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Zinner Syndrome with Chronic Refractory Lower Urinary Tract Symptoms: A Case Report

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Authors' contributions

This manuscript is the collaboration of all the authors. Author AKS has designed the outline of the manuscript and the production of the first version. Authors BM and MBA have helped to collect data and bibliographic research and have supervised the work. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Zinner syndrome is a rare congenital urogenital anomaly characterized by unilateral renal agenesis, ejaculatory duct obstruction and ipsilateral seminal vesicle cyst due to developmental arrest in early embryogenesis affecting the caudal end of Mullerian duct. Here, we report a case of 27-year-old male, who presented with burning micturition, pain at base of penis, painful ejaculation and painful defecation for 10 years with history of recurrent urinary tract infection. Physical examination, transrectal ultrasonography, and pelvic magnetic resonance imaging (MRI) showed right renal agenesis, ipsilateral ejaculatory duct obstruction and right seminal vesicle cyst. He was found refractory to the conservative management of 6 months duration. Transurethral resection of veru with right ejaculatory duct and deroofing of the right seminal vesicle cyst was done with bipolar loop. The patient was symptomatically better during the follow-ups. The cases of Zinner syndrome have been addressed adequately but we had not found such cases from Nepal.

Keywords: Ejaculatory duct obstruction; renal agenesis; seminal vesicle cyst; transurethral deroofing of seminal vesicle cyst; zinner syndrome.

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1. INTRODUCTION

Zinner syndrome (ZS) is a rare congenital urogenital anomaly characterized by a clinical triad of unilateral renal agenesis, ejaculatory duct obstruction and ipsilateral seminal vesicle cyst due to developmental arrest in early embryogenesis affecting the caudal end of Mullerian duct [1]. Less than 100 such cases have been reported so far. Therefore, we report a symptomatic case of Zinner syndrome managed with transurethral resection of veru and ejaculatory duct with deroofing of seminal vesicle cyst.

2. PRESENTATION OF CASE

A 27-year-old male presented with burning micturition, pain at the base of penis, painful ejaculation and painful defecation for 10 years. He also complained of the poor and bifid flow of urine and incomplete voiding with multiple urological consultations at several centers in the past. He had the history of culture-proven recurrent urinary tract infection. He had a surgical history of left open pyelolithotomy 9 years ago and ureteroscopic laser lithotripsy 3 years ago. He was the father of an 8-year-old child. His genitalia were normally developed. On digital rectal examination, there was a tender, smooth, firm prostate with a tender boggy swelling at the right side of the base of prostate. Routine analysis of urine was normal, whereas urine and semen showed the growth of Escherichia coli. The infection was treated accordingly. The ultrasonography of abdomen and pelvis showed absence of right kidney with right-sided seminal vesicle cyst of 6cmX2.6cmX2.8cm dimension. An obstructive pattern of uroflowmetry was observed (Qmax 8.2mL/sec) and absent right kidney was also appreciated on MRI [Image 1]. The transrectal ultrasonography and MRI showed ejaculatory duct obstruction in the right side and ipsilateral seminal vesicle cyst of 6 cm X 3.2 cm X 3.4 cm dimension [Image 2]. However, the serum level of renal function test was within the normal limit. Flexible cystoscopy appreciated cystic swelling compressing the posterior aspect of the urinary bladder near the right side of the bladder neck and the trigone with bilateral ureteric orifice was visible normally. Hormonal workup (Leutinizing Hormone, Follicle Stimulating Hormone, Prolactin, and Testosterone) was within the normal limit. The semen analysis showed normal parameters. The final diagnosis of symptomatic Zinner syndrome was carried. He was found refractory to the conservative management (Tamsulosin 0.4 mg) of 6 months duration. He was well explained about the possible complications of transurethral deroofing of the cyst (urethral stricture, urinary incontinence and retrograde ejaculation). Transurethral resection of veru with right sided ejaculatory duct and deroofing of the right seminal vesicle cyst was done with bipolar loop [Images 3a,3b]. The histopathology of the seminal vesicle cyst wall showed no any malignant features. He progressed well and was completely free of urinary symptoms on 2 weeks, 6 months and 12 months follow ups. He had no any remnant or recurrence of right-sided seminal vesicle cyst on ultrasonography imaging. His urine and semen samples were found negative (without any microbiological growth at 6 months follow up). The uroflometry showed normal parameters at 6 months (Qmax17mL/sec) and 12 months (Qmax19 mL/sec) follow ups. Currently, the patient is doing well and has been kept on follow ups (just in case of any necessity).

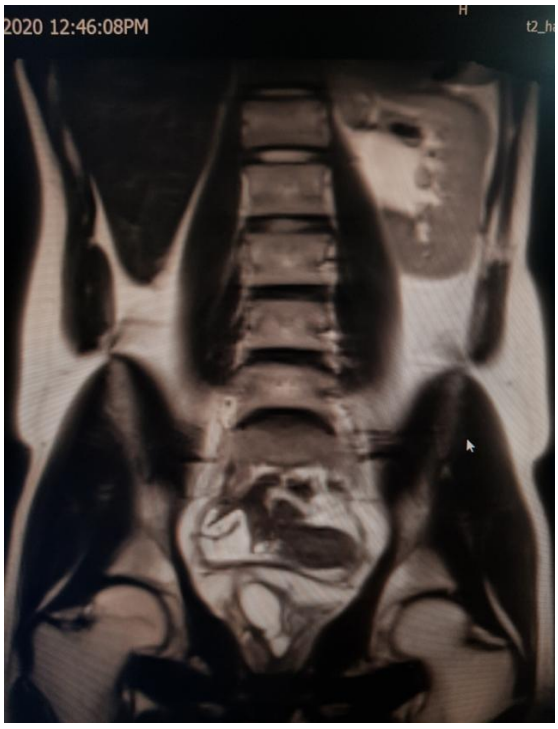

Image 1. MRI showing absent right kidney and right ureter

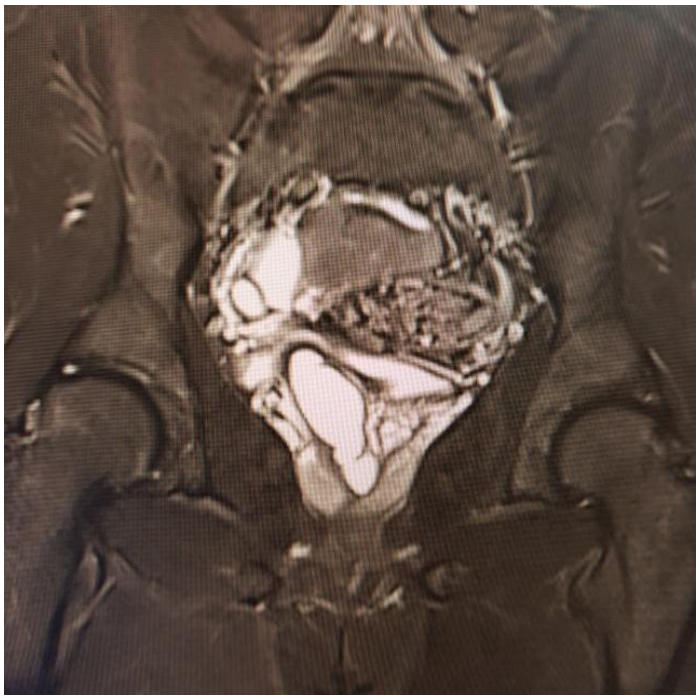

Image 2. MRI showing right-sided seminal vesicle cyst with ipsilateral ejaculatory duct obstruction, normal anatomical contralateral seminal vesicle and ejaculatory duct seen

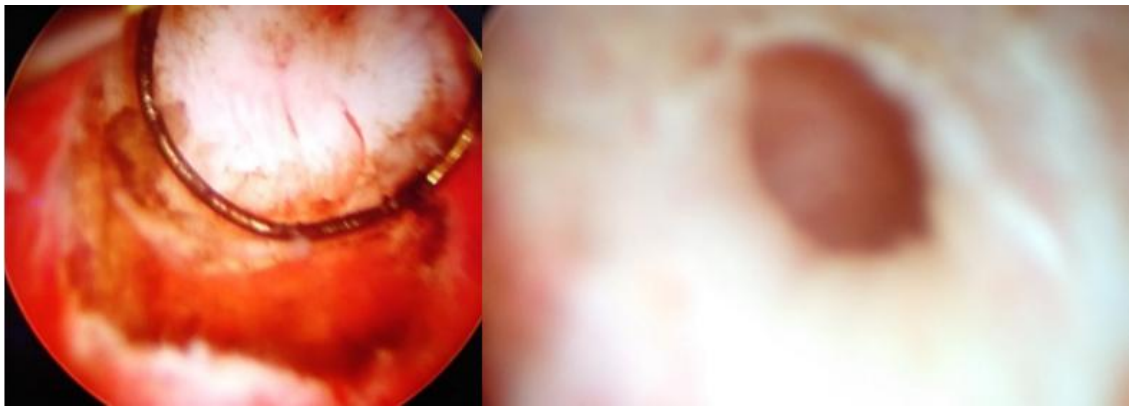

Image 3. a) Transurethral resection of right ejaculatory duct and veru, b) Transurethral deroofing of right seminal vesicle cyst

3. DISCUSSION

Zinner syndrome is a congenital anomaly characterized by the clinical triad of unilateral renal agenesis, ipsilateral ejaculatory duct obstruction and the seminal vesicle cyst due developmental arrest in early embryogenesis affecting the caudal end of Mullerian duct. It is a mesonephric duct anomaly occurring in 7 weeks of gestation. The first case was described by Zinner in 1914 [1–3]. Zinner syndrome is also considered to be the male counterpart of Mayer-Rokitansky-Kustner-Hauser (MRKH) syndrome (uterovaginal aplasia) seen in females [2].

The perineal pain, ejaculatory pain and dysuria are the most common presenting features; however, they may present with frequency, hematuria, recurrent urinary tract infections and intermittent scrotal pain. The clinical feature depends on the size and location of the cyst [4]. The patients usually present with infertility during

their third or fourth decade of life. Our patient had fulfilled all the criteria for the diagnosis, however, there was no fertility issue with him. His semen analysis was within normal limit, in spite of recurrent bacterial growth on culture. Cysts of less than 5 cm size are usually asymptomatic. In our case, the size of the seminal vesicle cyst was more than 5 mm and the patient was symptomatic as mentioned above. The largest cyst of up to 12 cm has been reported. Large cysts can also cause bladder outlet or colonic obstruction [3]. Malignant transformation in the seminal vesicle cyst has also been reported in a 17-year-old young boy [5]. However, the resected specimen did not show any features suggestive of malignancy on post-operative histopathology. The utrasonography of abdomen & pelvis, transrectal ultrasonography, intravenous urography, CT scanning, MR imaging and cystoscopy are the common modalities used for the diagnosis, anatomical assessment and the surgical planning. MRI scan gives excellent definition of the soft tissues and surgical anatomic study with multiplanar demonstration of the relations between the surrounding pelvic structures and is recommended for the confirmation of diagnosis [3]. We used all these modalities during the assessment of our patient. We began clinical correlation for ZS after the ultrasonographic findings. Latter, the MRI confirmed the syndrome.

Though the case had multiple history of renal stone formation, we could not perform the metabolic assessment for the recurrent stone formation because the patient was worried and suffering from lower urinary tract symptoms at this time. The imaging did not show any evidences of stone during our visits.

The cystoscopy is helpful to understand the anatomy of the urethra and its connection with the cyst. It further evaluates the possible associated bladder anomalies [1]. Percutaneous fine-needle aspiration may be useful to reconfirm the diagnosis by revealing spermatozoa from the cyst [6]. But we did not feel the necessity of doing aspiration of the cyst to reach the diagnosis in our case. The Prostatic Utricle Cyst, Mullerian Duct Cyst and Ejaculatory Duct Cyst are common differential diagnosis, which are easily excluded by common imaging modalities [1]. There is no such gold standard treatment for ZS. We also tried to manage this case conservatively. The conservative treatment is an efficient and safe option in the management of asymptomatic or poorly symptomatic patients [2,4]. However, our patient did not responded well after 6 months use of Tamsulosin (alfa 1a blocker) as conservative treatment. Transurethral intralesional instillation of some substances like alcohol and minocycline are also proposed as conservative treatment [7]. The surgical management is suggested in severely symptomatic and refractory to conservative treatment cases. Open and transurethral resection as well as transrectal aspiration has been proposed. We found transurethral resection and deroofing of the seminal vesical cyst effective and safe modality for the management of this condition. It managed the issues and made the case free of botherness which we evaluated routinely during follow up. The open approaches demand larger incisions with extensive dissection and therefore, they are associated with a high risk of morbidity in terms of erectile dysfunction and rectal injury [8]. These approaches are limited by the inability to treat upper urinary tract anomalies. Endourethral, laparoscopic and robotic approaches have been associated with minimal blood loss, quick convalescence, minimal postoperative pain and a good cosmetic result. The robot-assisted laparoscopic excision is a safe and feasible optionto treat large seminal vesicle cysts. Therefore, these approaches should be the gold standard for treating ZS [9].

4. CONCLUSION

A case of chronic and refractory lower urinary tract symptoms in a young male may be syndromic. Such refractory to conservative treatment case of Zinner syndrome can be managed by transurethral resection of veru and ejaculatory duct with deroofing of seminal vesical cyst safely and successfully with no complications. Infertility may not be an issue of typical ZS at all.

CONSENT AND ETHICAL APPROVAL

As per international standard or our national research guidelines participant's consent and ethical approval has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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