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Zinner Syndrome: Left Unilateral Renal Dysgenesis with Seminal Vesicle Cyst and Ipsilateral Testicular Atrophy Associated with Azoospermia - Case Report

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Abstract **Case Report**

Seminal vesicular cysts associated with ipsilateral renal agenesis or hypoplasia are a rare urological abnormality. This combination of urinary and genital abnormalities occurs due to the closely related embryological origin of these structures from the distal mesonephric duct [Wolff]. This case illustrates the discovery of this malformation in adulthood following chronic pelvic pain associated with infertility. The diagnostic approach consisted of a clinical examination with the performance of a set of imaging examinations including a suprapubic ultrasound, a scanner and an MRI. While the therapeutic management was limited to an ultrasound-guided evacuating puncture of the cyst of the seminal vesicle with a bacteriological and histological analysis of the puncture liquid. Given the complexity of the case, our surgical treatment is mainly aimed at relieving pain by evacuating puncture and offering medically assisted procreation for her infertility. We report the case of a young patient with Zinner's syndrome associated with infertility.

Keywords: Seminal vesicular cysts, urological abnormality, malformation.

I. BACKGROUND

Zinner syndrome is a rare congenital anomaly often diagnosed during the third and fourth decades of life, the period of greatest sexual and reproductive activity [1]. With fewer than 200 cases reported in the literature [2]. It is a combination of three abnormalities of the Wolffian duct, including a seminal vesicle cyst, ipsilateral renal agenesis and obstruction of the ejaculatory duct [3]. This syndrome owes its name to A. Zinner, who first described it in 1914 [4]. We report here the case of a 32-year-old man presenting with nonspecific genitourinary symptoms, who was subsequently diagnosed as suffering from the rare disease known as Zinner syndrome.

II. CASE PRESENTATION

It is about a 24-year-old unmarried patient with no past medicale history, who consulted our service for desire to have a child associated with chronic pelvic pain. He reported no hemospermia or lower urinary tract symptoms. Examination of the lumbar region was unremarkable, and rectal examination revealed a small, soft prostate with tenderness at the left bladder base. Examination of the scrotum revealed a diminutive right testicle, while the left testicle was normal size on palpation. Blood count was normal, renal function preserved, spermogram showed azoospermia, blood testosterone 4ng, FSH 2, inhibin B 162. CT scans supplemented by MRI (abdominal, pelvic and scrotal) showed 3 abnormalities: Absence of the left kidney in favor of Zinner syndrome - Hemorrhagic cyst of the left seminal vesicle - Right testicular hypotrophy.

Procedure: Ultrasound-guided puncture of seminal vesicle cyst.
Study of puncture fluid:
Bacteriology: sterile culture
Cytologyn: Absence of malignant cells, presence of spermatozoa
Spermogram after puncture: Azoospermia

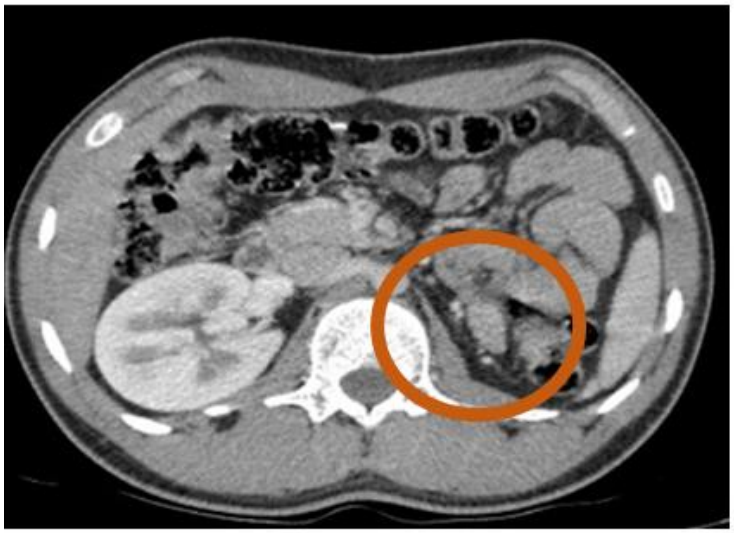

Figure 1: CT scan showed the absent left kidney in the right renal fossa

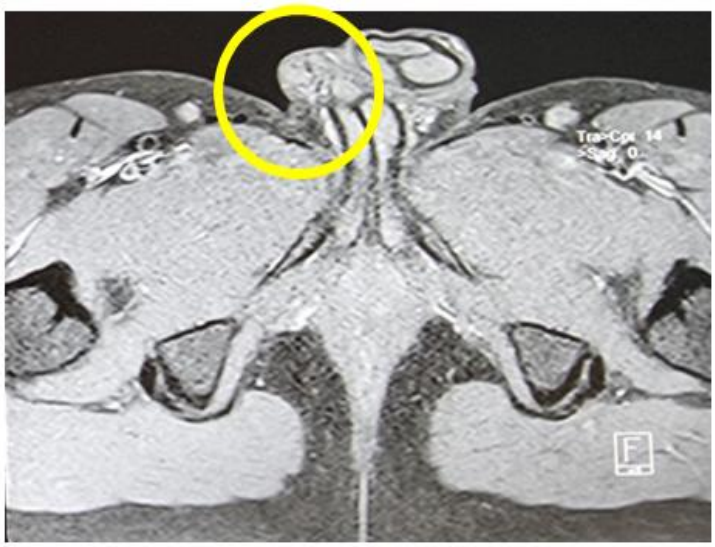

Figure 2: MRI with axial section shows right testicular hypotrophy

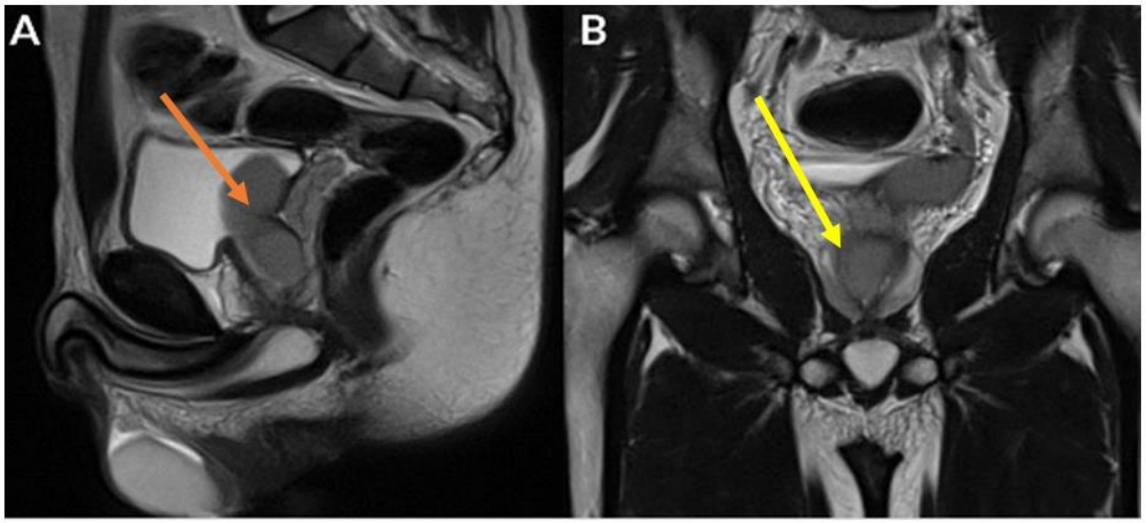

Figure 3: (A) T2 sagittal MRI cut: dilated left ejaculatory duct; (B) T2 sagittal MRI cut: obstruction of the right ejaculatory duct

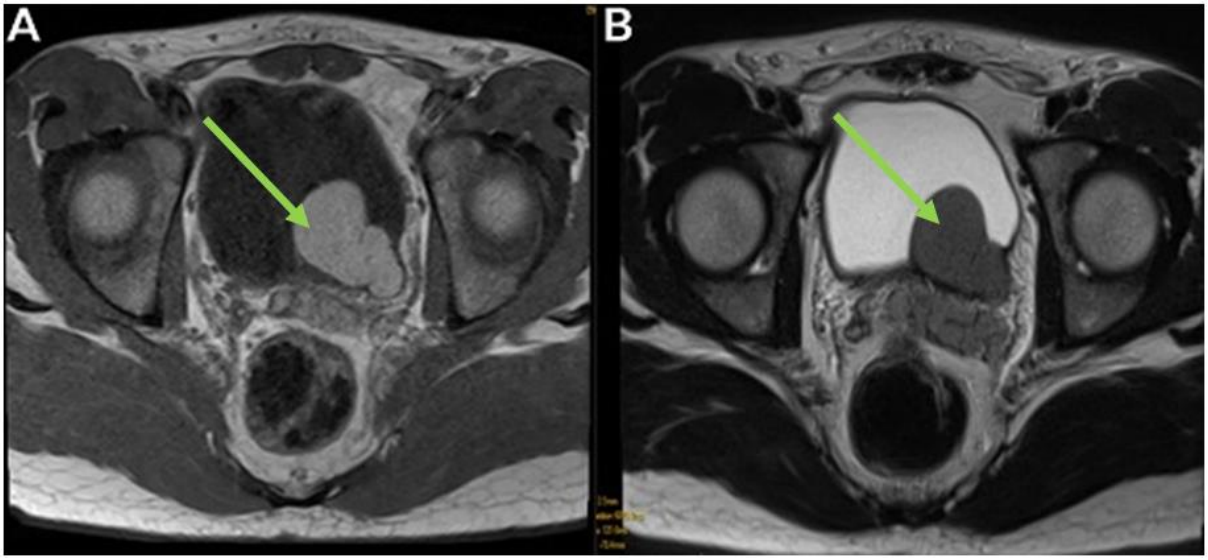

Figure 4: (A) T1 axial MRI cut: cystic formation of the left seminal vesicle which was in hyper signal on T1; (B) T2 axial MRI cut: cystic formation of the seminal vesicle with intermediate signal on T2

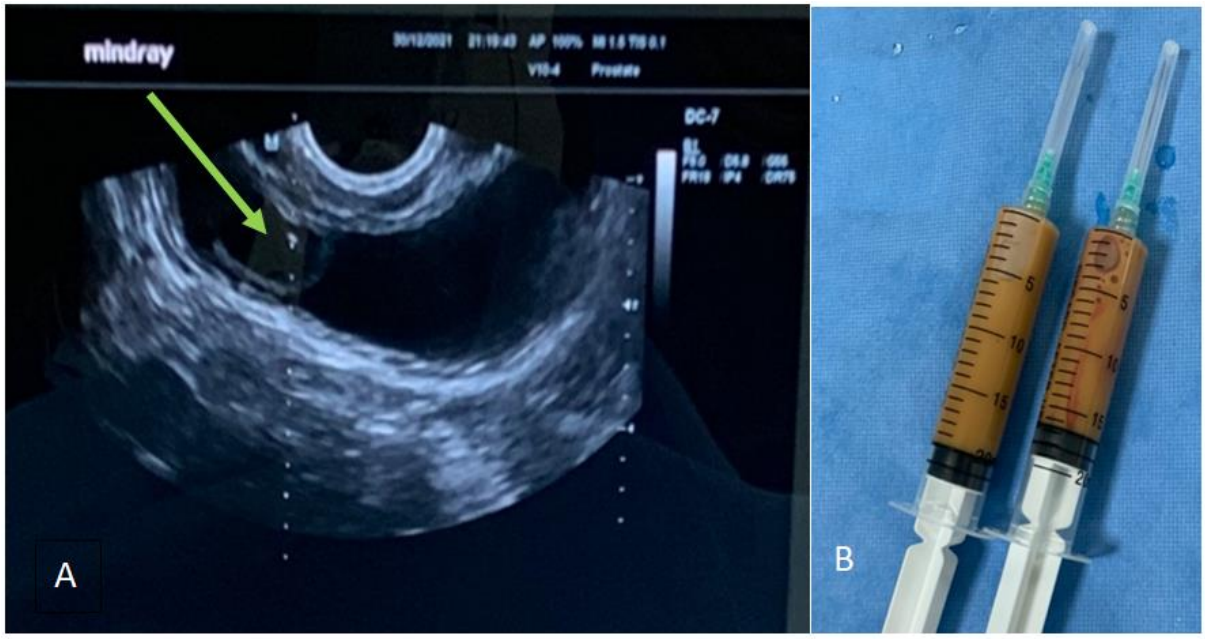

Figure 5: (A) Endorectal ultrasound images of the bladder, showing a cystic lesion protruding into the vesical lumen (red arrow) on the right side with non-visualization of the right seminal vesicle; (B) Buff-yellow appearance of the puncture fluid

III. DISCUSSION

Zinner syndrome is a Wolffian duct anomaly characterized by unilateral renal hypoplasia, homolateral seminal vesicle cyst and ejaculatory duct obstruction [5].

An unusual finding in our patient was that the obstructed ejaculatory duct and the hypoplastic testis were located on opposite sides. In Zinner syndrome, malformations are usually on the same side, although contralateral malformations have been described in the literature [6].

This combination of urinary and genital anomalies is caused by their common embryonic origin in the mesonephric canal. Ureteral buds emerge from the distal mesonephric duct as diverticula and develop cranially to contact the base of the metanephric blastema and induce its differentiation into the final adult kidney. Intrauterine transformation in early pregnancy can lead to abnormal development of the distal mesonephric duct [7].

In our patient, the main reason for consultation was infertility with chronic pelvic pain. This is consistent with the literature. Patients diagnosed with Zinner syndrome present symptoms such as dysuria, urgency, painful ejaculation and epididymitis [8]. But may be asymptomatic, or patients may have non-specific

perineal pain or infertility. Seminal cysts smaller than 5 cm are usually asymptomatic, but large cysts can cause bladder and colon obstruction [3]. Cysts larger than 12 cm are considered giant cysts [9]. Semen analysis of patients with Zinner syndrome has shown decreased sperm volume, oligospermia and azoospermia due to obstruction of the ejaculatory ducts, resulting in infertility in around 45% of patients [4]. Small homolateral testicles and ureterocele can also be reported [10], as can a small contralateral testicle on examination. Rectal examination may reveal well-circumscribed and palpable cystic lesions adjacent to the seminal vesicles, but this may go unnoticed on physical examination [11].

Imaging has an important role to offer the diagnosis of this syndrome. Transrectal ultrasound is the most widely used tool for evaluating seminal vesicles, and can reveal anechoic pelvic cystic lesions. Computed tomography is considered superior to ultrasound. MRI not only provides a definitive diagnosis of the cystic component, resulting in lesions with low signal intensity on T1-weighted images and high signal intensity on T2-weighted images; it is also of great help in decision-making and surgical approach. Due to the mass effect of large seminal vesicle cysts, cystoscopy may reveal an incomplete trigone with extrinsic compression [2, 12].

Treatment consists of monitoring asymptomatic patients, preferably by transrectal ultrasound. Surgical aspiration of seminal vesicle cysts perineally or laparoscopically, or percutaneous cyst drainage or transurethral cyst derotation in symptomatic cases to relieve ejaculatory duct obstruction [7, 8]. Another option is transvesical, extravesical or laparoscopic resection of the seminal vesicle cyst.

IV. CONCLUSION

Zinner syndrome is a rare congenital anomaly that is often asymptomatic. Diagnosis consisted of a clinical examination and a series of imaging studies including suprapubic ultrasound, CT scan and MRI.

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