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Zinner Syndrome: An Overlooked Congenital Anomaly in Male Reproductive Health - A Case Report

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Abstract

Zinner Syndrome, a rare congenital anomaly, involves unilateral renal agenesis, ipsilateral seminal vesicle cysts, and ejaculatory duct obstruction due to mesonephric duct maldevelopment. With approximately 200 reported cases, it typically presents in young adulthood with pelvic pain, urinary symptoms, or infertility.

A 22-year-old male presented with vague abdominal discomfort and occasional lower abdominal pain. Ultrasound revealed left renal agenesis, compensatory right kidney hypertrophy, mild hepatomegaly with fatty changes, and a midline prostatic cyst, initially considered a prostatic utricle cyst. MRI confirmed a bi-loculated left seminal vesicle cyst, dilated left ejaculatory duct and vas deferens, and absent left kidney, establishing Zinner Syndrome.

Resulting from embryologic mesonephric duct anomalies, Zinner Syndrome is diagnosed via ultrasound and MRI, with MRI offering superior anatomical detail. Management varies from conservative observation for mild symptoms to surgical intervention for pain or infertility. This case underscores the need to consider Zinner Syndrome in young males with renal agenesis and pelvic cysts, even with atypical symptoms like abdominal discomfort.

Early imaging-based diagnosis of Zinner Syndrome is critical to guide management and prevent complications such as infertility. This case highlights the importance of clinician awareness to include this rare condition in the differential diagnosis for young males with relevant imaging findings.

Keywords: Zinner syndrome; seminal vesicle cyst; renal agenesis; ejaculatory duct obstruction; male infertility.

Introduction

Zinner syndrome is a rare congenital disorder of the Wolffian (mesonephric) duct characterized by the classic triad of unilateral renal agenesis, ipsilateral seminal vesicle cysts, and ejaculatory duct obstruction[1]. First described by Austrian urologist Adolf Zinner in 1914, fewer than 200 cases have been reported in the literature [1, 2].

This anomaly arises from abnormal embryonic development of the distal mesonephric duct and ureteric bud early in gestation. Specifically, failure of the ureteric bud to induce the metanephros yields ipsilateral renal agenesis or hypoplasia, while concomitant

maldevelopment or atresia of the distal Wolffian duct leads to ejaculatory duct obstruction and cystic dilatation of the seminal vesicle [2,3]. Because the affected kidney and ejaculatory structures derive from a common embryologic origin, an insult before the 7th to 8th week of embryogenesis typically produces the Zinner phenotype[3]. Clinically, many patients are asymptomatic until young adulthood when they may present with pelvic pain, dysuria, infertility or genitourinary infection[4]. Radiological imaging plays a pivotal role in diagnosis, with ultrasound often first identifying renal agenesis and cystic pelvic lesions, and MRI providing detailed characterization of seminal vesicle cysts and associated anomalies

Here we present a case of 22-year-old male presenting with vague complains of lower abdominal discomfort.

Case Report

A 22-year-old male was referred for evaluation of pelvic discomfort and urinary hesitancy. A transabdominal ultrasound (USG) of the abdomen and pelvis demonstrated absence of the left kidney, with an empty left renal fossa and no renal tissue visualized on the left side. The right kidney was markedly enlarged consistent with compensatory hypertrophy. Adjacent to the posterior bladder wall, ultrasound revealed a well-defined, anechoic cystic mass, thought to arise from the left seminal vesicle. The left seminal vesicle was not separately visualized except as part of this cystic lesion. The proximal vas deferens on the left was tubular and dilated, and the distal left ejaculatory duct was not visualized, suggesting obstruction. No ureteric jets were seen on the left; the right ureter was normal in caliber. The bladder appeared normal.

To further characterize these findings, an MRI of the pelvis was performed. On MRI, a bi-loculated T2-hyperintense cystic lesion was seen in the left retrovesical space, contiguous with the expected location of the left seminal vesicle. The cyst contained fluid with low T1 signal intensity, consistent with simple fluid. The left ejaculatory duct was dilated proximally and abruptly terminated into the cystic mass. The prostate gland appeared normal but contained a small midline cystic structure at the prostatic urethra adjacent to the verumontanum, consistent with a prostatic utricle cyst. MRI confirmed left renal agenesis and showed compensatory hypertrophy of the right kidney. There was no evidence of an ectopic or pelvic kidney, a remnant of a left ureteric bud was not identified. No enhancing solid components or diffusion restriction were noted within the seminal vesicle cyst.

These imaging features (left renal agenesis, ipsilateral seminal vesicle cyst with ejaculatory duct obstruction, and dilated vas deferens) established the diagnosis of Zinner syndrome.

Figure 1:

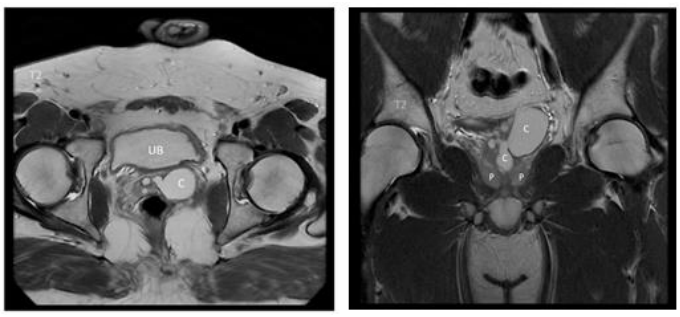
Ultrasound showing –

A) Bulky right kidney measuring ~14.1 x 6.8 cm

B) Empty left renal fossa

C) Ultrasound of pelvis showing anechoic cystic structure in midline slightly left side.

Figure 2:



Mri images showing:

A) axial T2

B) coronal T2 large well defined thin walled hyperintense biloculated cystic lesionin the left seminal vesicle

Discussion

Zinner syndrome results from embryologic malformations of the mesonephric (Wolffian) duct system. During normal development, the ureteric bud branches from the mesonephric duct and induces formation of the kidney, while the distal duct itself forms male reproductive structures (seminal vesicles, vas deferens, ejaculatory ducts, epididymis, part of the trigone) under testosterone and MIS influence [2,3]. Interruption of the ureteric bud–metanephros interaction leads to unilateral renal agenesis or dysplasia, while concomitant failure of the distal mesonephric duct to canalize produces ejaculatory duct atresia and seminal vesicle cysts[4]. This shared embryology explains the near-constant association of ipsilateral seminal vesicle cysts with renal agenesis. In fact, ipsilateral renal anomalies occur in up to 67–75% of congenital seminal vesicle cyst cases[4]. Prostatic utricle cysts, ectopic ureters, and vas deferens anomalies are reported less commonly [1].

Patients with Zinner syndrome are often asymptomatic until the second to fourth decades of life. When present, symptoms may include pelvic or perineal pain, dysuria, ejaculatory pain, hematospermia, recurrent epididymitis or infertility[5]. Infertility can result from ejaculatory duct obstruction and severely decreased semen volume, so fertility evaluation is important in these patients [2,5]

The patient in this report was 22 and had only mild urinary symptoms. Imaging is essential for diagnosis. Ultrasound (transabdominal and/or transrectal) is typically the first-line test. Transabdominal ultrasound may reveal an empty renal fossa and compensate contralateral hypertrophy, as in our case [6]. Transrectal ultrasound (TRUS) can delineate the seminal vesicles and ejaculatory ducts in detail, demonstrating cyst wall characteristics, internal septations, or debris. TRUS may also detect a dilated ipsilateral vas deferens or ejaculatory duct obstruction [2,5].

MRI is the modality of choice for definitive evaluation. MRI confirms the location, size, and contents of the seminal vesicle cyst and accurately characterizes associated structures. Seminal vesicle cysts are typically hyperintense on T2-weighted images and hypointense on T1 if simple fluid, although proteinaceous or hemorrhagic contents can alter the signal. Additional findings on MRI such as contralateral seminal vesicle hypoplasia or vas anomalies should be reported.

The differential diagnosis for a retrovesical cystic mass is broad. Paramesonephric (Müllerian) cysts, ejaculatory duct diverticula, prostatic utricle cysts, prostatic abscesses, or Gartner duct cysts (in females) may appear similar on imaging [2]. Key distinguishing features are laterality and association with renal anomalies[6]. Müllerian duct cysts are midline and not typically associated with renal agenesis. A prostatic utricle cyst is also midline and connects to the prostatic urethra. In our case, the cyst was clearly lateralized to the left seminal vesicle and there was absent left kidney, strongly favoring Zinner syndrome.

Clinical correlation (male patient, symptoms) and demonstration of ejaculatory duct obstruction complete the picture. Treatment of Zinner syndrome depends on symptoms and fertility considerations. Asymptomatic or mildly symptomatic patients may be managed conservatively

In symptomatic cases (e.g. recurrent infection, pain, infertility), surgical intervention is indicated. Options include transurethral resection of the ejaculatory duct

(TURED) to relieve obstruction, or excision of the seminal vesicle cyst. Minimally invasive approaches (laparoscopic or robotic vesiculectomy) have become standard due to deep pelvic location and complexity [1, 3].

Larger cysts or those with stones may require open or robotic excision. In our patient's case, symptoms were minimal and he deferred children, so we opted for observation. Should he develop more severe symptoms or desire fertility, options like TURED or cyst excision could be offered. Sperm banking may be considered preoperatively in young patients undergoing intervention [4].

Throughout management, imaging remains crucial. Ultrasound guides initial diagnosis and can monitor cyst size. MRI helps plan the surgical approach and assess adjacent anatomy. After treatment, imaging (e.g. MRI or ultrasound) is useful to detect recurrence or complications. Awareness of Zinner syndrome and its radiologic features is essential for radiologists, urologists, and surgeons, as early recognition can prevent misdiagnosis (e.g. as an abscess or neoplasm) and improve outcomes.

Conclusion

Zinner syndrome is a rare congenital Wolffian duct anomaly that presents a diagnostic challenge. The combination of unilateral renal agenesis, ipsilateral seminal vesicle cyst, and ejaculatory duct obstruction (along with possible associated findings such as vas deferens dilation or prostatic utricle cyst) defines the condition. Modern imaging allows prompt and accurate diagnosis of this entity. In this 22-year-old man, ultrasound and MRI clearly demonstrated left renal agenesis, a large left seminal vesicle cyst with ejaculatory duct dilatation, compensatory right kidney hypertrophy, and an associated prostatic utricle cyst. Recognition of this constellation on imaging avoided unnecessary biopsy or surgery. Management is individualized: asymptomatic patients may be observed, whereas symptomatic cases require intervention. Early identification is important to counsel regarding fertility and to treat complications. Radiologists should include Zinner syndrome in the differential for any young male with a unilateral renal anomaly and pelvic cystic masses.

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