One of the uncommon urogenital malformations in males, which presents late in the second or third decades of life, is the congenital malformation of the seminal vesicle. Zinner syndrome is a rare condition comprising a triad of unilateral renal agenesis, ipsilateral seminal vesicle obstruction, and ipsilateral ejaculatory duct obstruction. Very few cases in the literature describe the malignant change in seminal vesicle cyst in Zinner syndrome. To the best of our knowledge, this is the first report of a papillary neoplasm of the seminal vesicle cyst developed in a patient with Zinner syndrome.

**Keywords:** Papillary neoplasm in Zinner syndrome, seminal vesicle cyst with malignant transformation, unilateral renal agenesis, Zinner syndrome

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### Case Report

**Zinner Syndrome with Papillary Neoplasm: A Rare Case Report**

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### Introduction

Zinner syndrome was first described by Zinner in 1914 as the male equivalent of Mayer-Rokitansky-Kustner-Hauser (MRKH) syndrome described in females.¹ This case presents a unique variation of Zinner syndrome with papillary clear cell neoplasm of the seminal vesicle cyst that was successfully excised with open approach.

### Case Report

A 17-year-old boy was referred to our tertiary care pediatric surgery department with complaints of recurrent episodes of colicky pain in the lower abdomen, with associated dysuria and constipation for 6 months duration. The patient underwent noncontrast computerized tomography of the abdomen elsewhere on presentation which revealed a well-defined midline hypodense mass compressing right ureter with moderate hydroureteronephrosis and absent left kidney. At our center, the patient was further investigated with contrast-enhanced magnetic resonance imaging of the abdomen which revealed midline cystic lesion measuring 11 cm × 10.5 cm × 11.3 cm in the rectovesical pouch region. Dilated right ureter with hydronephrotic kidney was noticed. The left kidney was not visualized [Figure 1]. In view of compressive symptoms, the patient was taken up for surgery. Midline laparotomy was performed which revealed 15 cm × 15 cm thick-walled cystic mass in rectovesical pouch-containing amber-colored fluid with few tissue components inside [Figure 2]. The cyst was densely adhered to base of the urinary bladder anteriorly, and laterally, the mass was extending up till bilateral iliac vessels. The frozen section was done from the cyst wall and cystic contents which was confirmed as benign seminal vesicle cyst. In view of single kidney, close proximity to vital pelvic structures, younger age with dense adhesion of cyst to the urinary bladder anteriorly and with benign frozen section report, the cyst was removed subtotally and marsupialization of the remnant cyst wall was done. The cyst wall along with its contents was sent for the histopathological examination which revealed Wolffian duct remnant cyst-recto vesical neoplasm of papillary variant.

On follow-up on the 6th month, contrast-enhanced computed tomography (CT) scan of the abdomen was done, which revealed remnant cyst measuring about 4 cm × 6 cm with papillary projections in the cyst wall which enhanced on positron emission tomography scan. After further assessment with contrast-enhanced magnetic resonance imaging (CE MRI), we planned...
to do a completion surgery to avoid overt malignant change in the cyst. The midline cystic neoplastic mass was identified and was removed en mass preserving right ureter. Follow-up imaging with CE MRI abdomen was done a month after surgery which revealed no recurrence/collection. The patient was followed up after 06 months with ultrasound pelvis which revealed no collection/recurrence. The patient is presently on 06 monthly follow-up with ultrasound pelvis and yearly CE MRI of abdomen and pelvis.

DISCUSSION

The prevalence of congenital genitourinary abnormalities is estimated to be <1%. In one study in Taipei, the incidence of seminal vesicles with ipsilateral renal dysplasia was found to be 0.0046% (13/28,000). The patients are usually diagnosed in the 2nd-3rd decade of life and present with symptoms of dysuria (37%), frequency (33%), perineal pain (29%), and epididymitis (27%). The condition comes to the notice of the clinician in the second to fourth decade when patients present with symptoms due to compression or due to infertility. CT imaging enables accurate diagnosis and is better than sonography, findings are of a retrovesicular periprostatic cystic mass along with ipsilateral renal agenesis. MRKH syndrome, the female counterpart of Zinner syndrome, is reportedly associated with ovarian cancer and renal cell carcinoma in the genitourinary tract. Similarly, two cases of adenocarcinoma arising from a seminal vesicle cyst in Zinner syndrome patients have been previously reported in the literature.

Our case indicates that rational imaging, well timed surgery and long-term observation are mandatory for patients with Zinner syndrome presenting with nonspecific but rapid and progressive urogenital symptoms. In case of large-sized cyst with symptomatic presentation, complete excision of the cyst either open or laparoscopic or robotic-assisted laparoscopy will be the gold standard treatment.

CONCLUSION

The high index of suspicion should be raised if a young male patient presents with nonspecific symptoms of recurrent lower urinary tract or lower abdominal symptoms with unilateral renal agenesis. If the patient is symptomatic with cyst being larger in size, we recommend complete excision and further follow-up according to the histopathological examination.

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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