A Case Report on Zinner Syndrome – A Rare Congenital Malformation

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ABSTRACT

Zinner syndrome is a rare congenital triad of unilateral seminal vesicle cyst, unilateral ejaculatory duct obstruction or agenesis and ipsilateral renal agenesis. Here we present a case of a healthy 22 year-old male presenting with complaints of low back ache and painful ejaculation since 6 years. On radiological evaluation with MRI abdomen and pelvis, a horse shoe kidney with atrophy of right limb and a right seminal vesicle cyst was revealed. Because of persistence of symptoms after a trail of medical management, the patient opted for surgical intervention. Laparoscopic seminal vesicle cyst excision was done and vas deference was dismembered from communication with ureter. Post operatively, patient's symptoms were dramatically improved.

Key words: Zinner syndrome, Duct obstruction, Renal agenesis, Laparoscopy, Cyst excision.

INTRODUCTION

Zinner syndrome is a rare congenital triad of unilateral seminal vesicle cyst, unilateral ejaculatory duct obstruction or agenesis and ipsilateral renal agenesis that affects males.[1] It is believed that an embryologic developmental anomaly of Wolffian distal mesonephric duct that appears between 4 and 13 weeks of gestation and is considered as male counterpart of Mayer-Rokitansky-Kuster-Hauses syndrome seen in females.[2] The embryological origin of the kidneys and seminal vesicles is similar in males; the kidney is formed by the metanephric blastema which is induced by ureteral bud that originates from the dorsal aspect of the distal mesonephric duct. The mesonephric duct gives rise to most of the genital tract including epididymis, vas deferens, ejaculatory duct and seminal vesicle. Any malformation of the ureteral bud or mesonephric duct can cause Zinner syndrome.[3]

Most of the patients with Zinner syndrome are asymptomatic. In general symptoms are detected incidentally and they are commonly present and exhibit between puberty and fourth decade of life in which the person is sexually active and the seminal fluid that is produced gets accumulate due to ejaculatory duct obstruction.[4] Patients with Zinner syndrome commonly present with infertility, dysuria, urgency, prostatism and painful ejaculation.[5] The syndrome may often present with voiding symptoms after during the period of utmost sexual or reproductive activity i.e., mostly in second and third decade of life.[6] Diagnosis can be made by radiologic modalities such as intravenous pyelography, ultrasonography, vasovesiculography; contrast enhanced computed tomography and magnetic resonance imaging.[7] Selection of treatment options mainly depends on the presentation of symptoms. The treatment strategy mainly focuses on seminal vesicle cyst. Patients are mostly managed conservatively. The treatment of choice for seminal vesicle cysts is surgical resection and other methods such as transurethral puncture, aspiration, transurethral resection of seminal colliculus and vas deferens.[8]

CASE HISTORY

A 22 year old male patient was admitted to urology department at NRI Medical College

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and Hospital, Chinakakani, Guntur, with complains of back ache on right side since 6 years, painful erection, ejaculation and inability to retract foreskin. Patient was informed of a solitary left kidney two years back. On examination tenderness was noted in right iliac fossa. Based on these clinical findings patient was provisionally diagnosed as having Phimosis, seminal vesicle cyst, absent right kidney, dilated right ureter and is treated symptomatically with T. Amitryptyline 10mg BD, T. Pantoprazole 40mg OD, Syp. Lactitol monohydrate 10ml BD, T. Drotavarine SOS. Diagnostic Cystoscopy showed normal appearance of urethra, prostate, seminal vesicle and left vesicoureteral junction. MRI of abdomen (Figure 1 shows MRI report of Patient) and pelvis revealed a horse shoe kidney with atrophy of right limb and a right seminal vesicle cyst was revealed, MR triad s/o Zinner syndrome - ectopic and dysplastic right kidney with moderate hydroureterosis up to the distal end, right seminal vesicle cyst secondary to ejaculatory duct obstruction, mild right hydrocele.

Based on these reports patient was diagnosed as having Zinner syndrome. Because of persistence of symptoms patient opted for surgical intervention. Through laparoscopy seminal vesicle cyst excision was done (laparoscopic image was shown in Figure 2). Vas was dismembraned from communication with right ureter, pelvic drain placed and site closed. 10 ml of the fluid was drained on Day 1 and 20 ml on Day 2 and was treated with Inj.Cefoperazone sodium (1.5g) + Sulbactam (500mg) IV BD, Inj. Ranitidine 50mg IV BD, Inj. Paracetomol 1g IV TID, T. Amitryptyline 10 mg PO BD. Post operatively patient symptoms were dramatically improved.

DISCUSSION

Zinner syndrome was first described by Zinner in 1914 and 200 cases of seminal vesicle cysts associated with ipsilateral renal agenesis have been reported in the literature.[9] Incidence is 1 in 3000 to 1 in 4000 newborns. Patients with this condition are usually normal but sometimes presents with decreased urine output, increased frequency of urine output and pain over the perineum, or epididymitis.[10] The developmental anomaly can be explained by close embryonic relationship between genital and urinary tract. The Wolffian duct that forms the male reproductive system as well as the ureteric bud is a paired structure. Between 6th-8th gestational week, the orifice of the distal mesonephric duct and the ureteric bud separates and the ureteric orifice migrates toward the metanephric blastema, whereas the distal part of mesonephric duct under the influence of testosterone and anti-Mullerian hormone, forms the hemitrigone, the bladder neck, the urethra up to the external sphincter, the seminal vesicle, vas deferens, ejaculatory ducts, epididymis, paradidymis and appendix of the epididymis. During the 4th to 6th week of gestation, the metanephric blastema secretes growth factors, which induce the growth of the ureteric bud toward it. The ureteric bud also secretes growth factors and proliferates, fusing with the metanephric blastema and in turn inducing the blastema to become the primitive kidney. Disturbance in any of these inductive events during this period of embryogenesis such as mutation of metanephric blastema, or disruption of retinoic acid signaling, causes inhibition of ureteric bud growth with failure of fusion of the Ureteric bud with the metanephric blastema and renal agenesis.[11]For this rare anomaly several optional treatments are available. Existence of bothersome symptoms is a significant factor that can affect the treatment options and most investigators recommend treatment only for symptomatic patients.[12] Only the symptomatic forms of treatment are justifiable and approach may be made by trans-vesical way, extra bladder or laparoscopically.[3]

Logigan *et al.* described a patient dysuria, polakiuria, dribbling, nocturia, hyposeprmia, oligoasthenoteratozoospermia, ejaculatory pain, chronic

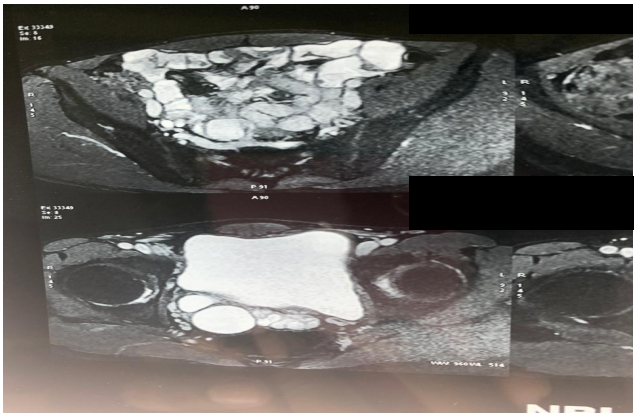

Figure 1: MRI Image Showing Ejaculatory Duct Obstruction and Seminal Vesicle Cyst.

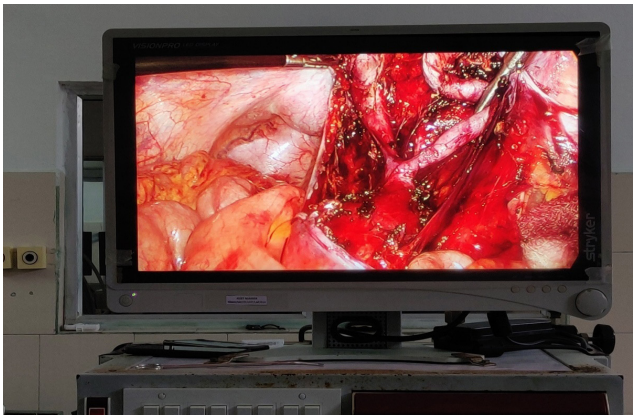

Figure 2: Laparoscopic Image Showing Junction of Ureter and Vas Deferens.

constipation and diffuse perineal and hypogastric pain, that started 9 weeks prior to admission. In this patient *vasa deferentia* have been identified, isolated from the cystic neovascular emergences and the cystic wall was extracted using a specimen bag. Jackson-Pratt drain was used for the pouch of Douglas, with the exteriorization of about 60 ml of serosanguinous liquid, which was removed after 24 hr. The actual console time was 85 min, estimated blood loss during procedure 50 ml and the patient was discharged day four postoperatively.[2] Slaoui *et al*. described a 39 year old father of five children presenting with terminal haematuria, pyuria associated with right lumbar pain radiating to external genitalia and a 35 year old patient hospitalized for subocclusion underwent Ultrasonography showed a retrovesicle liquid mass who was benefited from laparoscopic removal of cyst.[4] Kori R *et al*. described a 19 year old unmarried male patient presented with complains of recurrent urinary tract infections, poor urinary stream which slightly improved on straining from the past 1 year. Physical examination showed non-tender bulge just above the prostrate. Computed tomography (CT) scan revealed absent right kidney and well-defined peripherally enhancing cystic lesion with hyperdense contents arising likely from the seminal vesicle. Ultrasound examination of the abdomen showed absent right-sided kidney with mild degree of cystitis. Transrectal ultrasonography showed a cystic lesion of size 5 cm × 4 cm, with internal echoes arising from the right seminal vesicle abutting the prostrate and urinary bladder posteriorly. The patient did not agree for surgical removal, henceforth, transrectal ultrasound-guided aspiration was done yielding 8 ml thick viscid content with no sperms on microscopic examination. The patient followed up for 4 months and is healthy with adequate urinary stream and flow rate maintained at 19 ml/min.This patient presented with back ache on right side since 6 years, painful erection, ejaculation and inability to retract foreskin. MRI of abdomen and pelvis revealed a horse shoe kidney with atrophy of right limb and a right seminal vesicle cyst, MR triad s/o zinner syndrome - ectopic and dysplastic right kidney with moderate hydroureterosis up to the distal end, right seminal vesicle cyst secondary to ejaculatory duct obstruction, mild right hydrocele and was managed with laparoscopic seminal vesicle cyst excision and Vas was dismembraned from communication with right ureter, pelvic drain placed and port sites closed.

CONCLUSION

The combination of ipsilateral renal agenesis and ipsilateral seminal vesical cyst is a very rare combination and this triad is called Zinner syndrome. It is a rare congenital anomaly, with symptoms usually responding to conservative measures. A patient with persistent symptoms can be managed by minimal invasive surgical approach, with minimal manipulation, addressing only the core issue of patient symptoms.

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Declaration of Patient Consent

The authors certify that they have obtained the patient consent form. The patient has given his consent for images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal patient identity.

ABBREVIATIONS

MRI: Magnetic resonance imaging; **MR:** Magnetic resonance; **CT:** Computed Tomography.

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