Case Report

Zinner syndrome: A first case report from Ethiopia

Erko Beyene, MDa,* , Engidawork Tadele, MDa, Meti Negassa, MDb

a School of medicine, College of Health Sciences, Addis Ababa University, Addis Ababa, Ethiopia
b School of Medicine, Myungsung Medical College, Addis Ababa, Ethiopia

A B S T R A C T

A triad of unilateral renal agenesis, ipsilateral seminal vesicle cyst and ipsilateral ejaculatory duct obstruction has been called Zinner syndrome since its first description in 1914 by Zinner. It is a very rare congenital abnormality of the male genital tract due to abnormal embryologic development of the Wolffian ducts. There have been several case reports from different parts of the world about the clinical and imaging findings of these anomaly. In this case report, we present the first case report of Zinner syndrome in a 28-year-old male Ethiopian patient.

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Background

Zinner is syndrome is a very rare congenital urogenital tract with reported incidence of 0.046% [1]. It consists of triads of unilateral renal agenesis, with ipsilateral seminal vesicle cyst and ipsilateral ejaculatory duct obstruction. It was first reported in 1914 by Zinner. It occurs due abnormality of distal mesonephric duct and ureteric buds during early embryonic period [2]. Patients usually present in the 2nd to 4th decade of life with nonspecific urinary symptoms, local perineal symptoms or abnormal reproductive function [3,4]. The symptoms include pain, dysuria, frequency, perineal pain, postejaculation pain etc [4]. Some patients also present with infertility with reported incidence of up to 45% [4].

Case presentation

We report a case of a 27-year-old male patient, father to 1-year-old girl, working as a merchant with nonspecific lower abdominal pain of 1 year duration. He has no history of fever, lower urinary tract symptoms, no perineal pain or no post ejaculation pain. He has history of hernia surgery. Physical examination showed well looking young man with no gross abnormality. The vital signs were pulse rate 76 bpm, Respiratory rate 20, Blood pressure 120/75 mm Hg and Axillary temperature 36.9°C. There was old healed inguinal scar over the right inguinal region. He had non icteric slera and pink conjunctiva. The respiratory system, the cardiovascular system, and the lymphoglandular systems examination were all unremarkable. On laboratory investigations, complete blood count,
serum creatinine, serum electrolytes, urine analysis, and seminal fluid analysis were done and they were all within normal range.

On imaging, abdominopelvic ultrasound (Figs. 1 and 2) and CT urography study (Figs. 3, 4 and 5) were done. The abdominopelvic ultrasound findings were non-visualized left kidney with compensatory hypertrophy of the contralateral right kidney measuring 13.2 × 6.9 cm. There was also dilated bilateral seminal vesicles with left side tortuously dilated seminal vesicles with cystic structure at its most superior part. The cyst has internal floating low level echo debris.

On CT, there was right side enlarged functioning kidney with non-visualized left kidney and ureters. There was positive left side flat adrenal sign. There was left side non enhancing cystic lesion posterior to the bladder with minimal mass effect on the bladder that measured 2.5 × 2.5 cm in size. The bilateral seminal vesicles also seem enlarged tortuous and tubular dilatation bilaterally. On post contrast study there was enhancing component in the cystic lesion, no solid component, no septa.

This patient was put on follow up for observation and no intervention was planned.

**Discussion**

Zinner syndrome is a rare congenital abnormality that is often asymptomatic but could present in 2nd and 3rd decades of life with nonspecific lower urinary and genital tract symptoms [3,4]. The reported incidence from one pooled study which reviewed 200 cases of ZS was 0.046% (3). Majority of patients become symptomatic in the 2nd to 3rd decades of life when sexual activity heightens [5]. Symptoms develop during the years of sexual activity, when secretions are at their peak or drainage is inadequate owing to a malformed or secondarily stenosed duct system [6].

Zinner syndrome occurs due to developmental anomaly of the Wolffian duct which is a precursor to the seminal vesicles and the vas deferens. It also gives rise to ureteric bud which explains the association of seminal vesicle pathologies with renal anomalies [7]. In our case the patient has unilateral renal agenesis with ipsilateral seminal vesicle cyst with tortuously dilated ipsilateral seminal vesicle indicative of ipsilateral ejaculatory duct obstruction. The patient also has minimal enlargement of the contralateral seminal vesicle. This
Fig. 3 – 3DVR CT urography image shows unilateral renal agenesis with non-visualized left ureter; the transverse pelvic scan of the abdomen with color Doppler interrogation showed the presence of isolated right side ureteric jet and non-visualization of the left ureteric jet which indicated absence of the left ureteric orifice.

Fig. 4 – Coronal and axial 12 minutes delayed CT urography images show unilateral left side renal agenesis with compensatory right renal enlargement measuring 13.1 x 6.9 cm in size, with no hydronephrosis. On the left side, flat adrenal sign can also be seen.

could be due to partial obstruction of the contralateral ejaculatory duct.

The differential diagnosis of cystic lesions of the male pelvis are classified into intraprostatic and extraprostatic lesions [6]. The extraprostatic lesions include the seminal vesicle cysts, the vas deference cyst and Cowper duct cyst. Because of the anatomic location of our patient’s lesions we did not consider Cowper duct cyst in our differential diagnosis. Vas deferens cyst are also another possible cystic lesion in this region of the male pelvis. They are usually located along the course of the vas deferens superior to the prostate. However patients with vas deferens cysts have azoospermia with low ejaculate volume on seminal fluid analysis [6]. In our patient the seminal fluid analysis parameters were all in normal range. Seminal vesicles cysts are most commonly found as isolated findings, however, rarely, they could be associated with polycystic kidney diseases or renal agenesis [6]. In our patient we have associated findings of ipsilateral renal agenesis and tortuously dilated bilateral seminal vesicles likely due to bilateral ejaculatory duct obstructions.

On imaging evaluation the preferred modality of choice to diagnose ZS is MRI [3]. Before MRI was widely available in the early 1980s CT scan with other imaging modalities was used as an accurate diagnostic method for seminal vesicle abnormalities [8,9]. The utility of ultrasound lied in its ability to show the cystic nature of seminal vesicle mass lesion detected on CT [9]. In our case we were able to demonstrate the absence of left kidney with CT urography finding and also left seminal vesicle region mass lesion which has high density fluid attenuation which was also demonstrated on the ultrasound to have
cystic nature with posterior acoustic enhancement and low-level floating debris which could represent the seminal fluid’s higher density as compared to urine. Even though MRI is the recommended imaging modality especially for surgical planning, in our patient the ultrasound and CT urography findings were enough to make final diagnosis. In a low resource setup where MRI is not available, high-resolution ultrasound with CT urography can be used for diagnosis of Zinner syndrome.

Pooled analysis of 200 cases of ZS showed that the most common management options for Zinner syndrome included surgery, Aspiration or Observation [3]. The types of surgery that was done was seminal vesiculectomy with different approaches including open surgery, laparoscopic, or robot assisted surgery [3]. Aspiration was the other management option but it is associated with high degree of recurrence [10]. In patients who presented with infertility as a presenting symptom the preferred management option of transurethral resection of the ejaculatory duct [10]. Small size seminal vesicle lesions with no symptoms can be observed with regular follow up. The majority of seminal vesicle cysts described in Zinner syndrome are benign in nature with only single reported case of malignant seminal vesicle in a 28-year-old patient [10,11]. The longest documented follow up for a known Zinner syndrome patient was for 17 years with 80% increment in size of the lesion over the course [10].

Conclusion

Zinner syndrome is rare cause of lower abdominal pain in young adult men. Ultrasound and CT urography can be used compositely to diagnose Zinner syndrome and careful evaluation of images is required as to avoid potential misdiagnoses. If surgery is planned for symptomatic patients, MRI can be helpful for planning. In small size and asymptomatic cases observation with follow up ultrasound is appropriate course of action as, even though rare, there are reported incidences of malignant transformations.

Patient consent

Informed consent to prepare and publish this case report was obtained from the patient. Personal identifiers were not used in the case report.

REFERENCES

[1] Sheih CP, Hung CS, Wei CF, Lin CY. Cystic dilatations within the pelvis in patients with ipsilateral renal agenesis or dysplasia. J Urol 1990;144(2 Pt 1):324–7.
[2] Mehr S, Ranjan R, Garga UC. Zinner syndrome—a rare developmental anomaly of the mesonephric duct diagnosed on magnetic resonance imaging. Radiol Case Rep 2016;11(4):313–17.
[3] Liu T, Li X, Huang L, Li H, Cai K, Jiang J, et al. Zinner syndrome: an updated pooled analysis based on 214 cases from 1999 to 2020: systematic review. 2021. 2021;10(2):2271–82.
[4] Di Paola V, Gigli R, Totaro A, Manfredi R. Zinner syndrome: two cases and review of the literature. BMJ Case Rep 2021;14(6):e243002.
[5] Regmi AR, Mishra S, Gurung A, Acharya A, Paneru LD, Ghimire A. Zinner’s syndrome: a rare diagnosis of congenital seminal vesicle cyst and renal agenesis on basis of radiological imaging and its management. Int J Surg Case Rep 2022;97:107434.
[6] Shebel HM, Farg HM, Kolokythas O, El-Diasty T. Cysts of the lower male genital tractary tract: embryologic and anatomic considerations and differentiation diagnosis. Radiogr Rev Publ Radiol Soc N Am Inc 2013;33(4):1125–43.
[7] Ghonge NP, Aggarwal B, Sahu AK. Zinner syndrome: A unique triad of mesonephric duct abnormalities as an unusual cause of urinary symptoms in late adolescence. Indian J Urol IJU J Urol Soc India 2010;26(3):444–7.
[8] Drolshagen L, Sandler MA. Congenital seminal vesicle cyst: a specific diagnosis by computed tomography. Henry Ford Hosp Med J 1984;32(2):131–3.
[9] Kenney PJ, Leeson MD. Congenital anomalies of the seminal vesicles: spectrum of computed tomographic findings. Radiology 1983;149(1):247–51.
[10] Kelly NP, Fuentes-Bonachera A, Shields WP, Cullen IM, Daly PJ. Long-term surveillance and laparoscopic management of Zinner syndrome. Case Rep Urol 2021;2021:6626511.

[11] Bhat A, Banerjee I, Kryvenko ON, Satyanarayana R. Primary seminal vesicle adenocarcinoma: a lethal yet cryptic malignancy with review of literature. BMJ Case Rep 2019;12(12):e232994. doi:10.1136/bcr-2019-232994.