Sindrome de Zinner. A propósito de un caso clínico.


1er Ten “ec” (E. Med) Mariana Gay*, 1er Ten “ec” (E. Med) Luciana R. Peralta Davila*, Dr. Gustavo L. Decicco**

Workplace: Hospital Aeronáutico Córdoba, 479 Colón Ave., City of Córdoba

* Diagnostic Imaging Service Resident. Hospital Aeronáutico of Córdoba.
** Diagnostic Imaging Service Physician. Hospital Aeronáutico of Córdoba.

Abstract

Introduction: Zinner's syndrome is characterized by unilateral cystic dilatation of the seminal vesicle and ipsilateral renal atrophy or agenesis, which is the result of a Wolffian ducts congenital abnormality.

Objectives: Presentation of a clinical case and literature review.

Case Report: 18-year-old adolescent who complains of vaguely localized, dull, intermittent, testicular pain, accompanied by ejaculated seminal fluid volume reduction. We performed an abdominal ultrasound, which shows right renal agenesis and right seminal vesicle. With the clinical and image findings, Zinner's syndrome is diagnosed.

Discussion: Most patients presenting this group of mesonephric duct abnormalities are asymptomatic until their thirties or forties. Ultrasound is the initial diagnostic method due to its accessibility and it is useful for ruling out other causes of pelvic pain. It shows renal agenesis and pelvis cystic image. MRI is the method of choice to assess mesonephric ducts abnormalities.

Keywords: Zinner syndrome. Renal agenesis.

Received: 29th October, 2012. Accepted: 20th November, 2012.

Introduction

The described clinical case involves a 18-year-old adolescent who presented one year of vaguely localized, dull, intermittent, testicular pain, accompanied by ejaculated seminal fluid volume reduction noted by the patient. He brings 2 previous testicular ultrasound scans (performed in
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another institution) to the consultation which show no significant discoveries. Physical examination was unremarkable. CBC, serum biochemistry, coagulation profile test and urine test are ordered which show results within normal limits. Semen analysis showed an ejaculation volume lower than 1ml, sperm count of 400,000/ml, alkaline pH, and fructose 1.1g/l. Luteinizing Hormone (LH) analysis, follicle-stimulating hormone (FSH), and testosterone analysis results fell within normal parameters.

An abdominal ultrasound scan is performed which shows right kidney absent in renal fossa or in pelvis, with a right renal agenesis diagnosis. Left kidney presented normal characteristics and location.

Right renal fossa, absent right kidney; normal left kidney

It also describes a well-defined, anechoic round lesion with regular walls, located in the right periprostatic region and accompanied by multiple, satellite, smaller anechoic images, all with posterior wall reinforcement which indicates their cystic nature.

Prostate with peripheral anechoic image

Right seminal vesicle was not visualized. Both testicles were normal.

A pelvis, inguinal region and scrotum MRI was also performed for a more accurate anatomical evaluation. Right renal agenesis was confirmed.

The cyst content showed high signal intensity on both T1 and T2-weighted images, dismissing the presence of solid or hemorrhagic contents. It also showed the continuity between the proximal seminal vesicle dilatation and the right-region seminal vesicle cyst, and reported that the cystic mass does not exert mass effect on the adjacent structures, showing a good plane of separation among neighboring organs.
Abdomen coronal section in T2 w/gadolinium

Pelvis sagittal section T1 w/gadolinium

Pelvis axial sections in T2

Considering the previous clinical and imaging findings, ZINNER’S SYNDROME is diagnosed.

Discussion

This syndrome, characterized by unilateral cystic dilatation of the seminal vesicle, ureter with ectopic anastomosis and ipsilateral renal atrophy or agenesis, was first described by Zinner in 1914. It involves a congenital alteration of the Wolffian ducts that takes place when this embryological structure is affected by an injury before the 7th week of the pregnancy.

0.1% of newborns present unilateral renal agenesis; whereas 60-80% of patients with seminal vesicle cysts present ipsilateral renal agenesis. It is a rare congenital pathology that affects less than 0.003 % of the population. Only approximately 100 Zinner’s syndrome cases have been reported worldwide so far.

It can be accompanied by other genitourinary alterations: absent ureter, duplicated collecting system, ectopic ureter, trigone alterations, testicular ectopia, and remaining ureteric bud.

From an embryological point of view, the genitourinary system develops from a mesodermic fold called intermediate mesoderm, which originates the 3 renal systems (consecutive and overlapping): the pronephros, the mesonephros and the metanephros.

The pronephros appears by the 3rd week of the pregnancy and forms a common collecting duct or pronephros duct; it atrophies by the 4th week of the pregnancy. The mesonephros appears during the 4th week and forms the Bowman’s capsule and to the longitudinal collecting duct known as the mesonephric or Wolffian duct (which comes from the pronephros duct), and by the 10th week it atrophies, except in males where the causal end of the Wolffian duct remains and then forms the appendix of the epididymis, paradidymis, epididymis, deferential duct, ejaculatory duct, the seminal vesicle, and the bladder hemitrigone.
By the beginning of the 5th week, the 3rd system appears, the *metanephros* or permanent kidney; the ureteric bud appears (mesonephric duct outgrowth) which penetrates the metanephric tissue and it forms the collecting system when it dilates (ureter, renal pelvis, major and minor calyces and from 1 to 3 million of collecting tubules).

In turn, each collecting tubule is covered by metanephric tissue, and differentiates into glomeruli and forms the nephron or excretory unit. The definitive kidney starts working since the 12th week of the pregnancy.

A harmful agent during the 1st trimester negatively affects kidney embryogenesis, ureter, seminal vesicle and deferential duct. A disorder in the development of the mesonephric duct distal region leads to the ejaculatory duct atresia (to the seminal vesicle obstruction and cystic dilatation) and to an ureter mislocation (which results in renal dysplasia or agenesis). The obstruction at the ejaculatory duct level leads to the gradual accumulation of seminal vesicle secretions and the subsequent formation of cysts.

Most patients presenting this group of mesonephric duct abnormalities are asymptomatic until their thirties or forties and they are frequently manifested during high reproductive or sexual activity periods. Patients express pelvic, perineal or testicular pain, dysuria, painful ejaculation, repeated lower urinary tract infection, recurring prostatitis or epididymitis and infertility.

Ultrasound is the initial diagnostic method due to its accessibility and it is useful for ruling out other causes of pelvic pain. It shows renal agenesis and the pelvis cystic image. MRI is the method of choice to assess mesonephric ducts abnormalities since it makes it possible to determine the exact anatomic relation and the dependence of a pelvis cystic mass, as well as to show the cystic nature of lesions with content as regards pelvic pathology, its multiple plane imaging capacity, the excellent resolution of soft tissue details, and the use of non ionising radiation.

**DIFFERENTIAL DIAGNOSIS FOR CYSTIC MASSES IN MALE PELVIS**

- Müllerian duct cyst
- Deferential duct dilatation
Prostate cyst

Hydronephrosis

Ureteral duplication
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In conclusion, differential diagnosis are:
- cysts
- hydronephrosis
- bladder diverticulum
- ureterocele
- ureteral duplication
- Zinner’s syndrome

Conservative management is indicated for asymptomatic patients; otherwise, laparoscopic surgical resolution is indicated.

**References**