

UNILATERAL EJACULATORY DUCT OBSTRUCTION: A CASE STUDY OF ZINNER'S SYNDROME

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Abstract: Zinner's syndrome is a rare congenital mesonephric (Wolffian) duct anomaly due to developmental arrest in early embryogenesis, comprising unilateral ejaculatory duct obstruction or agenesis, ipsilateral seminal vesicle cysts, and ipsilateral renal agenesis. Here, we present a case of a healthy 17-year-old male in Pakistan presenting with complaints of mild, dull pain in the left lower abdomen. He had no history of lower urinary tract symptoms and regular bowel habits. Imaging revealed non-visualization of the left kidney with multi-cystic pelvic mass related to the left seminal vesicle in keeping with the diagnosis of Zinner's syndrome.

Keywords: Zinner's Syndrome, Congenital Anomaly, Unilateral Ejaculatory Duct Obstruction, Renal Agenesis

Introduction

Zinner's syndrome is a rare congenital malformation of the genitourinary tract. Because of first trimester insult, the embryogenesis of the seminal vesicle, vas deferens, kidney, and ureter could be altered. Due to the common embryological origin of seminal vesicle and ureter from the mesonephric duct, there is a co-occurrence of seminal vesicle cyst and ipsilateral renal agenesis (Florim et al., 2018). It is considered the Mayer-Rokitansky-Kuster-Hauser counterpart in males (Khanduri et al., 2017).

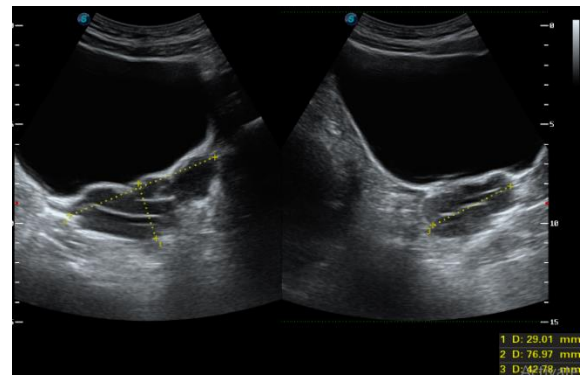
Case Presentation

A 17-year-old male presents with complaints of mild, dull, non-transferred pain in the lower abdomen on the left side. He did not have any associated lower urinary tract symptoms. The rest of the physical examination was unremarkable.

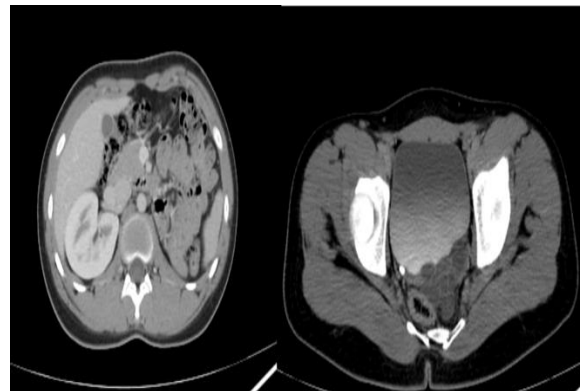
Abdominopelvic ultrasound (USG) revealed non-visualization of the left kidney and a complex multi-cystic lesion with echogenic content observed at the left posterolateral aspect of the urinary bladder measuring 76 x 29 mm [Fig. 1].

Multi sequential Multi-planner plain magnetic resonance imaging (MRI) pelvis revealed a multi-cystic mass high on T1W and T2W not suppressed on Fat saturation suggestive of hemorrhagic/proteinaceous content and dilated tortuous left-sided ureter inserted ectopically in seminal vesicle cyst. [Fig. 3a, 3b, 3c & 3d].

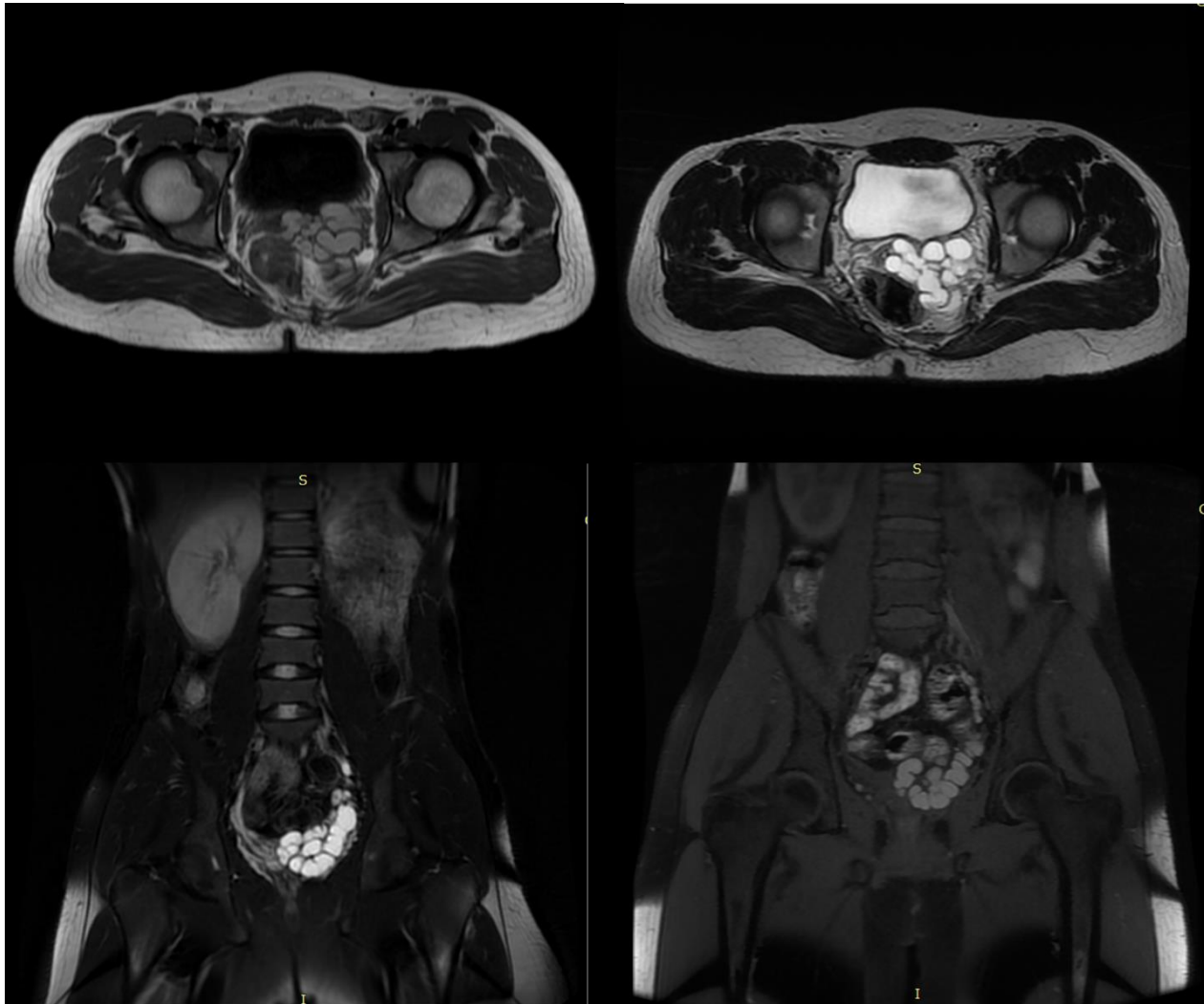
Abdominopelvic computed tomography (CT) axial slices revealed left renal agenesis associated with a large multilobulated non-enhancing cystic mass [Fig. 2a & 2b].



[Fig.1] Abdominopelvic USG showing multilobulated cystic lesion with echogenic content at the left posterolateral aspect of the urinary bladder.



[Fig. 2-a & b] CT images axial slices (a) showing non-visualization of left kidney. (b) Shows a large multilobulated non-enhancing cystic mass involving the left seminal vesicle.



[Fig. 3] (a & b) MRI T1W and T2W axial sections showing multilobulated hyperintense seminal vesicle lesions, respectively. (c) MRI T2W coronal section showing non-visualization of left kidney with left seminal vesicle multi-cystic lesion. (d) MRI T1W Coronal Fat Sat. image showing dilated tortuous distal ureter inserting ectopically in seminal vesicle cyst.

Discussion

Zinner's Syndrome is a rare congenital anomaly comprising seminal vesicle cyst, ipsilateral renal agenesis, and ejaculatory duct obstruction. The Ureter of this side can be atrophic or have an abnormal course, inserting ectopically into the seminal vesicle cyst as seen in our case (Ibrahimi et al., 2020). It was first described in 1914 by Zinner (Zinner, 1914). Seminal vesicle cysts can be congenital or acquired secondary to chronic inflammation (Kuo et al., 2011).

Only 200 cases in the literature have been reported (Florim et al., 2018; Ibrahimi et al., 2020). It may also be associated with other anomalies, such as ipsilateral testicular agenesis, polycystic kidney disorders, and hemivertebrae (Khanduri et al., 2017). Other associated anomalies, such as testicular ectopia, absent ureter, and trigone, may also be seen (Kuo et al., 2011). The frequency of ipsilateral renal abnormalities is higher, while seminal vesicle cysts are documented in 5% of the patients with renal agenesis. The incidence of seminal vesicle cysts is 0.005%, and almost two-thirds of the cases are related to ipsilateral renal agenesis (Khanduri et al., 2017). Most of the patients with cysts less than 5 cm in size are asymptomatic before 2 to 3rd

decade of life with a mean age of 30 years at presentation, which corresponds to the beginning of sexual life, and there is also an increase in cyst size. Symptoms are usually related either to the urinary tract, such as dysuria, hematuria, urgency, and frequency, or sometimes related to the reproductive system, such as painful ejaculation, hematospermia, or subfertility. Complications, which include epididymitis and prostatitis, are also seen (Ibrahimi et al., 2020; Khanduri et al., 2017; Van den Ouden et al., 1998).

Ultrasound, CT, MRI, and vaso-vesiculography help establish an accurate diagnosis. Ultrasound is a baseline modality that reveals the absence of one kidney and cystic structure in the ipsilateral hemipelvis. CT reveals better tissue details, such as confirmation of renal agenesis and the origin of the cystic lesion from the seminal vesicle. MRI is the modality of choice for diagnosis and planning surgery; MRI signals are specific for cyst components (hemorrhagic/proteinaceous) and precisely detect genitourinary abnormalities and anatomical relationships with the pelvic structures (AlArifi et al., 2019; Ibrahimi et al., 2020). Vaso-ventriculography can also be performed, which entails aspiration of cyst fluid followed by injection

of contrast; however, it is rarely performed to avoid complications (Kuo et al., 2011).

Differentials include prostatic cysts, prostatic utricle cysts (Müllerian duct cysts), ureteroceles, and abscesses. Diagnosis is primarily made on cyst position and other associated anomalies (Ibrahimi et al., 2020). Prostatic cysts are localized to the prostate at the level of verumontanum, can be midline or lateral, are usually small, and contain no sperms. These are unilocular and hypointense T1-weighted MRI images concerning the prostate (Gevenoio et al., 1990; Kuo et al., 2011; Van den Ouden et al., 1998). Müllerian duct cysts are midline cysts arising behind the verumontanum extending above the base of the urinary bladder with a beak-like projection into its lumen (Kuo et al., 2011; Pollack and McClennan). Ureterocele is a sacular dilatation of the terminal ureter that could be congenital or acquired (inflammation/trauma), usually unilateral, at the site of ureteric insertion. It gives a cobra head appearance on the urogram (Coplen and Duckett, 1995; Kuo et al., 2011). A seminal vesicle abscess appears as a cyst in a seminal vesicle with echogenic material in it; there will be a history of purulent ejaculation (Monzo and JM, 2005). Management includes a multidisciplinary approach, depending upon the presence of symptoms. In our case, as the patient was asymptomatic, conservative management of pain relief was advised with a 3-month clinical follow-up. Transrectal cyst aspiration or simple drainage of the cyst can be performed for symptomatic relief; however, it is associated with increased chances of recurrence. Surgical intervention will be carried out for symptomatic patients; one option is open surgical resection. Minimally invasive laparoscopic removal and robot-assisted surgeries are thought to be the best options due to better outcomes and short hospital stays (AlArifi et al., 2019).

Conclusion

In conclusion, Zinner's syndrome is a rare congenital anomaly that affects the genitourinary tract. The co-occurrence of seminal vesicle cysts, ipsilateral renal agenesis, and ejaculatory duct obstruction characterizes it. Symptoms are usually related to the urinary or reproductive systems and can include dysuria, hematuria, painful ejaculation, and subfertility. Imaging modalities such as ultrasound, CT, and MRI are helpful in establishing an accurate diagnosis. Early diagnosis and management are crucial in preventing complications and improving the patient's quality of life.

Declarations

Data Availability statement

All data generated or analyzed during the study are included in the manuscript.

Ethics approval and consent to participate

Approved by the department Concerned.

Consent for publication

Approved

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Conflict of interest

The authors declared absence of conflict of interest.

Author Contribution

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Conception of Study, Development of Research Methodology Design, Study Design,, Review of manuscript, final approval of manuscript

Coordination of collaborative efforts.

MARYIM NASREEN

Data acquisition, analysis.

Coordination of collaborative efforts.

ASIF HAMEED

Data entry and Data analysis, drafting article

Data acquisition, analysis.

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