

It is uncommon for congenital seminal vesicle cysts (CSVCs) to be linked to malformations in the opposite upper urinary tract.[3] The term "Zinner syndrome" has been used to describe this disorder, which is thought to be the male equivalent of the Mayer-Rokitansky-Kuster-Hauser syndrome.[4] ZS is asymptomatic and inadvertently discovered, or it can be linked to symptoms without a known cause, such as infertility, bladder dysfunction, dysuria, and urinary tract infection (UTI).[3]

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The syndrome typically manifests as voiding signs such as perineal discomfort or soreness, dysuria, prostatism, urgency, painful ejaculation, hematospermia, and dysuria, and occasionally infertility in the 2nd and 3rd decades of life (after the onset of sexual activity).[5] It can be difficult to identify infertility in adolescent patients. Clinical management of ZS is primarily focused on pain alleviation and protection of the contralateral ejaculatory duct to preserve fertility because surgical treatment is only prescribed for symptomatic patients.[6]
Diagnostic tools for this uncommon entity include Intravenous Pyelography (IVP), US, Vasovesiculography, contrast enhanced Computed Tomography (CT), and Magnetic Resonance Imaging (MRI). Here, we describe a remarkably uncommon Mullerian duct developmental abnormality that would go undetected without radiologic imaging.
Lower urinary tract irritation symptoms were found in the patient. [7]
In this report, we describe a patient whose primary complaints were recurrent epididymitis and infertility. Unfortunately, there is no agreement on the ideal therapeutic approach. We sum up disease mechanisms, the diagnostic procedure, and treatment strategies in the debate.

Case presentation:
A 25-years-old non married male, arrived to the urology clinics of Tadawi medical Centre at Abha city, complaining of history of dysuria, absence of second testis. The responsible doctor examined the patient and then asked for abdomino-pelvic US, a urine analysis, and a complete blood chart for examination.
Without any indications of UTI or crystals to suggest urinary tract stones, the results of the urine analysis, and CBC were normal.
Ultrasound results revealed right renal agenesis, with enlarged left kidney measures 12.5 x 6.7 cm) and utricle cyst measure (3.5 x3 cm), at right seminal vesicle, and right testicular a genesis.
CT was recommended for this patient for confirmation of the diagnosis and revealed same diagnosis; Zinner syndrome.
CT exam was done to the patient revealed that the findings were the same. The rest of the abdominal organs and bowel seemed ordinary, with the exception of minor compensatory enlargement of the left kidney. A CT pyelogram was then performed to the patient, confirming the right kidney's absence and demonstrating the absence of any renal arteries or veins. A large cystic lesion of the right seminal vesicle was also visible on contrast-enhanced CT without enhancement. The lesion, which was roughly (3.5 x3 cm), and showed a dilated right seminal vesicle cyst.

Discussion:
ZS is a rare congenital condition that was initially identified by A. Zinner in 1914.[8]
The diagnostic approach based on physical examination, abdominopelvic US, abdominopelvic CT and pelvic MRI. Both CT and MRI are reliable diagnostic modalities used to confirm the suggested diagnosis of US. The seminal vesicle cyst (SVC), ipsilateral renal agenesis or multicystic dysplastic kidney, and ejaculatory duct obstruction are the three symptoms of ZS,

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which is caused by an abnormality of the Wolffian duct during embryogenesis; however, other pelvic cystic structures must be ruled out with initial investigations.[4]

It is distinguished by an ipsilateral upper urinary tract and seminal vesicle deformity. The triad of an SVC, ipsilateral renal agenesis or multicystic dysplastic kidney, and ejaculatory duct obstruction can also be used to diagnose the illness. When the patients start having sexual activity in their second 2nd or 3rd decade of life, they are typically diagnosed. Epididymitis (27%), dysuria (37%), perineal discomfort (29%) and frequency (33%) are common symptoms upon presentation. [9]

Between the ages of 17 and 41, papillary adenocarcinoma and squamous cell carcinoma have been recorded seldom in SVCs associated with renal agenesis, but no patient in the pediatric age period has been described in literature as developing seminal vesicle malignancy. [10]

Clinical signs often peak when the time when sexual activity is at its highest level. The most typical accompanying symptoms are hematospermia, recurring episodes of apparent hematuria, and ejaculation failure. Additionally, documented symptoms include frequent urination, perianal soreness, and intermittent scrotal pain. [11,12]

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Fertility status is still a major worry that needs to be carefully explored. Infertility may continue after surgery, and assisted reproductive treatments seem to be the only solution. [4]

The majority of the time, ipsilateral urinary duct malformations resulting from the same embryonic anatomy are linked to congenital seminal vesicle problems. Male ectopic ureter is less common than female ectopic ureter because males typically present asymptomatically (ratio 1:2.9). Men frequently have an ectopic area in the seminal vesicle (37%) or posterior urethra (33%). In 58% of cases with ectopic ureters, renal abnormalities like renal agenesis and dysgenesis are found. [13]

Also, Van den study [14] showed a right-sided preponderance of the condition, which was not the situation in our instance. A 4-year-old boy who presented with a right paravesical cyst on ultrasound and was first identified as having a ureterocele was the subject of a case study in one of the previous studies.[3]

Ultimately, the diagnosis of ZS was made following 11 years of yearly US follow-up. Intriguingly, Pavan et al. [15] reported a singular case of the syndrome in a middle-aged male with a palpable, painless paratesticular lump that mimicked a varicocele that occurred in the same year. The successful robotic repair of the condition in a 23-year-old male with a history of lower urinary tract symptoms for two years, perineal pain, and recurrent UTI was documented by Kiremit et al. [16]

Although intrusive procedures could be necessary, the starting strategy should always be medical. Open surgery is no longer an option; laparoscopic and robotic procedures must be used instead. [4]

Conclusion:

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In order to assess complex urogenital tract developmental abnormalities, the right imaging modalities are crucial. We anticipate that employing an appropriate imaging tool, such as ultrasonography, because of its distinctive and recognizable imaging appearance should be sufficient for an experienced radiologist to be able to make an accurate diagnosis and occasionally even contribute to management. In this case ultrasound imaging alone has the potential to deliver an accurate diagnosis and confirmed by other radiological modalities. If there are renal anomalies, a comprehensive radiological imaging examination of the abdomeno-pelvis, including a CT and MRI, is required.

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Informed consent

Written & Oral informed consent was obtained from participant included in the study.

Conflicts of interest

The authors declare that they have no conflict of interest.

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Data and materials availability

All data associated with this study are present in the paper.

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