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ZINNER SYNDROME: A RARE CONGENITAL UROGENITAL ANOMALY

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A male child was referred to a Pediatric Nephrology consultation due to left kidney agenesis, which was initially suspected during the fetal period and later confirmed by an ultrasound and a Tc-99m dimercaptosuccinic acid renal scan performed at 1 month of age. The scan revealed a single functioning right kidney with normal morphology and function. Regular follow-up was maintained and in the absence of urinary symptoms, hypertension or microalbuminuria, emphasis was placed on promoting healthy lifestyle habits. At age 16, a follow-up ultrasound identified asymmetry in the seminal vesicles, indicating congenital agenesis of the left seminal vesicle. Suspecting Zinner syndrome, an abdominal-pelvic magnetic resonance imaging (MRI) was performed, confirming left renal agenesis (Figure 1), ipsilateral agenesis of the seminal vesicle (Figure 2) along with a periurethral cyst (Figure 3) and a hypotrophic left testicle, thus confirming the diagnosis. The patient reported no abdominal, pelvic or testicular pain and no lower urinary tract symptoms. Clinical, imaging and laboratory follow-up were maintained, with the patient remaining asymptomatic throughout and retaining a glomerular filtration rate within the normal ranges. At 18 years, upon reaching the maximum age for pediatric healthcare, the patient was referred to a Urology consultation for ongoing follow-up and fertility evaluation.

Figure 1. Coronal T2-weighted MRI of the abdomen demonstrating agenesis of the left kidney, with compensatory hypertrophy of the right kidney.

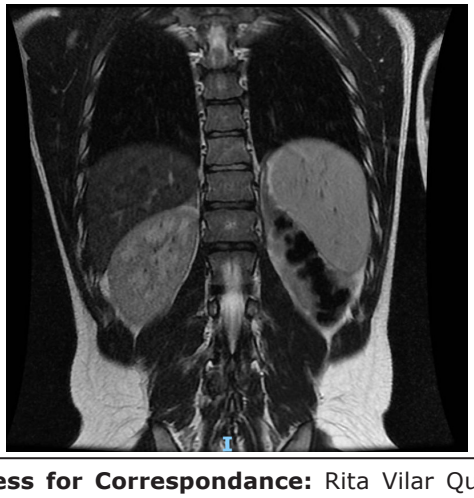

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Figure 2. Axial T2-weighted MRI of the pelvis demonstrating agenesis of the left seminal vesicle.

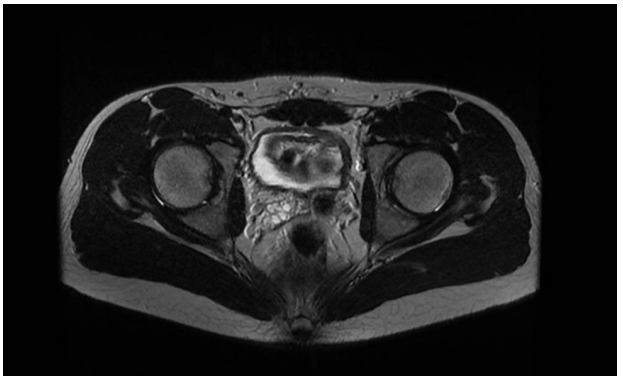
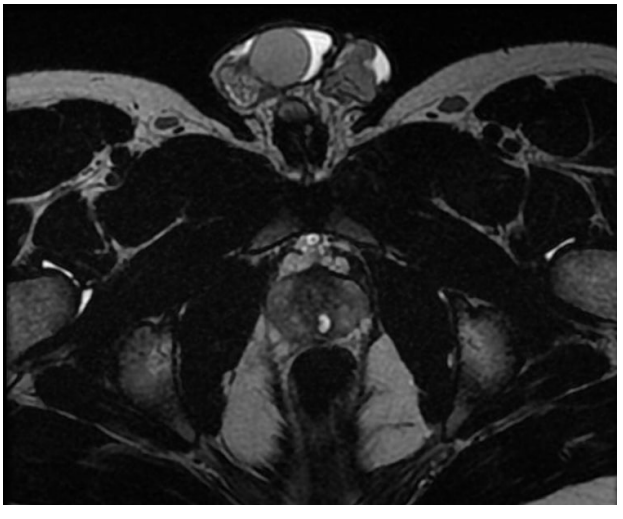


Figure 3. Axial T2-weighted MRI of the pelvis demonstrating a periurethral cyst.



What is Zinner Syndrome?

Zinner syndrome (ZS) is a rare congenital anomaly of the urogenital tract, with an estimated incidence of 0.00214%,[1,2,3] arising from abnormal embryonic development of the distal mesonephric duct,[4] occurring between the 4th and 13th weeks of gestation.[1,2,3,5] It is characterized by a triad of clinical manifestations: seminal vesicle cyst (SVC), ejaculatory duct obstruction and ipsilateral renal malformation, commonly presenting as kidney agenesis or multicystic dysplasic kidney (MCDK).[1,2,3,4,5] ZS is predominantly asymptomatic, with symptoms usually emerging after the onset of intense sexual activity[3,5] due to seminal fluid accumulation caused by ejaculatory duct stenosis.[1,2] Common symptoms

are generally non-specific and include abdominal, pelvic and scrotal pain along with lower urinary tract symptoms.[1,2,3,4,5] Painful ejaculation and recurrent epididymitis have also been reported.[4] Infertility is a concern in ZS, affecting approximately 45% of patients[1,2], primarily due to azoospermia.[1] Although the underlying pathogenesis remains unclear[1], it is speculated that unilateral testicular obstruction may lead to the production of anti-sperm antibodies.[1,2] Due to its asymptomatic nature and rarity, the diagnosis of ZS during the prepubertal period is mostly incidental, typically occurring during routine ultrasounds conducted to investigate abnormalities of kidney and urinary tract.[3] MRI serves as the confirmatory diagnostic tool[1,3,4,5], offering superior resolution without the risks associated with radiation exposure.[3,5] Management of ZS generally involves a conservative approach with watchful waiting for asymptomatic or minimally symptomatic patients.[1,2,3] Surgical intervention is reserved for individuals exhibiting significant symptoms or those who have not responded to conservative treatment.[1,2,3] Annual follow-up with clinical and imagological evaluations is recommended for all patients.[1,2] Upon reaching adulthood, a semen analysis should be conducted to assess fertility status.[1,4] With the increasing use of ultrasound, there has been a rise in the detection of renal agenesis, MCDK and urinary tract malformations.[3] Although these anomalies are common, they should prompt clinicians to consider associated genitourinary anomalies, such as SVC in male patients and, consequently, ZS,[2,3] since early detection and appropriate management are crucial in preventing its complications.[4] While further studies are necessary to substantiate the benefits of early surgical intervention, timely excision of the affected duct may help prevent infertility and reduce the impact on the contralateral genitourinary tract.[1]

Compliance with Ethical Standards

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Conflict of Interest: None

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