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ZINNER'S SYNDROME: A CASE REPORT WITH TYPICAL RADIOLOGICAL FINDINGS

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ABSTRACT

Zinner syndrome is a rare anomaly of the urogenital tract. The syndrome is related to delay in the embryogenesis of the ureteric buds and is associated with unilateral renal absence, cyst formation in the seminal vesicles, and ipsilateral obstruction of the ejaculatory.

KEYWORDS

Zinner syndrome, ultrasound, magnetic resonance imaging

LEARNING POINTS

- Zinner syndrome is a rare syndrome with delayed embryogenesis of the ureteric buds.
- Three features are typically present, absence of one kidney, cyst formation of the seminal vesicles, and ejaculatory duct obstruction at the same side.

INTRODUCTION

Zinner syndrome (ZS) is a rare congenital urological anomaly[1-3]. From the embryologic point of view, it is associated with failure in the connection of the incompletely migrated ureteric buds with the metanephros leading to renal agenesis. The seminal vesicle continues to develop, however, associated with dysfunction of drainage, cyst formation, and ipsilateral dilatation of the seminal vesicles and ejaculatory duct[4]. Most of the patients are asymptomatic, the rest complain of frequency of urination, dysuria, symptoms related to ejaculatory duct disturbance, and occasionally haemospermia and haematuria[5]. Patients may be asymptomatic until the third/fourth decades of life and symptoms usually appear at the age of increased sexual activity.

CASE PRESENTATION

A 22-year-old male presented with dysuria, repeated urinary tract infection, and suspected urinary tract obstruction. Urine and semen analysis were normal. Examination of the external genitalia revealed a mildly thickened right spermatic cord. Both testes and the left spermatic cord were unremarkable. Ultrasound of the abdomen was ordered

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revealing absence of the right kidney. Abdominal ultrasound revealed a well-defined rounded anechoic lesion in right periprostatic region. Further evaluation of anechoic lesion by transrectal ultrasound confirmed a large cyst measuring 25 mm in the region of right seminal vesicle. The normal right seminal vesicle could not be well delineated. There was significant compression over the right ejaculatory duct, which was not identified. The distal part of vas deferens was also compressed. The vas deferens was dilated all along its course in spermatic cord. Computed tomography (CT) and nuclear scan confirmed absence of the right kidney. Magnetic resonance imaging (MRI) revealed seminal vesicle cyst appearing hypointense on T1-weighted images and hyperintense on T2-weighted images. Presence of protein-rich contents or a previous episode of intracystic haemorrhage may lead to hyperintense signal on T1-weighted images and hypointense signal on T2-weighted images. Diagnosis of ZS was made based on the imaging findings.

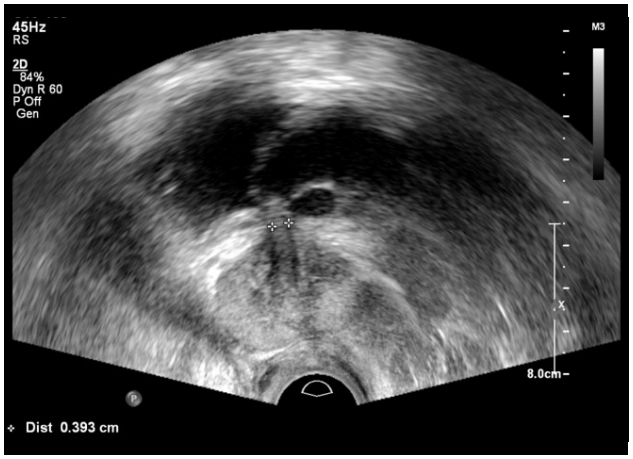


Figure 1. Transrectal ultrasound revealing a cystic lesion in the right peri-prostatic region related to the right seminal vesicles.

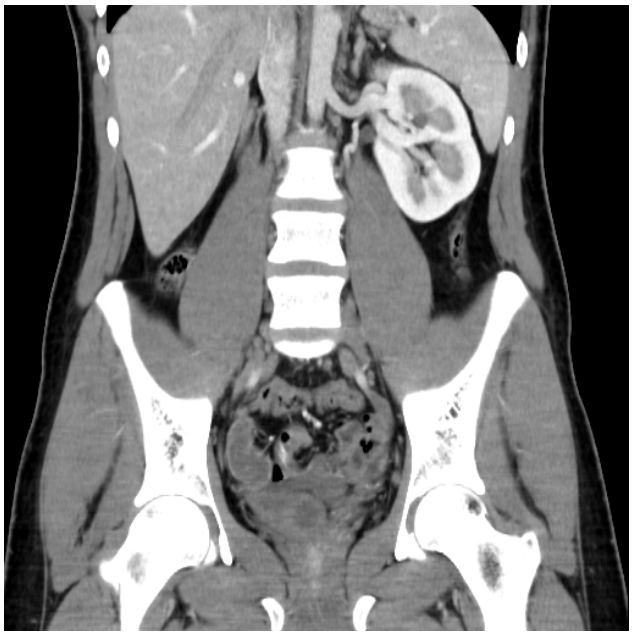


Figure 2. Coronal non-contrast computed tomography scan of the abdomen showing non-visualized right kidney.

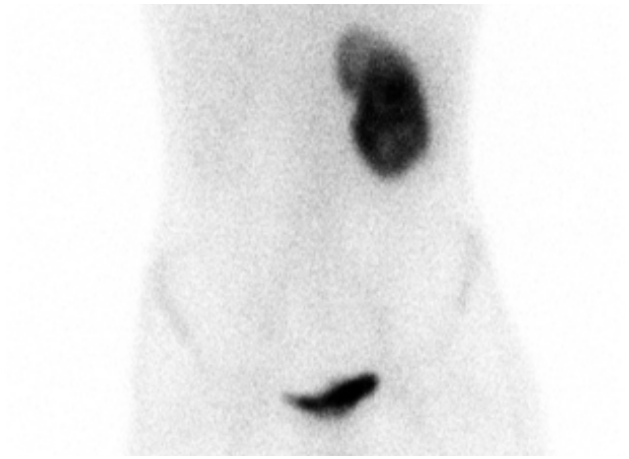


Figure 3. Coronal nuclear scan study confirming absence of the right kidney.



Figure 4. Axial T2 weighted magnetic resonance imaging scan showing a cyst connected to the right seminal vesicles by a convoluted tail.

DISCUSSION

Zinner syndrome (ZS) is a rare entity of the urogenital tract. This congenital malformation was first described in 1914[6]. It is the male counterpart of the Mayer-Rokitansky-Kuster-Hauser syndrome. ZS is characterized by ejaculatory duct atresia, seminal vesicle dilatation with or without cyst formation, and ipsilateral renal agenesis[7]. ZS can remain asymptomatic until periods of increased sexual activity which usually in the second decade of life[8]. The diagnosis of ZS needs multidisciplinary cooperation and a high index of suspicion[9]. Normally, in males both testosterone and anti-müllerian hormone help the mesonephric duct to give rise to the bladder neck and 50 % of the trigone epididymis, vas deferens, seminal vesicles and urethra. Any insult during the process of embryogenesis could influence the formation of different parts of the urogenital system[6]. Diagnostic imaging is crucial for the diagnosis of ZS. Abdominal ultrasound

evaluates both kidneys, urinary bladder and its relations. Transrectal ultrasound shows the prostate, seminal vesicles, vas deferens, and ejaculatory ducts. CT scan reveals kidneys status, and tracks both ureters. Nuclear scan study reveals absence of the kidneys. Nowadays MRI is considered the gold standard for diagnosis with excellent soft tissue contrast and multi-planar capability, in addition to accurate diagnosis of seminal vesicle cyst formation with ability to demonstrate the nature of the contents. MRI has been shown to be accurate in differentiating seminal vesicle cysts from other pelvic cystic malformations. Presence of a convoluted tail connecting the cystic abnormality to the seminal vesicle and the presence of fluid of high signal intensity on T1-weighted images favour the seminal vesicular origin of the cystic lesion. MRI remains the gold standard for the diagnosis confirming the presence and nature of the content of seminal vesicle cysts by different sequences.

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