CASE REPORT

Zinner's Syndrome: A New Asymptomatic Case Report

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ABSTRACT

Zinner's syndrome is a rare disease characterized by the presentation of the triad: Unilateral renal agenesis, ipsilateral seminal vesicle cyst, and ipsilateral ejaculatory duct obstruction. Small testis and ipsilateral ureterocele have also been reported. In most cases, patients are symptomatic and when diagnosed they require surgical treatment. We present a new case of asymptomatic Zinner syndrome that was diagnosed by chance in a 47-year-old man who consulted for vasectomy and who presented enuresis. In this case, conservative treatment and follow-up were decided.

Key words: Renal agenesis, seminal vesicle cyst, ureterocele, Zinner's syndrome

CASE REPORT

We present a 47-year-old male with no known drug allergies. His pathological history included left inguinal herniorrhaphy, circumcision, and sinusitis. He went to the urology outpatient clinic to request a vasectomy, being father of a child. In the anamnesis, he commented that on occasions, he had had enuresis. The vasectomy surgical technique and possible side effects were explained and he understood and accepted it.

On the other hand, and due to the presence of enuresis, we decided to perform the following complementary tests.

Blood test: Normal (glomerular filtration rate >90 ml/min, prostate-specific antigen 0.59 ngr/ml, and total testosterone 13.2 nmol/l). Basic urine test: Normal. Urine culture: Negative.

Urinary ultrasound: Absence of the left kidney. The right kidney with normal morphology and corticomedullary relationship that presented vicarious hypertrophy of 146.7 mm in size, with no evidence of tumor, significant cystic lesions, or nephrolithiasis. Urinary tract ectasia was not appreciated. Bladder presented some irregular area of mucosa at the left ureterovesical junction [Figure 1] that continued with a tubular cystic image of 30 × 17.8 mm in size in a extravesical location [Figures 2a and b]. There was no post-void residue and the size of the prostate was 25 cc.

Abdominal and pelvic magnetic resonance imaging (MRI): The absence of the left kidney and the compensatory hypertrophy of the right kidney were confirmed. The right kidney had no alterations. We diagnosed the presence of an elongated structure corresponding to the left ureter without associated renal parenchyma [Figure 3] that ended in an ectopic ureterocele [Figures 4a and b], in the left ejaculatory duct at the intraprostatic gland [Figure 5], conditioning a moderate ectasia of the left seminal vesicle. Both the ureterocele and the seminal vesicle on the left side contained hypertensive fluid in the T1 sequence of the MRI, corresponding to blood or hyperprotein material. The prostate gland and the pelvic structures did not present alterations.

These findings corresponded to congenital urinary malformations [Figure 6], in this case in a 47-year-old patient without the presence of associated symptoms. Therefore, we decided to carry out annual monitoring. Regarding enuresis, medical treatment was not advised because it was occasional and did not affect his quality of life.

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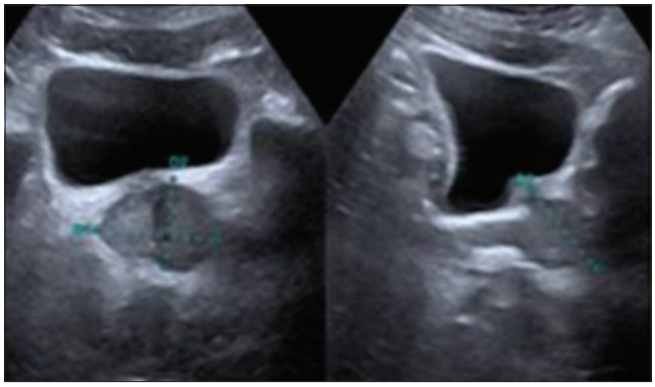

Figure 1: Irregular area of mucosa at the left ureterovesical junction

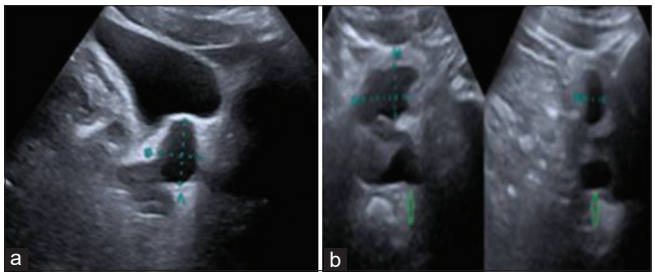

Figure 2: (a and b) Tubular cystic image of 30 × 17.8 mm in size in a extravesical location

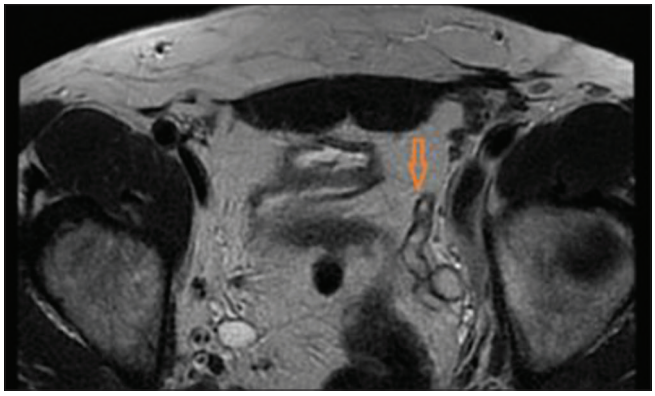

Figure 3: Elongated structure corresponding to the left ureter without associated renal parenchyma

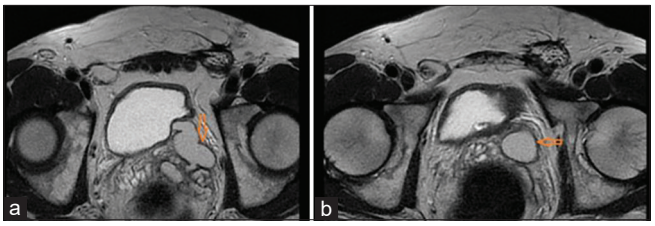

Figure 4: (a and b) The left ureter ending in an ectopic ureterocele

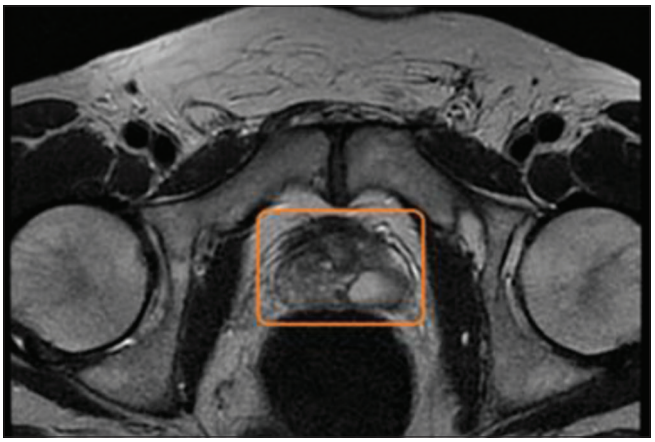

Figure 5: The presence of ureterocele ending in the left ejaculatory duct at the intraprostatic gland

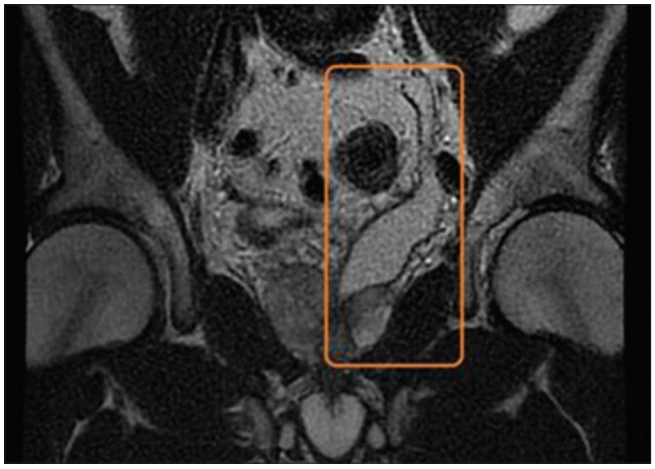

Figure 6: Congenital urinary malformation corresponding to the left ureter without renal parenchyma ending in an ectopic ureterocele at the ipsilateral ejaculatory conduit within the prostate gland

DISCUSSION

Renal agenesis is defined as the absence of the renal parenchyma and is caused by an embryological defect during metanephric development, appearing in 1 out of every 2900 births. Unilateral renal agenesis is estimated up to 5% of all renal malformations. The cause can be genetic but it can also be due to the teratogenic effect of cocaine or retinoic acid. It is frequently associated with the presence of vesicoureteral reflux. On the other hand, it has been associated with pyeloureteral and ureterovesical stenosis, with bicornuate uterus and with vaginal duplication. The differential diagnosis must be made with ectopic pelvic kidney, with crossed renal ectopia, and with horseshoe kidney.[1-3]

The presence of renal agenesis and ipsilateral cystic structure of the seminal vesicle was first described by Zinner in 1914 and is known as Zinner syndrome.[4,5] Patients are typically diagnosed at the third or the fourth decade of life and the most frequent symptoms are discomfort or pain in the perineum

and recurrent epididymitis even with the presence of abscess. Pain with ejaculation and infertility can also appear.[6,7]

The definitive diagnosis is made with MRI. Cystography can also be useful in indeterminate cases to rule out the presence of reflux. Most cases are symptomatic and require surgical treatment. The laparoscopic or robotic approach is recommended whenever possible.[8-11]

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