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Case Report

Zinner's syndrome: Masquerading as pyonephrotic ectopic kidney ☆,☆☆

Faiz Manzar Ansari, MS[a,], Sajad Ahmad Para, MS, MCh[a], Shashank Singh, MS[a], Mohammad Saleem Wani, MS, MCh[b], Arif Hamid Bhat, MS, MCh[a], Manjul Kumar Maurya, MS[a]*

[a] *Department of Urology, Sher-I-Kashmir Institute of Medical Sciences, Room No. F 05, Old SR Hostel, SKIMS, Srinagar, 190011, India*

[b] *Urology and Renal Transplant, Sher-I-Kashmir Institute of Medical Sciences, Srinagar, India*

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ABSTRACT

Zinner's syndrome is a rare developmental anomaly of Wolffian duct, comprising a triad of seminal vesicle cyst, ipsilateral renal agenesis and ejaculatory duct obstruction, first described by Zinner in 1914. Several aberrations have been reported like renal dysplasia, ectopic ureteric orifice in one of the derivatives of Wolffian duct. Usually it presents in second to fourth decade of life with symptoms of urinary bladder irritation/obstruction, cyst distension, ejaculatory duct obstruction. The diagnosis is principally based on imaging studies, usually confirmed by MRI. Treatment is based upon the persistent symptoms or complications related to it. Excision of cyst is gold standard.

Introduction

Cystic malformations are the most common congenital malformations of seminal vesicles. Both seminal vesicle and ureteral buds develop from Wolffian duct and an insult during the first trimester of gestation may alter the embryogenesis of kidney, ureter, seminal vesicle, and vas deferens. One such rare condition was first described by Zinner in 1914, comprising a triad of ipsilateral renal agenesis, seminal vesicle obstruction and ipsilateral ejaculatory duct obstruction [1]. Approximately 200 cases have been reported till now. Zinner's syndrome is also considered to be the male counterpart of Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome seen in females.

Abbreviations: CT, computed tomography; DWI, diffusion-weighted imaging; FSH, follicle stimulating hormone; LH, luteinizing hormone; LUTS, lower urinary tract symptoms; MRI, magnetic resonance imaging; TURED, transurethral resection of ejaculatory duct; UTI, urinary tract infection.

☆ Work attributed to - Department of Urology, Sher-I-Kashmir Institute of Medical Sciences (SKIMS), Srinagar, India.

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* Corresponding author.
E-mail address: faizmanzaransari@gmail.com (F.M. Ansari).

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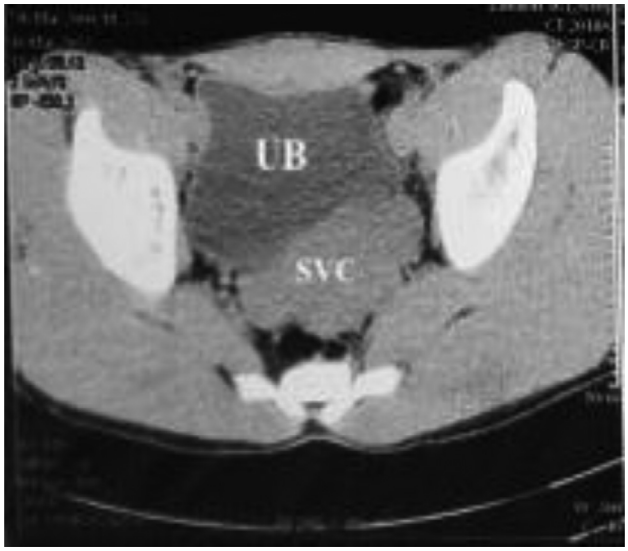


Fig. 1 – CT scan image (axial cut) showing urinary bladder (UB) and seminal vesicle cyst (SVC).

Case report

An 18-year-old male, unmarried, presented to our institute with c/o low backache and suprapubic discomfort. There was no history of lower urinary tract symptoms (LUTS), fever.

Abdominal and genitalia examination was unremarkable. Vas deferens was bilaterally palpable. On digital rectal examination prostate was normal with painless fluctuant left supraprostatic swelling.

USG was done which revealed normal right kidney, and left kidney not visualized in left renal fossa. There was evidence of cystic swelling akin to hydro/pyonephrotic kidney noted in pelvis posterior and superior to urinary bladder. Urine analysis showed only 4-6 pus cells/high power field.

CT Urography reported normal right kidney and right ureter with evidence of small reniform like structure in left renal fossa measuring 2.2 × 1.4cm with hypoplastic renal vein seen draining the structure into inferior vena cava. On excretory film no excretion of contrast seen with non-opacification of left ureter. Evidence of lobulated fluid density area seen in pelvis anterior to rectum and posterior to bladder, measuring 8.2 × 5.1cm showing no enhancement on post contrast images likely suggestive of seminal vesical cyst (Fig. 1).

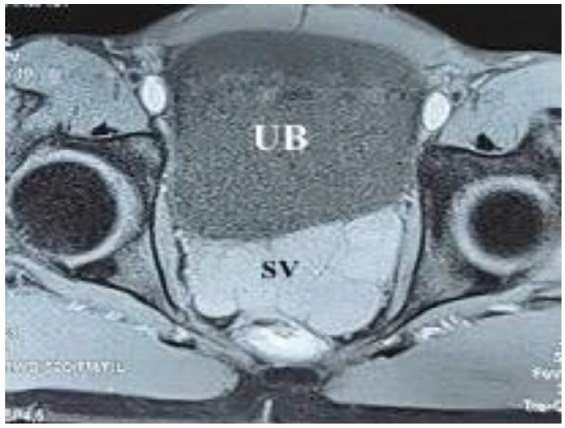
MRI pelvis revealed 94 × 44mm T2 hyperintense multicompartmental structure in retro-vesical pouch, suggestive of left seminal vesicle cyst (Fig. 2). On DWI there is mild restriction. Bilateral testes were normal.

In view of above mentioned clinical and radiological findings a diagnosis of Zinner's syndrome was made.

Semen analysis showed volume of 2 ml, with sperm count of 45 million/ml (50% actively motile), normal sperm morphology and 12-15 pus cells/high power field.

Serum testosterone, LH, and FSH were all within normal limits.

Initially, patient was managed conservatively with i.v. antibiotics and symptomatic treatment. Due to failure of con-



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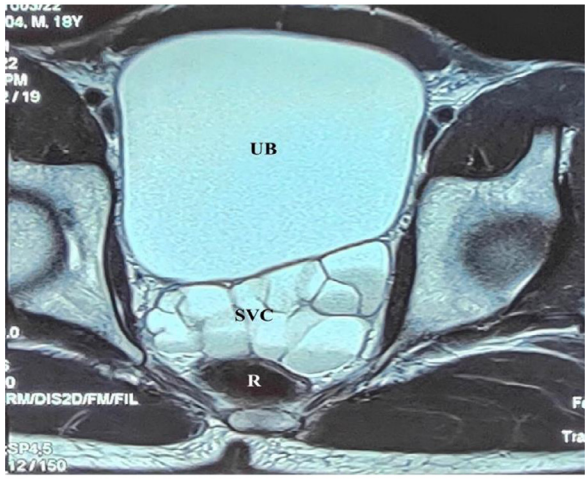

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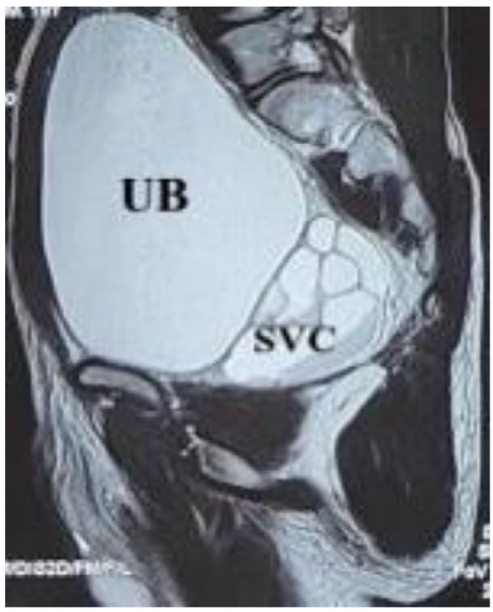

(c)

Fig. 2 – (A) T1-weighted MRI image (axial cut) showing hyperintense seminal vesicle cyst owing to proteinaceous content. (B) T2-weighted MRI image (axial cut section). (C) T2-weighted MRI image (sagittal section view). R, rectum; SVC, seminal vesicle cyst; UB, urinary bladder.

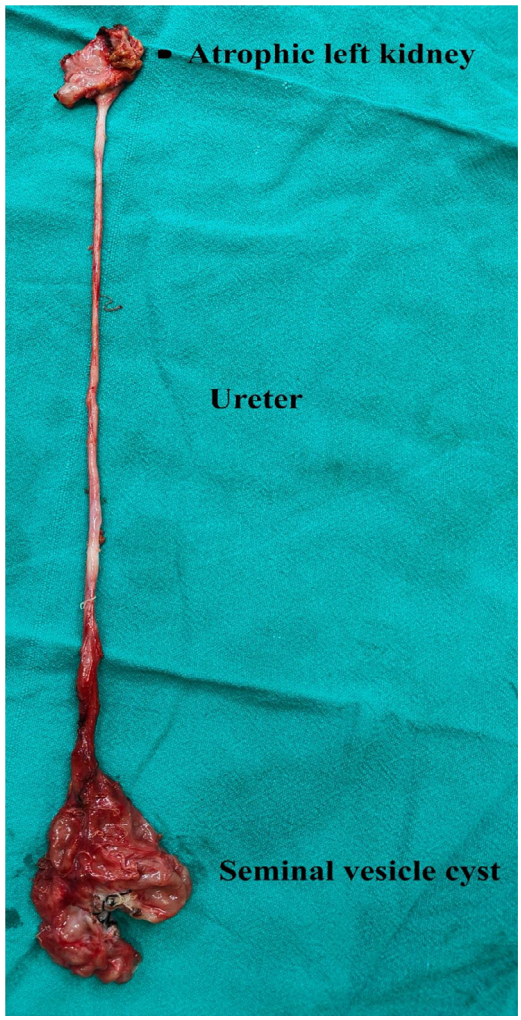

Fig. 3 – En-mass resected specimen of rudimentary left kidney with ureter draining into seminal vesicle cyst.

servative approach and persistence of symptoms, patient was planned for open exploration and cyst excision.

Cystopanendoscopy revealed raised left hemitrigone with absent left ureteric orifice.

Surgical exploration was done. Intraoperative findings were a large 11cmx6cm, multiloculated left seminal vesical cyst with ectopic insertion of left ureter into the cyst and atrophic 2.5 × 2cm left kidney. En-mass resection of left seminal vesicle cyst with rudimentary left kidney and left ureter was done (Fig. 3). Patient was discharged in satisfactory condition.

Histopathological examination revealed multiloculated seminal vesicle cyst, many of them filled with proteinaceous material with foci of calcification. A rudimentary kidney with few tubules and hyalinization with a luminal structure noted as ureter.

Discussion

Smith first identified seminal vesicle cysts in 1872, while the triad of ejaculatory duct obstruction, seminal vesicle cyst and ipsilateral renal agenesis was first identified by Zinner in 1914. This congenital anomaly is rare with an incidence of 214 by 100000 patients [2]. The ratio of right to left side is 2:1.

Zinner's syndrome is postulated to be a developmental anomaly of distal portion of Wolffian duct between 4th and 13th week of gestation [3]. As described by Tanagho and Schulman, it is essential that ureteric bud meets the center of metanephric blastema for normal development of kidney. If the ureteric bud develops too early, it will meet the mesonephros, located more cranially, which subsequently atrophy or disappears resulting in agenesis or dysplasia of the kidney, as seen in our case. The high location of ureteric bud results in delayed meeting of urogenital sinus, and this prolonged period of contact of the ureter with the Wolffian duct results in the ectopic orifice in one of the derivatives of the duct(seminal vesicles, ejaculatory duct, vas deferens, epididymis). In our case the ureter was found to enter the seminal vesicle cyst.

Patients with this anomaly usually presents in second to fourth decade of life corresponding to period of high sexual activity or reproductive activity [3]. Symptoms can be due to bladder obstruction (LUTS, strangury), bladder irritation (prostatitis like symptoms, urethritis, urethral discharge, hematuria, fever), cyst distension(pain lower abdomen, perineal pain, impaired defecation, scrotal pain), mesonephric duct obstruction(pain following ejaculation, diminished volume of ejaculate, infertility). Although one ejaculatory duct obstruction should not lead to azoospermia and infertility, as described in several literature reports. Several assumptions have been made to explain this feature like a congenital defect in the region of ejaculatory duct might block the free semen passage in the contralateral normal duct or unilateral testicular obstruction leads to formation of anti-sperm antibodies. Our patient presented with low backache and suprapubic discomfort. A high index of suspicion should be raised if a male patient presents with LUTS and has unilateral renal agenesis.

The diagnosis is typically based on imaging findings. Evaluation includes blood analysis, endocrine profile (FSH, LH, Testosterone), urine analysis, urine culture. Transrectal ultrasonography is now the recommended method for diagnosis and initial evaluation of seminal vesicle cysts [4]. Findings include an anechoic pelvic mass with thick irregular wall and occasional wall calcification, or the mass may contain internal debris reflecting prior hemorrhage and infection. However, our ultrasound was suggestive of hydro/pyonephrotic ectopic left kidney.

The suspected cases of Zinner's syndrome are usually confirmed by MRI [5]. Seminal vesicle cysts appear as hyperintense on T2-weighted images and hypointense on T1-weighted images. Presence of protein rich contents or previous episode of intracystic bleed may lead to hyperintense signal on T1-weighted images and hypointense signal on T2-weighted images. Seminal vesicle cyst needs to be differentiated from other pelvic cystic lesions like Mullerian duct cyst, prostatic cyst, diverticulum of ejaculatory duct. MRI gives excellent definition of soft tissues and relationship between the surrounding pelvic structures required for planning of the surgical management.

Urethrocystoscopy may show a hemitrigone due to underdevelopment of the homolateral ureteric unit or a complete

trigone with extrinsic compression of posterior wall of bladder. In our case the left ureteric orifice was absent with bulge on left posterolateral wall of bladder.

The findings of vasovesiculography include dilatation, deformity of seminal vesicle, ejaculatory duct stenosis, and reflux of contrast material in an ipsilateral ectopic ureter. The communication between the components of mesonephric duct can be delineated.

Additional information can be obtained by means of seminal vesicle aspiration which reveals a large number of sperms in the aspirate.

The management of this syndrome is clinically oriented. Asymptomatic and minimally symptomatic patients should be kept on regular follow up [5]. Antibiotics, transurethral aspiration of the seminal vesicle cyst or transurethral aspiration combined with substance instillation (alcohol, minocycline) are also proposed as conservative treatment. Transrectal aspiration should be discouraged because only 30% of patients become cured and there is considerable risk of infection and relapse. Perineal or abdominal route is preferred. Invasive treatment should be restricted to (1) symptomatic cases or patients who failed conservative treatment; (2) are young and therefore have a long life expectancy (to eliminate the risk of recurrence); and (3) if a malignancy cannot be excluded and histological examination of the cyst is necessary [3]. The various modalities include transurethral resection of ejaculatory duct (TURED), excision of the cyst.

TURED consist of resection of the distal prostatic portion and removal of proximal verumontanum until the opening of ejaculatory duct, using pure cutting current. The creation of transurethral fistula and transvesical drainage are also reported.

Excision of the cyst is gold standard. Open procedures have been described via transvesical, retroperitoneal, transperineal or posterior transcoccygeal approach.

Laparoscopic approach was first described by Kavoussi et al. In experienced hands laparoscopic resolution gives good results and is an effective and safe technique with minimal blood loss and minimal postoperative morbidity. Robotic assisted excision of seminal vesicle cyst has also been described with similar advantages of laparoscopy.

Our patient presented with pain lower back with multiple episodes of UTI with fever within a span of 2 months, not responding to conservative management and hence managed with open surgical exploration.

Conclusion

Seminal vesicle cyst combined with ipsilateral renal agenesis is a rare anomaly in the development of urogenital system. Zinner's syndrome should be suspected if a young patient presents with multiple and unspecific pelvic symptoms and has a known or incidentally detected renal agenesis. The diagnostic workup consists of transrectal ultrasonography, CT scan, MRI. Treatment is clinically oriented.

Ethical approval

This work is approved by the institution ethical committee with protocol number IEC/SKIMS Protocol #EC32/2022.

Patient consent

Written information consent was taken from patient for publishing the data related to him.

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