Case Report

# Incidental detection of Zinner syndrome in a patient with nonseminomatous germ cell tumor of testis

Jeevitesh Khoda, Saugata Sen, Argha Chatterjee

Department of Radiology, Tata Medical Center, Kolkata, West Bengal, India

**Abstract** Zinner syndrome is a rare congenital abnormality occurring in males comprising a triad of unilateral renal agenesis, ipsilateral ejaculatory duct obstruction, and seminal vesicle cyst. Most patients remain asymptomatic, and some may present with lower urinary tract symptoms or infertility. We present a case of incidentally detected Zinner syndrome in a patient with nonseminomatous germ cell tumor of testis, an association that is not reported in literature to our knowledge.

**Keywords:** Mesonephric duct, Mullerian aplasia, testicular germ cell tumor, Zinner syndrome

**Address for correspondence:** Dr. Argha Chatterjee, Department of Radiology, Tata Medical Center, 14 Main Arterial Road (E-W), New Town, Action Area, Kolkata - 700 156, West Bengal, India.
E-mail: arghachat84@gmail.com
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## INTRODUCTION

Zinner syndrome is a rare congenital abnormality occurring in males comprising of a triad of unilateral renal agenesis, ipsilateral ejaculatory duct obstruction and seminal vesicle cyst. In this report, we present a case of incidentally detected Zinner syndrome in a patient with non-seminomatous germ cell tumour of testis.

A 37-year-old male patient presented with right-sided scrotal swelling. Ultrasound and high inguinal orchiectomy done outside showed a right testicular mass. Histopathology examination revealed nonseminomatous germ cell tumor. Complete para-aortic lymphadenectomy was done, and then, the patient was referred to our center for further management. The patient was asymptomatic. On examination, the external genitalia were significant for surgical scar. Blood investigations were within normal limits including serum alpha-fetoprotein and beta-human chorionic gonadotropin. The patient did not have any significant past medical history.

Contrast-enhanced computed tomography (CECT) thorax and abdomen was performed for baseline metastatic workup before starting chemotherapy. On CECT, the right kidney was not visualized. A solitary left kidney was seen in the left renal fossa that was supplied by multiple left renal arteries arising from the aorta and left common iliac artery and drained by multiple renal veins [Figure 1]. Single left renal moiety and left ureter ending normally in the left vesicoureteric junction with normal contrast excretion were identified. The right seminal vesicle was enlarged and multicystic in appearance. The left seminal vesicle was normal. An elongated blind-ended cystic structure was seen extending from the seminal vesicle cyst at the level of prostate and seminal vesicle to the level of the aortic bifurcation above passing anterior and medial to common iliac vessels [Figure 2]. This structure was presumed to be

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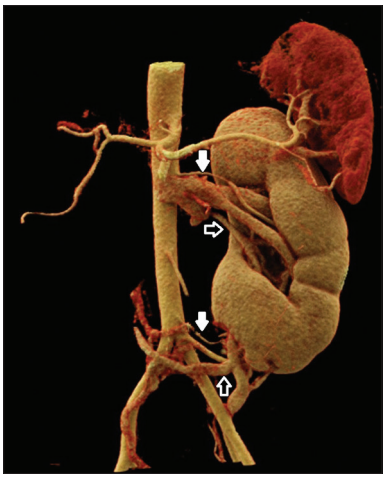

**Figure 1:** Cinematic volume rendering arterial phase computed tomography of the kidneys showing enlarged solitary left kidney supplied by arteries arising from the aorta at multiple levels (solid arrows) and drained by multiple veins (open arrow). Notice absence of the right kidney and right renal artery. Branches of the superior mesenteric artery are removed for better visualization

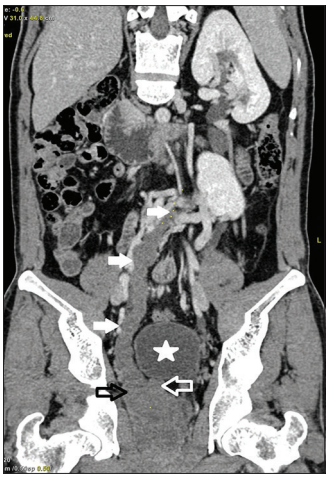

**Figure 2:** Curved multiplanar reformation of the nephrographic phase of computed tomography of the kidneys and urinary bladder (asterisk) showing a cystic attenuation tubular structure (solid arrows) extending superiorly from the multicystic seminal vesicle (between open arrows) below to end as a blind-ended tube at the midline at the level of the aortic bifurcation. The urinary bladder is marked by an asterisk

ectopic remnant of the right ureter. No lymphadenopathy was identified.

In view of the constellation of findings, a diagnosis of Zinner syndrome was made. The patient is currently receiving bleomycin-etoposide-carboplatin chemotherapy and on routine follow up.

## DISCUSSION

Zinner syndrome is an extremely rare congenital abnormality of distal mesonephric duct with reported incidence as low as 0.0021%.[1] It is considered to be the male counterpart of Mayer–Rokitansky–Küster–Hauser syndrome seen in women and consists of seminal vesicle cyst, ipsilateral ejaculatory duct obstruction, and unilateral renal agenesis. Mesonephric ducts are paired structures that develop into hemitrigone of the urinary bladder, proximal urethra, seminal vesicle, and vas deferens. An obstruction at the level of ejaculatory duct may lead to seminal vesicle cyst formation. An insult before 7 weeks of gestation before the appearance of the ureteric bud interferes with renal development leading to renal agenesis or dysplastic kidney. A ureteric remnant or ureterocele may be present. It is often an incidental finding. However, large cysts may cause obstructive symptoms.[2] The most common presentations include lower urinary tract symptoms and infertility in the second or third decade of life.[3] Ultrasound and cross-sectional imaging such as CECT abdomen are the imaging modalities of choice in such cystic pelvic lesions in men. Transurethral drainage and deroofing of cyst give good result in symptomatic cases.[3] Rare associations with primary seminal vesicle adenocarcinoma and renal cell-type tumor of the prostate are reported in literature.[4,5] However, to our knowledge, no association with testicular germ cell tumor is reported before this case. Considering the need for retroperitoneal nodal surgery in such tumors, knowledge of this anomaly is beneficial in surgical planning.

### 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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Nil.

### Conflicts of interest

There are no conflicts of interest.

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