

Zinner's Syndrome

Ahmed E Bakheet^{1*}, Ala. M. Abd Elgyoum², Mogahid M A Zidan³, Waleed H. Mohammed²

¹Zaharat Al-Amal poly Clinic-1, Western Ring Street, exit28, Al-Madeena street, Riyadh-Saudi Arabia.

²Faculty of Radiology & Nuclear Medicine Sciences, Radiology department, National Ribat University, Nile Street Burri, Postal Code-11111, Khartoum, Sudan

³Medical Imaging Technology Department, Al-Ghad International College for Applied Medical Science, Abha, KSA

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Email: ahmedezzeldin65@gmail.com

*Corresponding author: Ahmed Ezz Eldin Abdelrazig Bakheet

Abstract

Case Report

Is an uncommon urogenital malformation of congenital seminal vesicle cyst associated with ipsilateral renal agenesis. It is considered to be the anomaly development in early embryogenesis affecting the distal part of Mullerian duct. A 25 year old man, Had history of monorchism, presented with left loin and ejaculatory pain, an abdomen-pelvis ultrasound (US) was performed and demonstrated left atrophic kidney and ureteric obstruction, the MRI images showed high signals left seminal vesicle cysts on T2 and T2 fat sat images which reflect inspissated secretion with ectopic insertion of the tortuous ureters into the cystic masses. Also, the left ejaculatory duct was untraceable suggesting obstruction (Zinner's Syndrome). The patient was conservatively treated as he refused any surgical intervention including cystic mass aspiration.

Keywords: Urogenital, malformation, seminal,agenesis and Ultrasound.

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INTRODUCTION

Embryologically the urogenital organs are derivatives from mesonephric duct which is also known as wolffian duct. This explains the associated congenital anomalies of reproductive and urinary systems. Amid these anomalies is the rare Zinner syndrome, which unilateral ipsilateral seminal vesicle cyst, ipsilateral renal agenesis and ejaculatory duct obstruction [1-3]. Though most of the published cases reported typical findings, some described coexistent of other urogenital anomalies such as remnant ureteric bud [2, 4]. Usually patients with Zinner anomaly are diagnosed in their 2nd or 3rd decade when they are being sexual active [2, 5, 6]. Moreover, most of the Zinner syndrome complaints are related to the cyst size and consequence mass-effect, in addition infertility [6]. Medical imaging plays crucial role in diagnosis of Zinner syndrome, however, the choice of imaging modality depends on accessibility and the results of preceded examinations. Therefore, in most cases ultrasonography (US) was the first choice. In this case report we aim to enrich current published reports concerning Zinner syndrome finding, and to explicitly demonstrate the roles of medical imaging in the diagnosis of such cases.

CASE REPORT

A 25 year old man, who had been married for one year and he and his wife never had conception. Had history of monorchism and surgically treated left inguinal hernia 15 year ago. Presented with left loin and ejaculatory pain. While previous reports were inaccessible, the patient denied any history of lower urinary tract or genital infections or trauma. Physical examinations of the reproductive organs revealed single right testis, which was normal in size, and excluded the possibility of recurrent inguinal hernia. In addition to, laboratory tests were normal except for semen analysis, which confirmed hypospermia. An abdomen-pelvis ultrasound (US) was performed and demonstrated left atrophic kidney and ureteric obstruction, compensatory hypertrophic right kidney and cystic pelvic mass in supra-prostatic region (FIG1- A). Therefore, to further delineate the pelvic mass trans-rectal ultrasonography (TRUS) scan was conducted. The scan reported an anechoic grapes-like dilated cystic seminal vesicle masses measured 60*30 mm, and dilated left ejaculatory duct with internal debris (FIG1-B). Furthermore, in order to assess relationship between the pelvic mass and adjacent organs and to rule out ectopic ureteric insertion, magnetic resonance imaging (MRI) pelvis and urography were performed. The MRI images showed high signals left seminal vesicle cysts on T2

and T2 fat sat images which reflect inspissated secretion with ectopic insertion of the tortuous ureters into the cystic masses. Also, the left ejaculatory duct was untraceable suggesting obstruction (FIG 2). The patient was conservatively treated as he refused any surgical intervention including cystic mass aspiration.

DISCUSSION

Zinner syndrome is one of the rarest congenital anomalies and its defined as condition comprising a triad of unilateral renal agenesis, ipsilateral seminal vesicle obstruction and ipsilateral ejaculatory duct obstruction. ZS was discovered and diagnosed by Zinner in 1914, and 200 cases have been reported in the literature [1]. This rarest congenital anomaly is usually discovered and diagnosed in the 2nd-4th decade of life [7].

The incidence of ZS in the pediatric age is difficult to define [8]. Sheih *et al.* Found 13 cystic dilatations within the pelvis (six CSVCs in male and seven Gartner's cysts in female) associated with ipsilateral renal anomalies during a renal US screening among 280,000 infants and children in Taipei. Thus, the estimated frequency of ZS is about 0.00214% [7]. To the best of our knowledge, ZS has been reported in 19 papers and 50 patients in pediatric age, with a median age at diagnosis of 12.5 years (range 0–18 years) [9-13]. The side was right in 23, left in 24, bilateral in one and not reported in two patients. Associated ipsilateral urinary anomalies were 19 MCDK, 26 renal agenesis/hypoplasia, two urinary tract duplications—bilateral in 1 case⁽¹⁴⁾ -one ureteral drainage into the prostatic urethra, and one ureteral drainage into the ED (8). No anomalies were reported in three patients [14]. ZS may be associated with other malformations, such as cardiac (ventricle septum defects), anorectal, PUV, and contralateral MCDK [15]. Only in 12 cases the malformation was diagnosed in the first year of age as in our experience, with a median age of 5 months (range 0–10 months) [12, 13, 15-18].

The diagnosis of Zinner syndrome is based principally on imaging techniques, finding the characteristic cystic dilatations of the seminal vesicle and ipsilateral renal agenesis. The differential diagnosis is wide and includes mainly other causes of pelvic cysts, including true cysts of the prostate, prostatic utricle cysts, ejaculatory duct cysts, mullerian cysts, bladder diverticuli, and ureterocele. Other causes to consider are pelvic abscesses, hydronephrotic pelvic kidneys, and regional cystic neoplasms. To the authors' knowledge, no studies have analyzed the risk of malignant degeneration. The diagnostic procedures to evaluate a patient with Zinner's syndrome may include blood analysis and endocrine profile (FSH, LH, and

testosterone), urinalysis and urine culture, renal and pelvic/transrectal ultrasonography, CT or MRI, sperm analysis and eventually cyst aspiration [1].

The management of this syndrome should be clinically oriented and follow-up in asymptomatic and minimally symptomatic cases[13]. Antibiotics, transurethral aspiration of the seminal vesicle cyst or transurethral aspiration combined with substance instillation (alcohol and minocycline) are proposed as conservative treatment[19]. Invasive treatment should be restricted to symptomatic cases or patients who failed conservative measures and usually consists in seminal way disclosure through transurethral resection of the ejaculatory duct or balloon dilatation and open, laparoscopic or robotic vesiculectomy.

MRI to better characterise anatomic relationship of SV cyst diagnosis is based on imaging technique, finding characteristic of cystic SV and ipsilateral renal agenesis. Differential diagnosis, prostatic cysts, utricle cyst, ejaculatory duct cyst, UB diverticula, Trans rectal US most recommended for SV cysts, finding includes: an anechoic pelvic mass with thick and irregular wall and occasional wall calcifications, or the mass may contain internal debris reflecting prior haemorrhage or infection. MRI the best in making definitive diagnosis of SV cyst without the need to resort for more invasive techniques finding: SV cyst appears hyper intensity on T2 and hypo intensity on T1. MRI gives excellent soft tissue definition which permits the best anatomic study with multiplayer demonstration of the relation between pelvic structures. MRI recommended confirming diagnosis and planning of surgical management.

CONCLUSION

Seminal vesicle cysts combined with ipsilateral renal agenesis are a rare anomaly in the development of the urogenital system. These usually occur in males between the second to fourth decades of their life. The usual symptoms that are caused by the seminal vesicle cysts are bladder irritation and obstruction as well as pain in the perineum and scrotum. The diagnostic work-up consists of transrectal and abdominal ultrasonography, CT scan and MRI. For the removal of the seminal vesicle cyst there are techniques that are used nowadays due to advanced technologies such as open surgery and transurethral deroofting of the cyst.

Although Zinner syndrome is an infrequent condition, its characteristic presentation should be enough for the knowledgeable radiologist to be able to make a satisfactory diagnosis and contribute to its management.

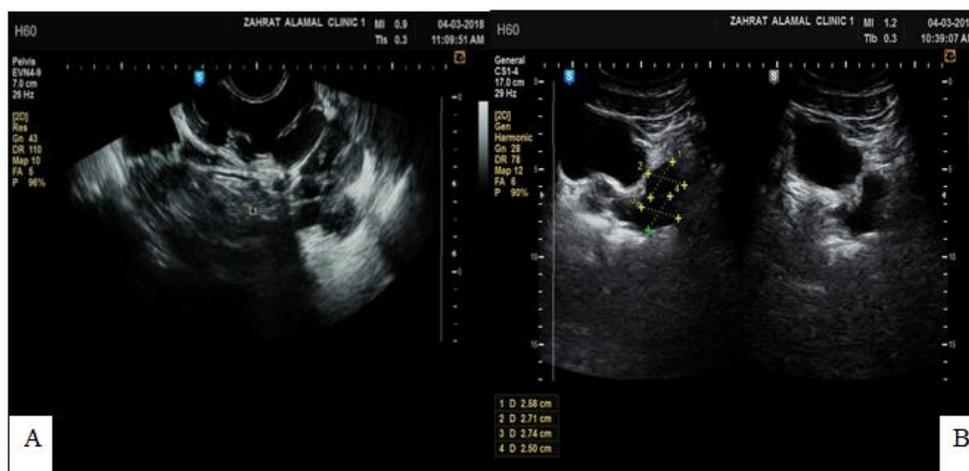


Fig-1: A. Transrectal: normal prostate and 6 cm SV cyst with echogenic content usually related to intracystic infection. B. Transabdominal US: Showed SV cyst associated with or renal agenesis

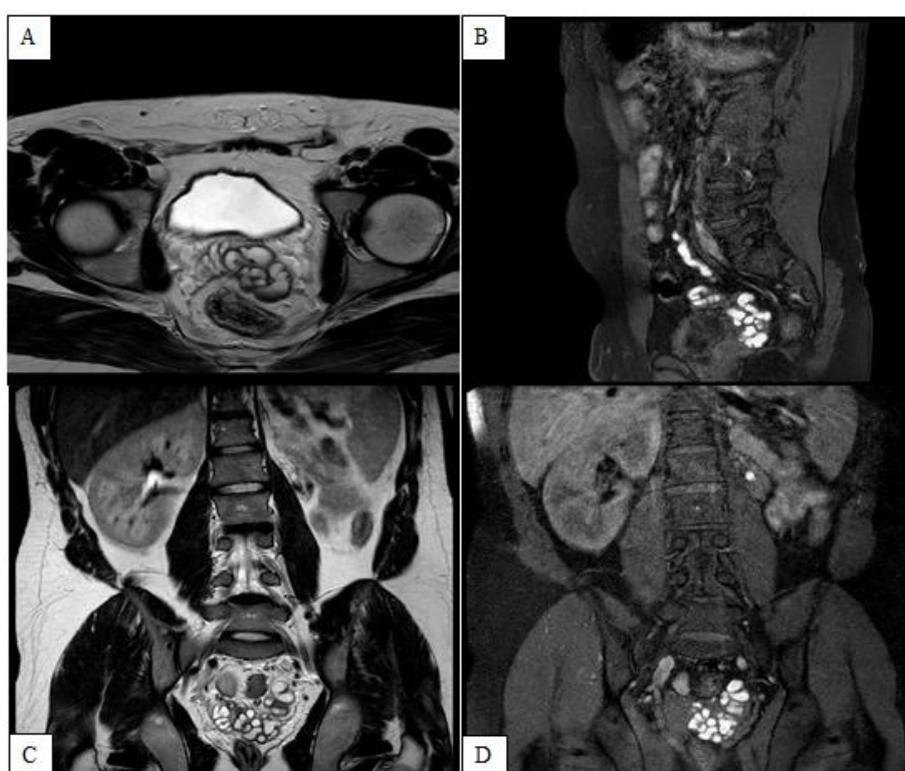


Fig-1: (A) MRI axial section T2. (B) MRI sagittal section T2 fat sat, (C) MRI coronal section T2 and (D) MRI coronal section T2 fat sat showing diffuse dilatation of left seminal vesicles with high signal intensity

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