

CASE REPORT

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A rare case of Zinner syndrome with unilateral testicular agenesis and aberrant course of left common iliac artery

Dixit Varma^{1*} , Ibrahim Khalil Ullah¹ and Madhurjya Kumar Thakur¹

Abstract

Background Zinner syndrome is a rare Wolffian duct anomaly presenting as a triad of unilateral renal agenesis, ipsilateral seminal vesicle cyst and ipsilateral ejaculatory duct obstruction. The condition is due to a developmental anomaly leading to malformation of the mesonephric duct. Magnetic resonance imaging is considered to be the gold standard for the diagnosis of Zinner syndrome. Due to the limited number of cases reported in the literature, the associations of Zinner syndrome are not well established.

Case presentation In this case report, we are presenting a case of a 38-year-old male with dysuria, intermittent pain in the scrotum and lower abdomen and painful ejaculation for the past six months. Imaging findings led to the diagnosis of Zinner syndrome, with unilateral testicular agenesis and an aberrant course of the left common iliac artery.

Conclusions Apart from the known associations of Zinner syndrome, testicular agenesis in a patient with Zinner syndrome has not been reported in the literature and is an important addition to the limited literature available.

Keywords Zinner syndrome, Seminal vesicle cyst, Renal agenesis, Testicular agenesis, Case report

Background

Zinner syndrome is a rare congenital malformation of the urogenital tract. The syndrome is due to maldevelopment of the distal part of the Mullerian duct during early embryogenesis. It is characterised by a triad of unilateral renal agenesis, ipsilateral seminal vesicle cyst and ipsilateral ejaculatory duct obstruction. It was first described in 1914 by A. Zinner [1]. A total of 6 cases of seminal vesicle cysts with ipsilateral renal agenesis or dysplasia were diagnosed during renal ultrasound screening of 280,000 children in a study in China [2]. Magnetic resonance imaging (MRI) is the gold standard of diagnosis. Due to the limited number of cases reported in the literature, the

associations of the syndrome are not fully established. Our case is the first in the literature on Zinner syndrome with ipsilateral testicular agenesis and an aberrant course of the left common iliac artery.

Case presentation

A 38-year-old male presented with dysuria, intermittent pain in the scrotum and lower abdomen and painful ejaculation for the past six months. There was no relevant significant past medical history or intervention. Palpation revealed an absent testis in the left scrotal sac. The rest of the general examination and vital signs were all within normal limits. Palpation of the abdomen revealed slight pain in the pelvic region. Trans-abdominal ultrasonography (USG) revealed non-visualisation of the left kidney in the left renal fossa or elsewhere in the abdominal cavity, which implicated the diagnosis of left renal agenesis. The right kidney was mildly enlarged, suggesting compensatory hypertrophy. The left testis was absent in the patient. In the left

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peri-prostatic region, a well-defined rounded anechoic lesion was noted measuring about 69×70×84 mm in its maximum anteroposterior, transverse and cranio-caudal dimensions, respectively (Fig. 1). On the left side, the left testis was not sonographically visualised in the scrotal sac. The patient underwent a contrast-enhanced computed tomography (CECT) study of the abdomen, which confirmed the absence of the left kidney, and the left ureter (Fig. 2). The left renal artery and vein were also absent. A low-density cystic lesion was noted in the left peri-prostatic region that did not show any contrast uptake (Fig. 3). The left common iliac artery had an aberrant course, winding around the seminal vesicle cyst (Fig. 4). The rest of the findings were consistent with the USG. The rest of the solid abdominal organs and bowel loops appeared unremarkable. For a more thorough examination, an MRI of the pelvis and the inguinoscrotal area was conducted. MRI revealed a cystic lesion in the left seminal vesicle, demonstrating T2-hyperintense signal and T1-hypointense signal with dilatation of the left ejaculatory duct (Fig. 5). Thus, inclusive findings of USG, CECT abdomen and MRI of left renal agenesis with ipsilateral seminal vesicle cyst and dilated left ejaculatory duct led to the diagnosis of Zinner syndrome. The patient was advised to undergo laparoscopic excision of the cyst. However, the patient opted for conservative management with transrectal aspiration of the cyst and is currently undergoing follow-up with relief of symptoms.

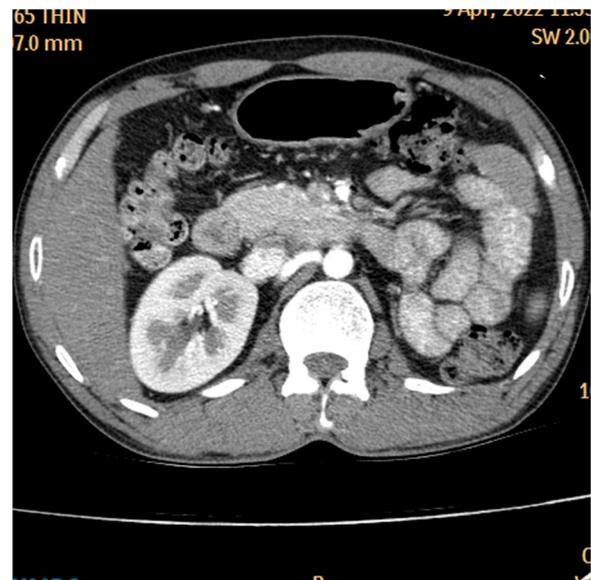


Fig. 2 Axial contrast-enhanced CT study of the abdomen in the arterial phase reveals the absence of the left kidney and the left renal artery. The right kidney and the right renal artery are visualised

Discussion

Zinner syndrome is a rare birth defect affecting the urogenital system and is characterised by a combination of three conditions: the absence of one kidney, cyst in the ipsilateral seminal vesicle and blockage in the corresponding ejaculatory duct. The affected ureter may be

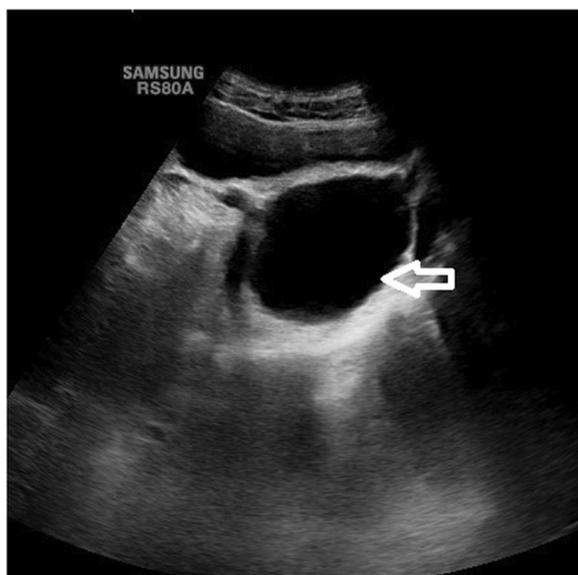


Fig. 1 Sagittal section of transabdominal USG of the pelvic region shows a well-defined anechoic cystic lesion (arrow) in the left peri-prostatic region



Fig. 3 Axial contrast-enhanced CT study of the abdominopelvic region in the arterial phase reveals a low-density cystic lesion (arrow) in the pelvic region that does not show any contrast uptake

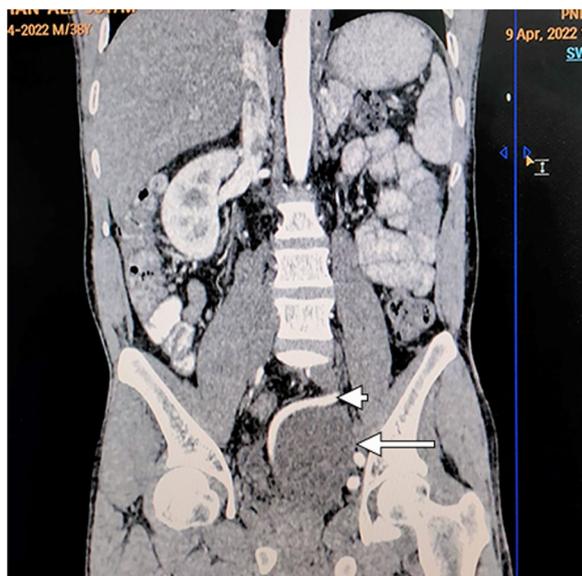


Fig. 4 Coronal contrast-enhanced CT study of the abdominopelvic region in the arterial phase reveals the absence of the left kidney in the left renal fossa and a low-density cystic lesion in the pelvic region slightly off-centred to the left side. The cystic lesion does not show any contrast uptake (long arrow). An aberrant left common iliac artery is also noted (short arrow) as it wraps the medial aspect of the cystic lesion. (Additional file 1: Movie 1)



Fig. 5 T2-weighted coronal section of the pelvic region reveals a hyperintense cystic lesion (long arrow) in relation to the left seminal vesicle, compatible with a seminal vesicle cyst. The ipsilateral ejaculatory duct is also dilated (short arrow), suggestive of ejaculatory duct obstruction

incomplete, absent or follow an abnormal course. The medical literature has reported fewer than 200 cases of this syndrome up to 2018 [3]. The condition can be present without symptoms or may manifest between the

second and fourth decade of life [4]. When symptomatic, painful ejaculation, scrotal pain, perineal pain, abdominal pain, urinary urgency, urinary frequency, haemospermia, epididymitis, testis swelling, dysuria or infertility are the usual clinical findings. In our case, the cause of pain is likely caused by the pressure effects of the cyst on adjacent structures and the obstruction of the left ejaculatory duct.

The mesonephric ducts are embryonic tubes that originate from the primitive kidney and connect to the cloaca. From these ducts, the ureteric buds grow out. The primitive kidney is formed when the ureteric bud migrates and fuses with the metanephric blastema under the influence of growth hormones. In males, the mesonephric ducts develop into various structures, including the trigone of the urinary bladder, urethra, seminal vesicle, vas deferens, epididymis and other parts of the male reproductive system [5]. During foetal development, the ureteric bud should separate from the mesonephric duct, but in Zinner syndrome, this separation does not occur, leading to blockages in the seminal vesicle and ejaculatory duct, resulting in cyst formation. The lack of separation also causes renal agenesis. This condition is believed to be the male equivalent of Mayer–Rokitansky–Kuster–Hauser syndrome.

Zinner syndrome is suspected when a male patient presents with recurrent urinary tract infections and unilateral renal agenesis. Imaging is crucial for making a diagnosis, and ultrasound is commonly used. Ultrasound reveals an anechoic lesion in the region of the seminal vesicle and the absence of the kidney, but it cannot accurately distinguish seminal vesicle cysts from other cystic lesions. Computed tomography (CT) urogram confirms the absence of the kidney and ureters and reveals a cystic lesion in the retrovesical peri-prostatic region. MRI is the gold standard for diagnosis, as it can confirm the seminal vesicle cyst and obstruction of the ejaculatory ducts. Fine-needle aspiration of the cyst may reveal spermatozoa, which can differentiate it from other cysts. The differential diagnosis includes prostatic cysts, prostatic utricle cysts, diverticulosis of the vas deferens, ectopic ureterocele, cystic neoplasms and pelvic abscesses. The position, intralésional material, related urogenital system abnormalities and imaging features are used to differentiate between the entities. Mullerian duct cysts and ejaculatory duct cysts are located in the midline, and seminal vesicle cysts are typically hyperintense on T2-weighted imaging and hypointense on T1-weighted imaging. Diverticulosis of the vas deferens ampulla and ectopic ureterocele is found laterally [6].

Because the number of reported cases is limited, there is a lack of research on the associations of Zinner syndrome. Cases were reported in which there was obstruction of the ejaculatory duct, a contralateral hypoplastic testicle, a small-sized testis, an ipsilateral ureterocele and

other conditions like ectopic ureter and megaureter [7–9]. Some studies reported unique combinations of abnormalities, including cystic dysplasia of the rete testis, partial hemitrigonal development and epididymal dilatation [10]. Two cases of Zinner syndrome reported by Cascini et al. had prenatally been diagnosed with multicystic dysplastic kidneys [11]. Fiaschetti et al. reported a patient with situs inversus, renal agenesis, ipsilateral seminal vesicle cyst, ectopic ureter and contralateral ejaculatory duct obstruction [12]. A systematic review found several comorbidities associated with Zinner syndrome, such as adenocarcinoma of the seminal vesicle, paraganglioma, squamous cell carcinoma of the seminal vesicle, renal cell carcinoma and non-Hodgkin lymphoma [13]. Treatment options depend on the size and clinical features of the cyst and may be conservative or surgical. Conservative management includes antibiotics and transurethral aspiration of the cyst or instillation of alcohol and minocycline, while excision of the cyst is preferred due to the high likelihood of recurrence following cyst puncture [14].

Conclusions

Zinner syndrome, although a rare disorder, is seen in adult males who report nonspecific symptoms such as dysuria, infertility, perineal pain and painful ejaculation. This triad of unilateral renal agenesis, ipsilateral seminal vesicle cyst and ejaculatory duct obstruction is best diagnosed with radiological imaging. Though ultrasonography and CT scans can be useful in suggesting this diagnosis, MRI is the preferred modality that confirms the diagnosis. The known associations of Zinner syndrome include conditions like hypoplastic testicles, ectopic ureter, megaureter and ureterocele. Testicular agenesis in a patient with Zinner syndrome has not been reported in the literature and is an important addition to the limited literature available on Zinner syndrome.

Abbreviations

CECT	Contrast-enhanced computed tomography
CT	Computed tomography
MRI	Magnetic resonance imaging
USG	Ultrasonography

Supplementary Information

The online version contains supplementary material available at <https://doi.org/10.1186/s43055-023-01045-6>.

Additional file 1: Movie 1: Scrolling sections of axial contrast-enhanced CT study of the abdominopelvic region in the arterial phase follows the aberrant course of the left common iliac artery, which is displaced medially and winds around the left seminal vesicle cyst.

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Author contributions

DV is the corresponding author and contributed to data collection, diagnosis and MRI findings. IKU and MKT were involved in manuscript writing. All authors read and approved the final manuscript.

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Availability of data and materials

Data sharing is not applicable to this article as no datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate

Written informed consent was obtained from the patient's family for publication of this case report and accompanying images, and the study is approved by the ethics committee at Fakhruddin Ali Ahmed Medical College and Hospital.

Consent for publication

Case publication was approved; consent form was signed by the patient's family.

Competing interests

The authors declare that they have no competing interests.

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