Massive seminal vesicle cyst with ipsilateral renal agenesis – Zinner syndrome in a Saudi patient

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INTRODUCTION

Seminal vesicle cysts associated with ipsilateral renal anomalies (Zinner syndrome) are an extremely rare phenomenon; the reported frequency is approximately 0.0046%, with only a few case reports available in the literature confirming their presentation and outlining their management worldwide.[1,2] Symptoms leading to diagnosis are nonspecific including long-standing urinary tract symptoms and painful ejaculation, and many a times, the diagnosis is made incidentally during abdominal imaging for nonrelated diagnosis.[2‑4] This case report is one of the first cases of Zinner syndrome in the Saudi population managed with a laparoscopic approach. A brief literature review is also provided.

CASE REPORT

A 28-year-old Saudi gentleman presented to our outpatient clinic with nonspecific symptoms of malaise, fatigue, and myalgia. No significant past medical or surgical history was noted. Physical examination was within normal. The patient mentioned having an ultrasound scan of the abdomen at his local hospital, which had demonstrated right renal agenesis; however, no other abnormalities were noted; at that time, no further investigations were done as he had normal laboratory analysis.

Repeat ultrasound scan abdomen along with basic laboratory analysis was recommended. Complete blood count, kidney profile, liver function tests, and thyroid profile were within normal limits. Urine analysis was negative for signs of hematuria or infection. Ultrasound abdomen demonstrated an incidental finding of a large midline heterogeneous pelvic cyst measuring 11.5 cm × 9 cm × 9.5 cm, with a smooth wall, localized anterior to the rectum and not separated

Abstract

Zinner syndrome is a rare male genitourinary tract disorder associated with seminal vesicle cysts and ipsilateral renal agenesis. Clinical presentation often involves symptoms of the genitourinary tract. We present a case report of a young Saudi male, presenting with nonspecific symptoms of fatigue and malaise. Ultrasound visualized a massive seminal vesicle cyst associated with ipsilateral renal agenesis. The cyst was managed using a laparoscopic technique without any immediate complications and an uneventful postoperative period.

Keywords: Laparoscopic surgery, renal agenesis, seminal vesicle cyst, urology, Zinner syndrome

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from the seminal vesicle. Also noted was a solitary left kidney (14.7 cm × 7 cm × 3 cm) with compensatory hypertrophy, normal corticomedullary differentiation, and no hydronephrosis; the urinary bladder along with other internal abdominal structures was reported to be within normal limits [Figure 1]. The patient was referred to urology services where further imaging was requested.

Magnetic resonance imaging pelvis and abdomen confirmed the presence of complete agenesis of the right kidney associated with a retro-vesicle midline cyst with high-protein component likely related to seminal vesicle cyst with ejaculatory duct obstruction [Figures 2 and 3].

Cystoscopy demonstrated absent right hemitrigone, absent right ureteric orifice, normal left ureteric orifice, normal left hemitrigone, and normal urinary bladder. Retrograde pyelogram confirmed the absence of right ureter. The patient underwent laparoscopic resection of the seminal vesicle cyst under general anesthesia.

Histopathology of the excised cyst demonstrated a cyst lined by keratinized stratified squamous epithelium with focal papillomatosis. Also seen were areas of ulceration and benign squamous inclusion cysts. There was no dysplasia or signs of malignancy. The patient had an unremarkable postoperative period and was discharged without complications.

**DISCUSSION**

In 1914, Zinner reported the first case of seminal vesicle cyst associated with renal agenesis. Since then, only a few case reports have been published worldwide documenting this rare Müllerian duct abnormality. In 1990, Sheih et al. published a study where massive ultrasound screening for 280,000 children for renal anomalies was carried out to detect associations of cystic dilatations in the pelvis with renal agenesis. Of these children, only 13 were observed to have a combination of cystic dilatations in the pelvis with ipsilateral renal agenesis or dysplasia, of which only 6 were seminal vesicle cysts. The frequency of this rare phenomenon in the study was <0.004%.

Clinical presentation at the time of diagnosis for Zinner syndrome often involves lower urinary tract symptoms. Jarzemski et al. described a young patient presenting with lower abdominal pain, perineal pain, and dysuria. Sundar and Sundar and Haddock and Wagner described a patient with Zinner syndrome initially presenting with symptoms of painful ejaculation. Pavan et al. published a case where
the presentation was mimicking that of a varicocele. Alharbi et al.[9] described a case associated with peritoneal dialysis failure. On rare occasions, gastrointestinal symptoms have also been reported.[10]

Minimally invasive surgery involving laparoscopic or robotic assisted excision of the cyst appears to be the modality of choice for symptomatic patient and is associated with a favorable outcome and results in resolution of symptoms in the majority of the cases.[11]

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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