

ZINNER SYNDROME- A CASE REPORT AND REVIEW OF LITERATURE

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PRESENTATION OF CASE

A 38 year old male patient presented with complaints of dysuria, increased frequency of micturition, scrotum and perineal pain intermittently, painful ejaculation, recurrent episodes of urinary tract infections and hematuria since 2 years.

RADIOLOGICAL FINDINGS

The patient came for ultrasonography of the abdomen and pelvis and was investigated further with CT scan and MRI of the abdomen and pelvis.

Trans-abdominal ultrasound showed a well-defined rounded anechoic cystic lesion detected in left superolateral aspect of the prostate gland; in left periprostatic region, abutting the inferolateral aspect of the left bladder base/posterior aspect of the fundus of the urinary bladder and non-visualisation of left kidney in the left renal fossa or elsewhere in the abdominal cavity or pelvis suggestive of left renal agenesis. The right kidney also showed compensatory hypertrophy. The testes were normal on both sides.

Computed tomography of the abdomen and pelvis revealed an isodense cyst measuring 4.2 cm in diameter, posterior to the bladder wall and anterior to the left seminal vesicle, continuous with the seminal vesicle on the left side suggestive of a seminal vesicle cyst with ipsilateral renal agenesis.

MRI of abdomen and pelvis was performed which showed left renal agenesis and compensatory hypertrophy of right kidney and a left seminal vesicle cyst displaying hyperintense signal on T1-weighted images and isointense signal on T2-weighted MRI suggestive of proteinaceous/hemorrhagic contents. MRI also showed the continuity between the dilated proximal seminal vesicle and the seminal vesicle cyst on the left side. The vas deferens was dilated all along its course in spermatic cord with significant compression of the left ejaculatory duct and distal part of vas deferens. The seminal vesicle was normal on right side

with no definite dilatation of vas deferens or ejaculatory duct on right side. The testes were normal on both sides.

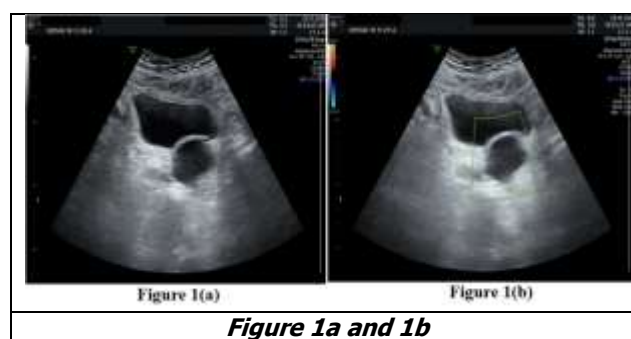


Figure 1(a). Transabdominal ultrasound shows a well defined rounded anechoic cystic lesion with posterior acoustic enhancement abutting the inferolateral aspect of the bladder on the left side, superolateral aspect of the prostate in the left periprostatic region.

Figure 1(b). On transabdominal ultrasonography, the lesion shows no color uptake/ vascularity on color doppler study.

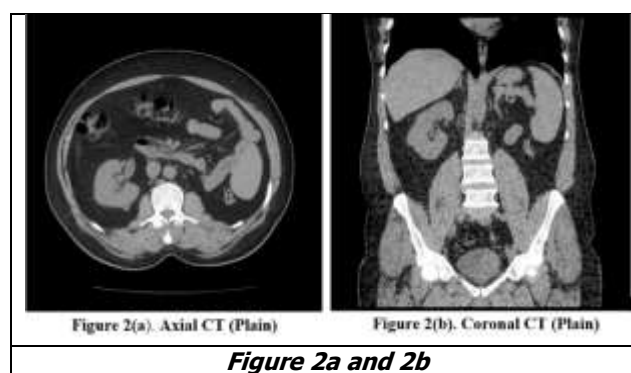


Figure 2(a) and 2(b). Axial and Coronal CT scan of the abdomen and pelvis shows left renal agenesis with compensatory hypertrophy of the right kidney.

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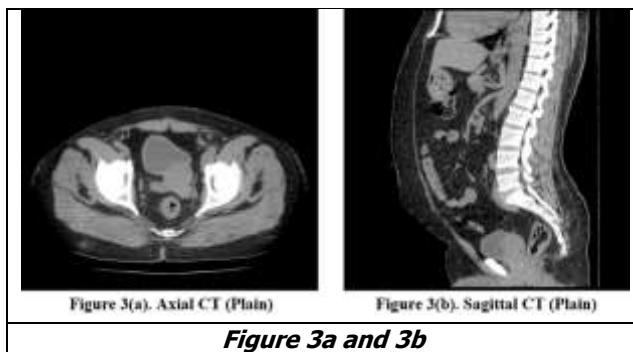


Figure 3(a). Axial CT scan of the pelvis shows an isodense cystic lesion arising from the left seminal vesicle abutting the inferolateral aspect of the bladder on the left side.

Figure 3(b). Sagittal CT scan shows the isodense cystic lesion located posterior to the bladder suggestive of a seminal vesicle cyst.



Figure 4(a). Axial T2W MRI of the pelvis shows an isointense/mildly hyperintense cystic lesion arising from the left seminal vesicle abutting the inferolateral aspect of the bladder on the left side.

Figure 4(b). Sagittal T2W MRI shows the well defined cystic lesion located posterior to the bladder wall with isointense/ mildly hyperintense signal intensity suggestive of hemorrhagic/ proteinaceous contents - Seminal vesicle cyst.

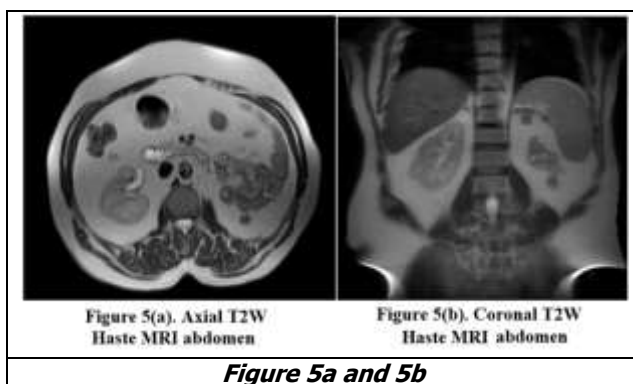


Figure 5(a) and 5(b). Axial and Coronal T2W Haste MRI of the abdomen shows left renal agenesis with contralateral compensatory hypertrophy of the right kidney.

In view of the above-mentioned clinical and radiological findings, the diagnosis of Zinner syndrome was made, a triad of Wolffian duct abnormality comprising of unilateral renal agenesis, ipsilateral seminal vesicle cyst and ipsilateral ejaculatory duct obstruction.

CLINICAL DIAGNOSIS

- Ureterocoele.
- Benign Prostatic Hypertrophy.
- Bladder Mass.

DIFFERENTIAL DIAGNOSIS

- Ectopic Ureterocoele
- Mullerian Duct Cyst
- Ejaculatory Duct Cyst

DISCUSSION OF MANAGEMENT

Trans-urethral resection and surgical excision of ejaculatory duct opening and TRUS guided aspiration of seminal vesicle cyst was advised as a definitive treatment option.

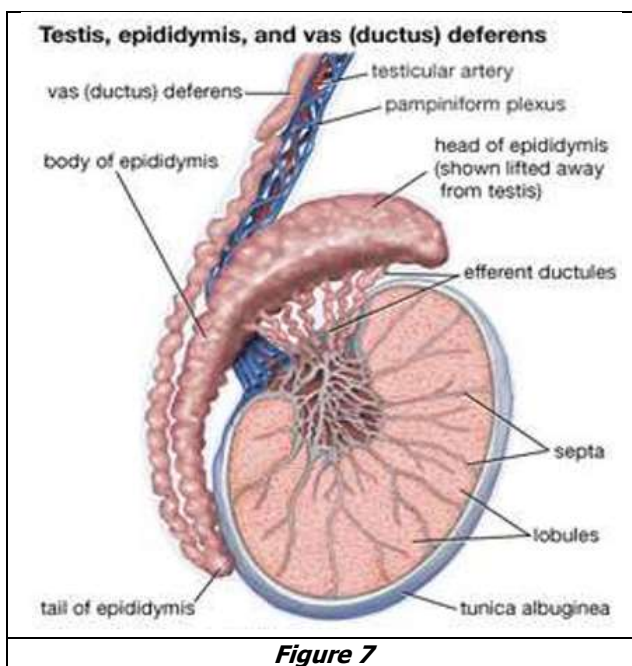
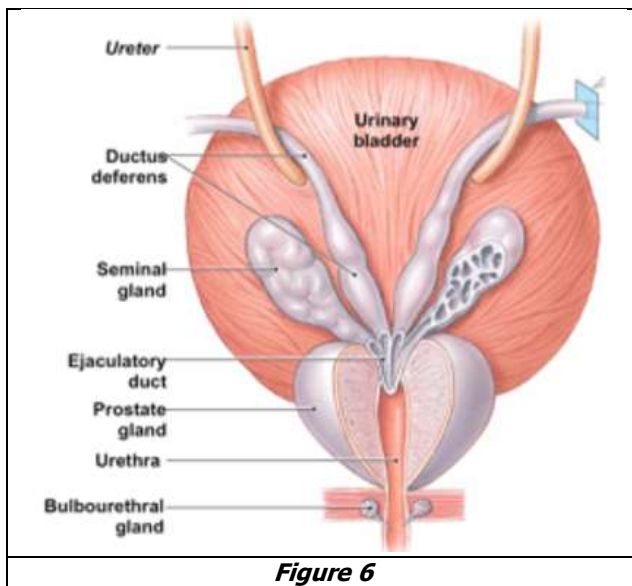
Discussion

At the fifth week of embryogenesis, the ureteric bud develops as an outgrowth from the caudal mesonephric (Wolffian) duct. The ureteric bud grows laterally and invades the metanephric blastema. When the ampulla of the ureteric bud and the metanephric blastema meet, the metanephric blastema begins to develop and mature into adult kidney. The urogenital ridge differentiates into pronephros, mesonephros and metanephros. The metanephros forms the definitive adult kidney. The glomeruli, proximal tubules and distal tubules is formed by the metanephric blastema. The renal pelvis, infundibulae, calyces and collecting tubules is formed by the ureteric bud. Renal agenesis occurs when there is failure to induce ureteric bud outgrowth.¹ The embryogenesis of kidney, ureter, seminal vesicle, and vas deferens is affected if an insult occurs during the first trimester of pregnancy.

Renal agenesis or dysplasia is likely if the insult during the embryogenesis is prior to 7 weeks of gestation, before the ureteric bud appears.² Zinner's syndrome is a congenital anomaly growth of the distal part of the Wolffian duct during the first trimester of embryogenesis between the 4th and 13th gestational week, comprising of unilateral renal agenesis, ipsilateral seminal vesicle cyst and ipsilateral ejaculatory duct obstruction. Patients usually present in the third or fourth decade of life. In males, the hemitrigone, bladder neck, urethra, seminal vesicle, vas deferens, ejaculatory duct, epididymis, paradidymis, and appendix epididymis develop from the paired mesonephric (Wolffian duct).³

It occurs due to common embryological origin of the ureteral buds and seminal vesicles from the mesonephric (Wolffian) duct.³ It was first described by Zinner in 1914 and 250 cases have been reported till date.^{4,5} Atresia of the ejaculatory duct leads to the obstruction and cystic dilatation of seminal vesicle and ipsilateral renal agenesis or dysplasia occurs due to abnormal ureteral budding, occurring due to

maldevelopment of the distal part of mesonephric duct (Wolffian duct).⁶



Bladder irritation leading to symptoms of dysuria, recurrent urinary tract infections, infertility, urinary urgency, painful ejaculation, epididymitis and prostatitis occurs as the seminal vesicles are located directly posterior to the bladder.⁷ Cyst formation in the seminal vesicle occurs due to obstruction at the level of ejaculatory duct, which may lead to azoo/oligozoospermia, resulting in primary infertility.⁸ Patients with seminal vesicle cysts less than 5 cm are usually asymptomatic and incidentally detected on cross-sectional imaging or ultrasound.⁹ Bladder and colonic obstruction occurs if the seminal vesicle cysts are larger than 12 cm and are termed as giant cysts and may lead to pelvic or perineal pain.¹⁰

On excretory urography, seminal vesicle cyst appear as an extrinsic smooth-walled filling defect along the inferolateral bladder surface. On ultrasonography, the

seminal vesicle cyst appears as an anechoic cystic pelvic mass with a thick and irregular wall in the inferolateral aspect of the bladder base. Presence of internal echoes within the cysts suggests prior hemorrhage or infection and sometimes the cyst may show calcification.¹¹ On computed tomography (CT scan) seminal vesicle cyst appear as a well-defined retrovesicular mass of water or near-water attenuation seen in the superolateral aspect of the prostate gland. On CT scan, the cyst may appear hyperdense due to proteinaceous or hemorrhagic contents.¹² On MRI seminal vesicle cyst usually appears hypointense on T1-weighted and hyperintense on T2-weighted images suggestive of fluid signal intensity. Presence of protein-rich contents or hemorrhage within the cysts lead to hyperintense signal on T1-weighted images and iso/hyperintense signal on T2-weighted images. Presence of high signal intensity on T1-weighted images with a convoluted tail connecting the cystic abnormality to the seminal vesicle suggests seminal vesicular origin of the cystic lesion.¹³

Treatment of seminal vesicle cyst includes TRUS guided aspiration of seminal vesicle cyst and surgical excision of the cyst. Transurethral resection of the ejaculatory duct (TURED), exploration, laparoscopic and robotic vesiculectomy are performed for symptomatic patients.¹⁴

True cysts of prostate gland, prostatic utricle cysts, ejaculatory duct cysts, Mullerian duct cysts, diverticula of the ampulla of vas deferens, bladder diverticula, and ureterocele are important differential diagnoses to be considered in the presence of seminal vesicle cyst. Mullerian duct cysts and ejaculatory duct cysts are midline in location, while diverticulosis of ampulla of vas deferens and ectopic ureterocele are more laterally located.¹⁵

FINAL DIAGNOSIS

Left Seminal Vesical Cyst with Ipsilateral Renal Agenesis and Ipsilateral Ejaculatory Duct Obstruction Suggestive of Zinner Syndrome.

Conclusion

Zinner syndrome, a triad of Wolffian duct abnormality is an uncommon but important diagnostic consideration in 3rd to 4th decade of life if the patient presents with recurrent urinary symptoms, painful ejaculation and abdominal or perineal pain. Zinner syndrome comprises of unilateral renal agenesis, ipsilateral seminal vesicle cyst and ipsilateral ejaculatory duct obstruction.¹⁶

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