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Oddities

An Unexpected Clinical Feature of Zinner's Syndrome – A Case Report

Yu-Cing Juho, Sheng-Tang Wu, Shou-Hung Tang, Tai-Lung Cha, En Meng*

Division of Urology, Department of Surgery, Tri-Service General Hospital, National Defense Medical Center, 325 Cheng-Kung Road Section 2, Taipei 114, Taiwan, ROC

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ABSTRACT

We present a case of a healthy 43-year-old man who experienced right lower abdominal mass with gastrointestinal upset for 6 months. A series of imaging studies revealed a large lobulated cyst in the right pelvis and retroperitoneum. Because of the persistent symptom, surgical intervention was performed to remove the cystic lesion. The final pathology report demonstrated a large seminal vesicle cyst with agenesis of kidney. It is compatible with the diagnosis of Zinner's syndrome. However, right lower abdominal mass is a rare manifestation of this syndrome. This case reminds us a unique differential diagnosis of a right lower abdominal mass.

Introduction

Zinner's syndrome is a congenital malformation which includes seminal vesicle cyst, ejaculatory duct obstruction, and ipsilateral renal agenesia.[1] It is believed that an embryologic development abnormality of the Wolffian duct will induce ipsilateral renal agenesis and atresia of the ejaculatory duct.[2] The syndrome often occurs in the second and third decade of life and presents with voiding symptoms after beginning of sexual life.[3] The diagnostic tools include intravenous urography (IVU), trans-rectal ultrasonography (TRUS), magnetic resonance imaging (MRI) and cystoscopy. The principle of treatment is focusing on decreasing the mass effect of seminal vesicle cyst. The strategy comprises TRUS guided aspiration of seminal vesicle cyst, transurethral resection of the ejaculatory duct opening and surgical excision.[1] Choosing of treatment method depends on the patient's symptom mostly. In this paper, we present an unusual variant of this rare condition and describe the treatment strategy of this syndrome.

Case presentation

A 43-year-old male suffered from gastrointestinal upset for 6 months. Dry ejaculation and urinary frequency were also noted for 1 year. Two months before coming to our hospital, the patient found a palpable mass over right lower abdomen. He received intravenous urography (Fig. 1) which showed no contrast excretion from the right kidney and a filling defect appeared in the right lateral wall of bladder. CT scan of abdomen was performed and a large lobulated cystic lesion without contrast enhancement in the right pelvis and agenesis of right kidney were found.

He came to our hospital and the physical examination demonstrated a palpable, non-tender soft mass over the right lower abdomen. The digital rectal examination revealed normal size of prostate without firm nodule. MRI of pelvis (Fig. 2) was performed and showed a large lobulated tubular shape cystic lesion sized about 15 cm × 7 cm over the right pelvis and retroperitoneum. On the basis of history and image studies, an obstructive right seminal vesicle cyst with dilatation was suspected, but a right ureter tortuosity and dilatation due to UVJO could not be ruled out.

We performed the surgery of exploration with removal of the right pelvic cystic mass. The specimen (Fig. 3) contains a cystic lesion which connected with a small solid mass. The pathology is compatible with an enlarged seminal vesicle cyst and agenesis of the kidney. Hence, the diagnosis of Zinner's syndrome was confirmed.

Discussion

Zinner's syndrome is a congenital anomaly growth of the distal part of the Wolffian duct between the 4th and 13th gestational week.[3] The embryologic abnormal development results ipsilateral renal agenesis and atresia of the ejaculatory duct.[2] The syndrome was first described by Zinner since 1914.[1] In Taipei, 280,000 children underwent ultrasound screen and 13 cases of seminal vesicle cyst with ipsilateral renal agenesis were revealed. Therefore, the incidence on the basis of the paper was 0.0046%.[3]

* Corresponding author. Tel.: +886 2 8792 7169; fax: +886 2 8792 7172.
E-mail address: en.meng@gmail.com (E. Meng).

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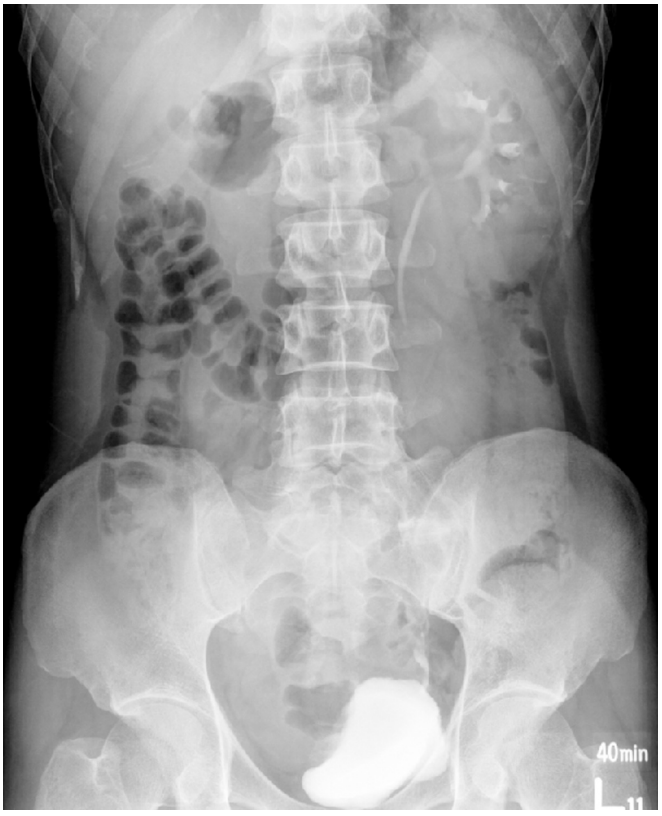

Figure 1. IVU showed no contrast excretion form right kidney and filling defect over right lateral wall of bladder.

The syndrome often occurs in the second and third decade of life and may present with voiding symptoms after beginning of sexual behavior.[3] Van den Ouden D analyzed 52 patients with Zinner's syndrome, and found that the most common symptoms include dysuria (37%), frequency (33%), perineal pain (29%), epididymitis (27%), pain following ejaculation (21%).[4] Our case presented with a right lower abdominal mass complicated with gastrointestinal upset which is a very rare manifestation of Zinner's syndrome.

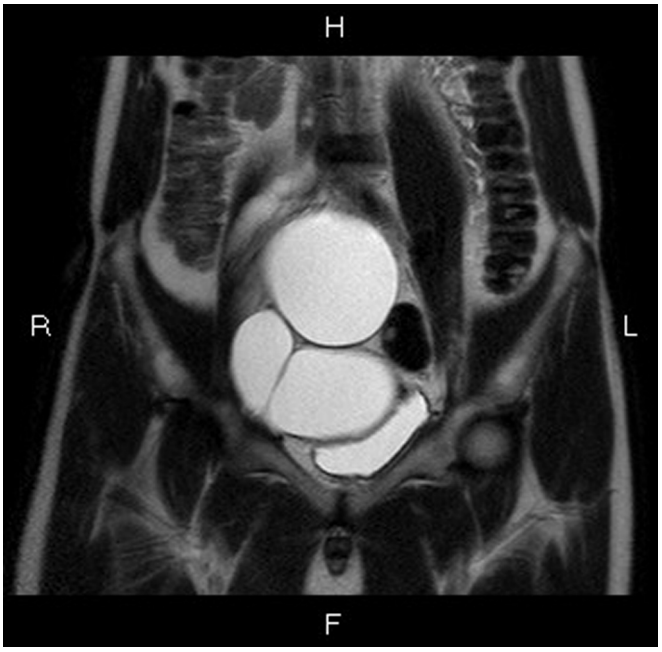

Figure 2. MRI of pelvis showed a cystic lesion within pelvic cavity which compressed the right border of bladder.

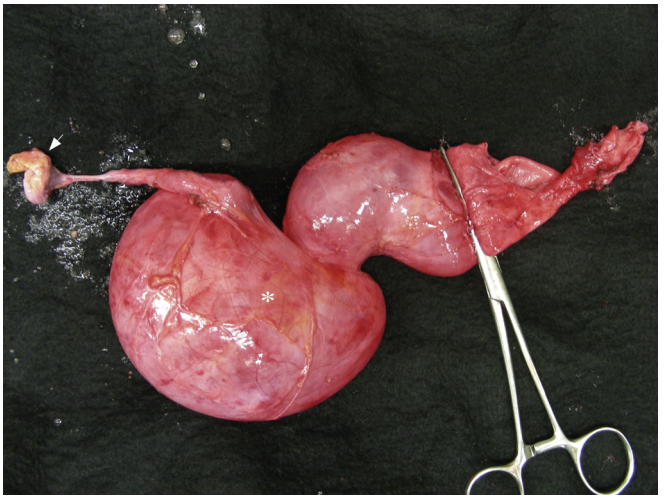

Figure 3. The surgical specimen composed of seminal vesicle cyst (asterisk) which connected to an atrophic kidney (arrow).

The diagnostic tools include IVU, TRUS, MRI and cystoscopy. IVU will show no contrast excretion and smooth filling defect of bladder over the lesion side. TRUS is the most wildly used tool for the seminal vesicle cyst evaluation and will revealed an anechoic cystic pelvic lesion in this syndrome. MRI not only provides a definite diagnosis but also demonstrates peripheral pelvic structure for surgical plan decision. Typical findings of seminal vesicle cysts are high signal intensity lesion on the T2-weighted image and low signal intensity lesion on the T1-weighted image.[5] Cystoscopy might show incomplete trigone with extrinsic compression due to the mass effect of a large seminal vesicle cyst.[2] Our case had a seminal vesicle cyst measured about 15 cm $\times$ 7 cm. So far as we know, it is the biggest seminal vesicle cyst which has ever been found in Zinner's syndrome.

If the symptom is mild, conservative treatment with antibiotic or transurethral needle aspiration of cyst is suitable for the patients. For symptomatic patient, invasive treatments including transurethral resection of the ejaculatory duct (TURED), exploration, laparoscopic and robotic vesiculectomy are suggested.[3] TURED is performed by removal of the proximal verumontanum until the opening of the ejaculatory duct with cutting current.[1] Exploration with removal of cystic lesion is still the most effective method. However, complication such as impotence, urinoma and pelvic organ injury should be considerated.[5] Therefore, minimal invasive surgery with laparoscopy and robotic surgery had been applied for removing the seminal vesicle cyst. Decrease of complication, blood loss and hospital day are the benefit of minimal invasive surgery.[3]

Exploration with removal of seminal vesicle cyst was performed for our patient due to the large retroperitoneal and pelvic cystic lesion. After the operation, the symptom subsided and no complication was noted during outpatient follow-up.

Conclusion

Zinner's syndrome is a rare congenital malformation which includes seminal vesicle cyst, ejaculatory duct obstruction, and

ipsilateral renal agenesia. We presented a 43-year-old man who suffered from a large lower abdominal mass and the diagnosis of Zinner's syndrome was confirmed after series of examinations. Surgical interventions successfully relieved his symptoms. This case reminds us that a middle-aged man with a large lower abdominal mass should be aware of the unique presentation of Zinner's syndrome.

Conflict of interest

The authors declare that they have no conflict of interest.

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