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## LETTER TO THE EDITOR

Male Health

# Zinner's syndrome: clinical features and imaging diagnosis

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Asian Journal of Andrology (2018) 20, 316–317; doi: 10.4103/aja.aja\_21\_17; published online: 11 July 2017

Dear Editor,

Zinner's syndrome 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.<sup>1</sup> After the initial report by Zinner in 1914,<sup>2</sup> fewer than 100 cases have been reported in literature.<sup>3</sup> On the basis of a children research in Taipei, the incidence of seminal vesicle cyst with ipsilateral renal agenesis was 0.0046% (13/280 000).<sup>1</sup> For symptomatic patients, an integrated and accurate diagnosis from radiologists can benefit clinicians' therapeutic decision-making. And for those young patients, finding the potential cause of obstructive azoospermia early can promote an early management. Therefore, our study aimed to summarize the computed tomography (CT) and magnetic resonance imaging (MRI) findings of this case series to draw attention of either urologists or radiologists, thus avoiding the misdiagnosis and missed diagnosis in clinics or even preventing the unnecessary invasive examination for this disease.

Between June 2006 and May 2015, the clinical records of 14 male patients (age range: 14–43 years, median: 25 years) with clinically confirmed Zinner's syndrome according to typical imaging appearance in our single center were retrospectively reviewed. Ethical permission was given by the Institutional Ethics Committee of the First Affiliated Hospital of Sun Yat-sen University, Guangzhou, China, and informed written consents were obtained from all participants. Among the 14 patients, abdominopelvic CT was performed with and without contrast media in 6 and 2 patients, respectively. Abdominopelvic contrast-enhanced MRI was performed in 12 patients. Two experienced radiologists reviewed the images together and recorded the appearance of the seminal vesicle cysts including location, number, largest diameter, margins (well defined or ill defined), shape (round, oval, or tubulosaccular), CT attenuation or magnetic resonance (MR) signal intensity of cystic content, presence of hemorrhage, and compression of adjacent urinary bladder, rectum, or prostate gland.

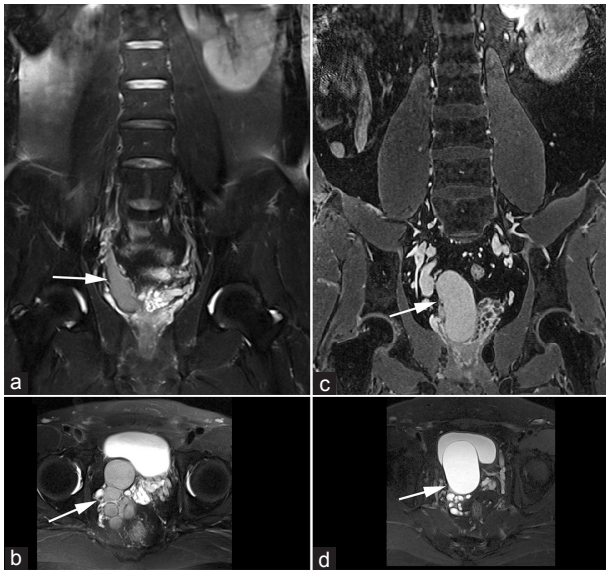
Symptoms included hemospermia ( $n = 1$ ), infertility ( $n = 2$ ), abdominal discomfort ( $n = 3$ ) or distention ( $n = 1$ ), and urinary frequency and dysuria ( $n = 7$ ). A bulging mass superior to the

prostate gland was found in all patients on digital rectal examination. Ultrasound showed seminal vesicle cysts associated with or without renal agenesis. Open exploration with unilateral vesiculectomy ( $n = 5$ ) or seminal vesiculoscopy ( $n = 6$ ) confirmed the seminal vesicle cysts, and partial decompression was performed. The remaining three patients with mild urinary complaints did not undergo surgery and the cysts were stable on follow-up ultrasound.

Abdominopelvic CT and/or MRI showed seminal vesicle cysts associated with ipsilateral renal agenesis in all the 14 patients with a 4:3 ratio of right-side predominance. All cysts arose from the seminal vesicles with an average cyst diameter of  $6.1 \pm 2.5$  cm (range: 4.2–7.1 cm) (Figure 1). The solitary cysts had irregular margins (9/14, 64.3%) or tubulosaccular shape (5/14, 35.7%). The cyst density was slightly greater than that of urine with an average CT value of  $8 \pm 5$  HU (range: 3–15 HU) and no contrast enhancement. On MRI, the cysts were T1 isointense or hyperintense and T2 hyperintense without contrast enhancement. Other features included fluid-fluid level in 4/14 cysts, compression on posterior aspect of the bladder ( $n = 9$ ), prostate gland ( $n = 3$ ), anterior lateral aspect of the rectum ( $n = 2$ ), and adjacent ureteric remnants ( $n = 6$ ).

Congenital seminal vesicle cysts are more common than secondary cysts and are associated with other congenital genitourinary anomalies, especially ipsilateral renal agenesis (67%–75%), because of their common embryologic origin.<sup>4</sup> Incomplete migration of ureteric bud can result in agenesis or dysplasia of the kidney. Developmental anomaly of the Wolffian duct affects the ipsilateral seminal vesicle, ejaculatory duct, vas deferens, and epididymis. The ejaculatory duct forms abnormally and congenital seminal vesicle cyst formation is believed to be secondary to obstruction of the duct. Similar to the meta-analysis by van den Ouden *et al.*,<sup>4</sup> we found right-sided Zinner's syndrome to be more common. The age of onset varies from 16 to 68 years and the cysts become apparent usually in the second to third decades with the greatest sexual or reproductive activity. Symptoms coincide with cyst enlargement from seminal fluid accumulation in the seminal vesicles.<sup>4</sup> Congenital seminal vesicle cyst with secondary infection has been reported in an infant.<sup>5</sup> In our study, the patients presented during adolescence ( $n = 3$ ) or period of exuberant sexual activity ( $n = 11$ ). Small cysts may remain asymptomatic whereas cysts larger than 5 cm usually result in bladder irritation and obstruction with dysuria, increased urinary frequency, perineal pain, epididymitis, ejaculatory and sacrococcygeal pain.<sup>1,4,6</sup> In this study, infertility (17%) and long-standing history of hemospermia up to 7 years ( $n = 1$ ) would have been caused by secondary inflammation of seminal

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Received: 28 April 2017; Accepted: 23 May 2017



**Figure 1:** A 43-year-old man with dysuria and a 34-year-old man with urinary frequency and nocturia. (a) Coronal T2-weighted image demonstrating the origin of the cyst from the right seminal vesicle with ureteric remnant (arrow). (b) Axial T2-weighted image showing a tortuous dilated tubulosaccular structure (arrow) with hyperintensities posterior to the urinary bladder on the right. (c) Contrast-enhanced coronal fat-saturated T1-weighted image showing that the cyst (arrow) does not enhance with intravenous contrast and is associated with right renal agenesis. (d) Axial T2-weighted image showing a large hyperintense proteinaceous cyst in the right seminal vesicle (arrow).

vesicle cysts.<sup>4</sup> For symptomatic patients, invasive treatments are recommended.<sup>7</sup> Seminal vesiculoscopy ( $n = 6$ ) or open exploration with vesiculectomy ( $n = 5$ ) was performed due to the compression on bladder and/or ejaculatory duct caused by the large pelvic cystic lesion.

Both CT and MRI can demonstrate the cysts with superior soft tissue resolution. MR with no radiation is superior to CT for delineating the genitourinary anomalies and demonstrating peripheral pelvic structure for surgical plan decision.<sup>8,9</sup> Simple seminal vesicle cysts without associated renal agenesis result from local expansion of the seminal vesicle tubes and are usually round or ovoid with regular margins. Seminal vesicle cysts of Zinner's syndrome derive from the remaining mesonephric duct and are usually larger, round, or tubulosaccular with irregular margins. In our study, 6/14 (42.9%) patients had ureteric remnant and is higher than the reported incidence of 27%,<sup>3,4</sup> suggesting that it is not an uncommon characteristic. The seminal vesicle cysts can be distinguished from other Wolffian and Müllerian duct cysts by the location, shape, and compression on adjacent structures. Seminal vesicle cysts are single or multiple, unilateral, surrounded by duct of seminal gland, and separated from the prostate by prostatic capsule. The cysts may be large, protrude into the posterior aspect of bladder, or compress the prostate and rectum.<sup>4</sup> Ejaculatory duct cysts are Wolffian duct cysts, usually unilateral, oval or tubulosaccular, and located at the level of the verumontanum posterior to the prostatic urethra. Ejaculatory duct cysts have posterolateral to anteromedial orientation in the axial plane and superolateral to inferomedial orientation in the coronal plane, rarely compressing the bladder and rectum. Müllerian duct cysts result from incomplete degradation of the Müllerian duct. It is located in the midline verumontanum area between the bilateral seminal vesicles and is posterior to the prostate. It is usually single and contains no spermatozoa. The cyst extends to the base of prostate and compresses the middle posterior aspect of bladder. It has typical inverted teardrop shape on sagittal or coronal

images with T1 hypointensity, T2 hyperintensity, and rarely intracystic hemorrhage.<sup>2</sup> Unilateral seminal vesicle agenesis with single kidney causing obstructive azoospermia is quite different from Zinner's syndrome. It is not palpable with physical examination and shows seminal vesicle agenesis on the image.

Zinner's syndrome should be considered when a tubulosaccular or a large rounded cystic lesion is found in a male pelvis, especially in seminal vesical region. Moreover, a further upper abdominal imaging examination should be performed to exclude the possibility of ipsilateral renal absence. Zinner's syndrome can be diagnosed when a remnant ureter is found in region adjacent to seminal vesical cyst, which is a unique sign found in six cases accounting for 42.9% of patients in our study (6/14), while a previous study has mentioned this only with few case reports.<sup>3</sup> Moreover, a coronal reconstruction image of abdominopelvic CT or a coronal MR scanning can benefit us to determine whether the cystic lesion is associated with Zinner's syndrome, which can help us avoid the misdiagnosis and missed diagnosis and further to provide the evidence for clinical management.

#### AUTHOR CONTRIBUTIONS

XSJ performed the clinical studies and wrote the paper. HJW collected the data and helped to organize the manuscript. JHL, YG, and CHS analyzed the data and interpreted the image. LL and JG guaranteed the integrity of the entire study and proposed study concepts and design. All authors read and approved the final manuscript.

#### COMPETING INTERESTS

All authors declare no competing interests.

#### ACKNOWLEDGMENTS

Our study was funded by Science and Technology Planning Project of Guangdong Province (2014A020212480) as well as Medical Science and Technology Research Fund of Guangdong Province (A2017008). The authors would like to thank Dr. Margaret H Pui for assistance with the manuscript polishing.

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