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Case Report

Zinner syndrome: A rare congenital cause of infertility ☆

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ABSTRACT

Zinner syndrome is a rare congenital malformation characterized by cystic seminal vesicles and ejaculatory duct obstruction in association with ipsilateral renal agenesis. It appears to be frequently linked to infertility. However, recent advances in imaging, notably MRI, have led to an increase in the diagnosis of this pathology. We describe the case of a 39-year-old patient receiving examination for primary infertility who was identified with Zinner syndrome using ultrasound, CT, and MRI; the patient did not report hemospermia, lower urinary tract symptoms, or perineal pain. Examining the abdomen and external genitalia revealed no abnormalities, and examining the rectal area revealed none at all. Zinner syndrome is a rare congenital condition; we report this case to highlight the etiopathogenesis of this seminal anomaly, its relationship with renal dysgenesis, and to illustrate the imaging of this condition through various diagnostic methods

Introduction

About 214 cases of Zinner syndrome, a rare congenital abnormality of the seminal vesicles and ipsilateral upper urinary tract, have been published in the literature between 1999 and 2020 [2]. Dr. Zinner initially characterized Zinner syndrome in 1914 [1]. Although it is usually discovered in the third or fourth decade of life, early detection is possible because of the growing applications of magnetic resonance imaging (MRI) and computed tomography [3]. Infertility affects up to 45% of people with Zinner Syndrome [4]. In the course of evaluating a 39-year-old patient for primary infertility, we report a case of Zinner syndrome, using ultrasound, CT, and MRI to illustrate the imaging of this particular condition.

Case presentation

We report the case of a 39-year-old patient, married and without children, with no notable medical history, who was referred to the radiology department for pelvic ultrasound as part of a primary infertility assessment. He did not report

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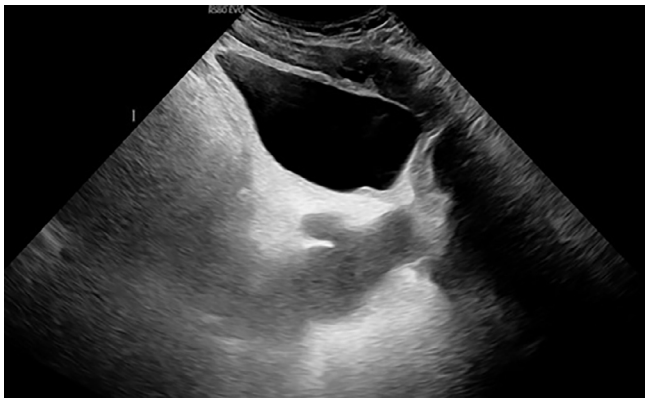

Fig. 1 – Pelvic ultrasound axial image showing a cystic left lateralized formation (arrow).

perineal pain, hemospermia or lower urinary tract symptom. Abdominal and external genital examination were unremarkable and rectal examination had no detectable abnormality. Pelvic ultrasound revealed a cyst in the left seminal vesicle measuring 70 × 39 mm. The cyst had a thin wall, regular contours and anechoic contents (Fig. 1). The scrotal ultrasound performed also revealed bilateral testicular hypoplasia. The abdominal-pelvic CT scan performed (Fig. 2) showed, in addition to the left seminal cyst, agenesis of the left kidney. The complement by pelvic MRI allowed a better analysis of the seminal cyst (Fig. 3). The association of seminal vesicle cyst with ipsilateral renal agenesis led to the conclusion of Zinner syndrome. The patient was referred to the urology department, where treatment by trans-urethral resection of the seminal bladder cyst was proposed given the size of the cyst.

Discussion

Since the ureteric buds and seminal vesicles come from the mesonephric (Wolffian) duct, congenital anomalies of these structures are frequently linked to abnormalities of the ipsilateral upper urinary tract. This duct is essential to the development of the human embryo. Under the influence of testosterone and anti-Müllerian hormone, it gives birth to the hemitrigone, bladder neck, urethra, seminal vesicle, vas deferens, epididymis, and epididymal heads in males. Thus, every anomaly that occurs in the first trimester may have simultaneous effects on the kidney, ureter, seminal vesicle, and vas deferens development [5]. The syndrome typically becomes apparent in the second and third decades of life, particularly with the initiation of sexual activity. After analyzing 214 patients with Zinner syndrome, Tianzhu Liu found that the most common symptoms were: frequency (24.3%), dysuria (23.1%), perineal pain (20.2%) [2]. Zinner syndrome was only accidentally discovered in our case, even though the cyst grew to a size of 7 centimeters. Infertility (13.9%) and other abnormalities of reproductive function were also noted [2]. Because the contralateral ejaculatory duct is intact in these people, the pathophysiology of infertility in these patients is poorly known. Nevertheless, it has been proposed that autoantibodies against sperm may develop as a result of unilateral blockage [6]. Transrectal ultrasonography, MRI, CT scan, and cystoscopy are examples of diagnostic instruments.

Imaging is crucial in the diagnosis of Zinner syndrome. Abdominopelvic ultrasonography, for example, shows the absence of an ipsilateral kidney and verifies the mass's cystic nature while also identifying its location and size. Transrectal ultrasonography reveals a cystic pelvic lesion with anechoic content, barring superinfection, and gives additional information on the cyst wall and its contents. It also aids in directing the surgeon in the case that the cyst needs to be removed transrectally. The ipsilateral ureter and kidney are confirmed to be absent by a CT urogram, yet the origin of the cyst is typically not confirmed. The most efficient diagnostic procedure is magnetic resonance imaging (MRI) [2]. This technique helps with surgical decision-making and approach in addition to definitively diagnosing the cystic component, which is characterized by low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. Additionally, MRI enables a more thorough examination of the glands and can verify ejaculatory duct blockage [7]. Other cystic pelvic lesions such as abscesses, ectopic ureterocele, diverticulosis of

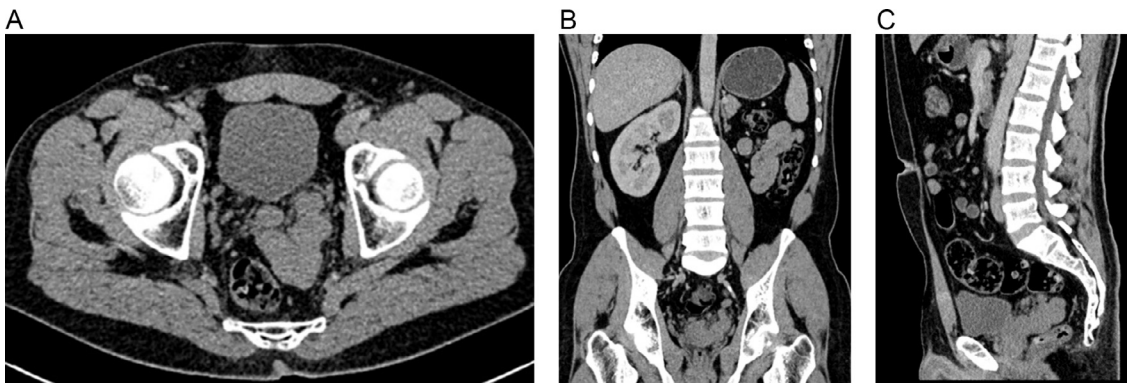

Fig. 2 – Contrast-enhanced abdominal-pelvic computed tomography, axial image (A) showing a posterior cystic lesion of the bladder (arrow). Coronal image (B) showing the left renal agenesis and the pelvic left cystic lesion (arrow). Sagittal image (C) showing the left renal agenesis and the pelvic left cystic lesion (arrow).

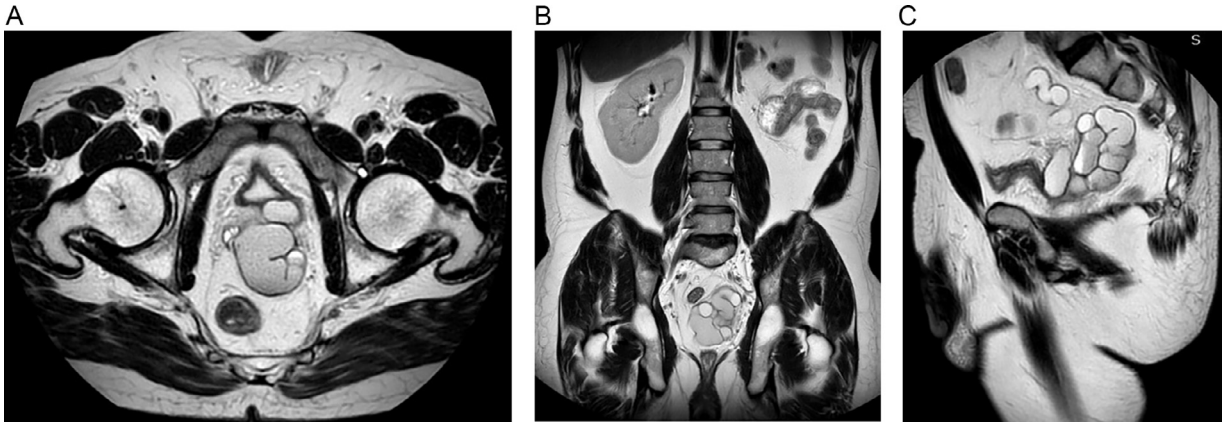

Fig. 3 – Pelvic MRI T2-weighted; axial image (A) showing a large smooth walled cyst in left seminal vesicle with hyperintense contents and presence of fluid-fluid level (arrowhead) and hypoplastic right seminal vesicle (arrow), coronal image (B) showing the left renal agenesis and the pelvic left cystic lesion (arrow), Sagittal image (C) showing the pelvic left tubular cystic lesion (arrow).

the vas deferens, and laterally situated prostatic cysts should be included in the differential diagnosis [8]. An incomplete trigone with a sizable cyst of the seminal vesicle in charge of extrinsic compression can be seen via cystoscopy [9].

For asymptomatic patients basic observation is advised. Treatment for symptomatic patients includes transurethral excision of the seminal vesicle cyst and ejaculatory duct, as well as ultrasound-guided aspiration [10]. For seminal vesicle cysts, laparoscopic surgery seems to be the best suitable surgical procedure at the moment. Compared to open surgery, it provides superb depth imaging and direct access to the seminal vesicle. Furthermore, it permits the seminal vesicle to be separated from the peritoneum surrounding the prostate and bladder without injuring the bladder or the lower intestines [11].

Conclusion

A rare urological anomaly is seminal vesicle cysts linked to ipsilateral renal agenesis or hypoplasia. This case illustrates the incidental discovery of this malformation in adulthood. A clinical examination and several imaging procedures, such as transpubic ultrasound, CT scan, and MRI, were part of the diagnosis process.

Patient consent

Regarding the manuscript titled "Zinner syndrome, a rare congenital cause of infertility''. I would like to confirm, on behalf of my co-authors and myself, that we have obtained all the consents required by the current legislation for the publication of personal data or images of patients, subjects of investigation, or other individuals depicted in the materials submitted to your Journal.

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