Zinner syndrome – case report

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Abstract

We present the case of a 51-year-old male with Zinner syndrome, which is a rare disease, resulting from an abnormal evolution of the mesonephric (Wolffian) duct. It consists in cystic dilations of one seminal vesicle and/or ejaculatory duct and ipsilateral renal agenesis. It leads to symptoms related to urination, ejaculation, even infertility, and to low-abdomen and perineal pain. The diagnosis is set by ultrasonography, CT scan and, mainly, MRI. Usually it is treated conservatively, but certain cases require surgery, nowadays minimally invasive.

Keywords: rare disease, seminal vesicles, renal agenesis

Introduction

Zinner syndrome is a rare disease, affecting males. It is secondary to an abnormal evolution of the mesonephric (Wolffian) duct during embryogenesis. The ureteric bud fails to migrate and join with the metanephros to form the kidney. On the caudal end of the mesonephric duct malformations also occur [1]. The syndrome is characterized by atresia of the ejaculatory duct, leading to its dilation and/or seminal vesicle cysts, associated with ipsilateral renal agenesis [1,2]. Clinically it is generally manifested by ejaculatory disturbances, local pain, and infertility, but it can display a wide range of presentations [1,2]. The diagnosis may be set by ultrasonography or CT scan, but MRI gives a complete image of the local anatomy [3]. It may be treated conservatively, but most of the cases require surgery, nowadays minimally invasive [4-6]. Zinner syndrome may lead to infectious complications, such as seminal vesicle abscess, or it may be associated with other developmental abnormalities or tumors [1].

Case report

A 51-year-old patient, without any relevant medical past, presented to the emergency department of a general hospital for dysuria, hypogastric pain, perineal and, occasionally, left testicular pain. Physical examination revealed a normal body weight, normal vital constants, no significant modifications of the cardiovascular and respiratory systems. There was slight tenderness of the hypogastrium and the rectal examination was very painful, making the assessment of the prostate impossible. The testicles were normal, without tenderness. The laboratory tests showed no abnormality. Urinary infection was ruled out.

The abdominal ultrasonography revealed a cystic lesion of the prostate, on a transverse section (Figure 1). When turning the transducer to a sagittal section, to measure the supposed prostatic cyst, the surprise was to find out that the cyst was communicating posteriorly with a duct-like structure (Figure 2). We suspected a dilation of the ejaculatory duct in its terminal segment, pre- and intraprostatic. The left kidney was not found in its normal, lumbar position. The pelvis was scanned, but no structure compatible with an ectopic kidney was found.

The next day, after anti-inflammatory treatment, the patient was in less pain and intrarectal ultrasonography was possible, but it did not bring any supplementary elements (Figure 3).

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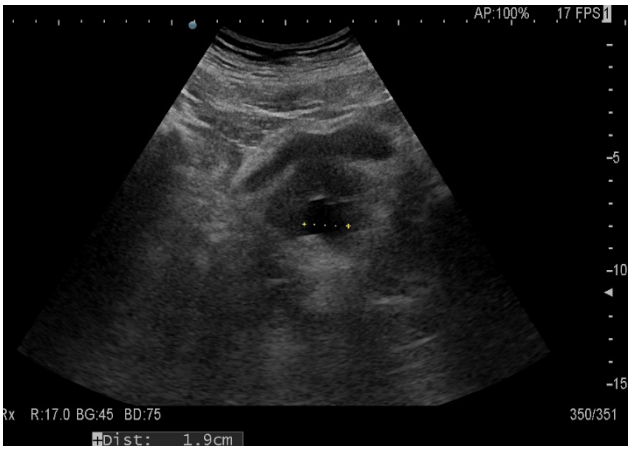

Figure 1. Transverse section through the prostate, showing a cyst-like lesion.

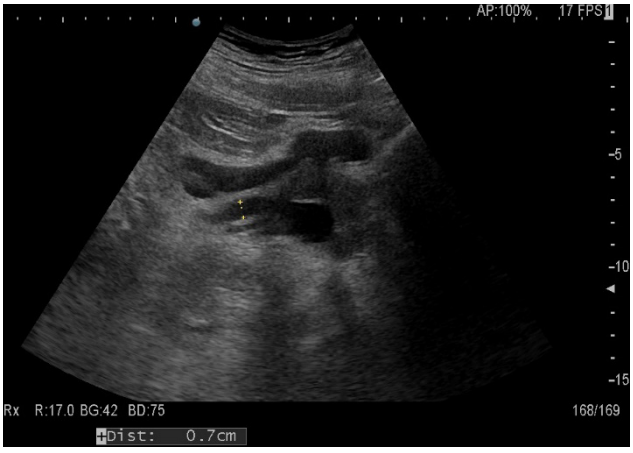

Figure 2. Sagittal section through the prostate, showing a dilation of the left ejaculatory duct.

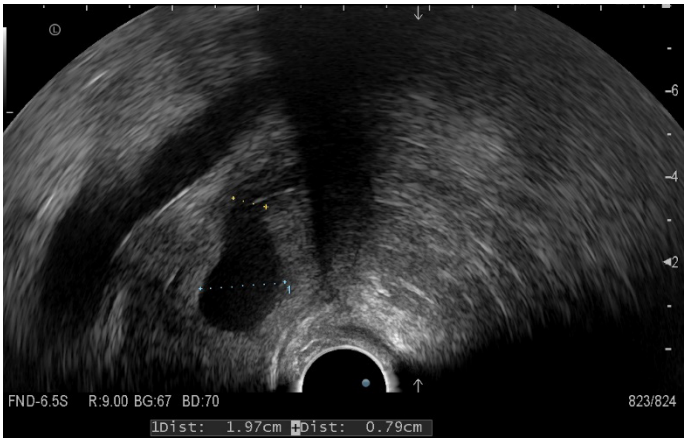

Figure 3. Endorectal ultrasonographic section through the prostate, showing the dilation of the ejaculatory duct.

Contrast-enhanced abdominal and pelvic CT scan was performed, confirming the absence of the left kidney (Figure 4A) and the cystic-like lesion in an axial section of the prostate (Figure 4B), but it also revealed the cystic structure of the left seminal vesicle (Figure 4C).

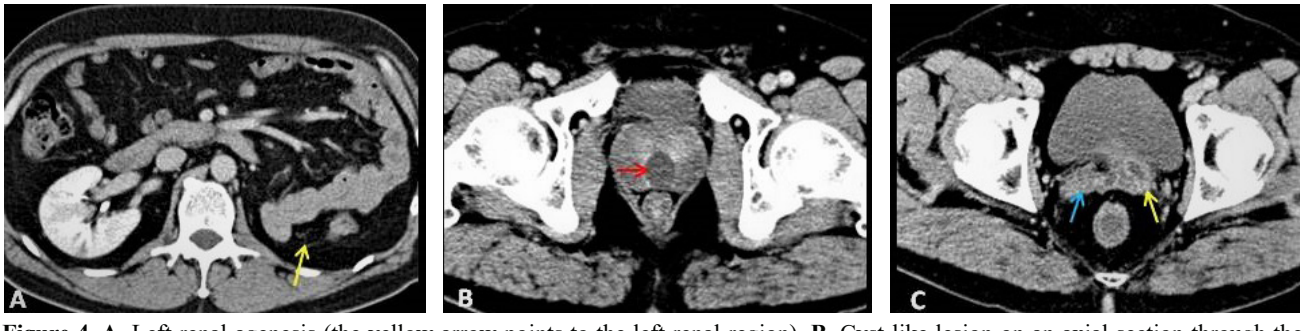

Figure 4. A. Left renal agenesis (the yellow arrow points to the left renal region). **B.** Cyst-like lesion on an axial section through the prostate (red arrow). **C.** The left seminal vesicle has a cystic structure (yellow arrow), but a lower volume compared to the right one, which has normal volume and structure (blue arrow).

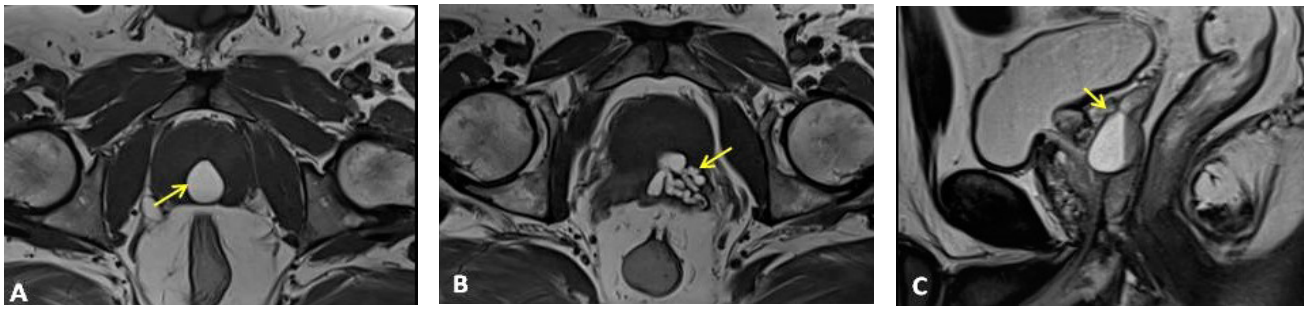

Figure 5. A and B – axial sections through the prostate, showing a T1 hypersignal tubular lesion communicating with the left seminal vesicle (dilated left ejaculatory duct). C. Sagittal section through the dilated ejaculatory duct, displaying fluid-fluid level. Yellow arrows point to the dilated duct.

Contrast-enhanced pelvis MR confirmed a cyst-like intraprostatic lesion (Figure 5A), which was communicating with the left seminal vesicle (Figure 5B). Hypersignal in the T1 sequence (Figures 5A and 5B) suggested a proteinaceous or hematic content, as did the fluid-fluid level visible in the T2 sequence (Figure 5C).

Flexible cystoscopy showed a narrowing of the lumen of the prostatic urethra by a posterior bulging.

The diagnosis of Zinner syndrome was set and anti-inflammatory treatment was given, leading to a quick improvement and discharge from the hospital.

Discussion

To our knowledge, this is the first case of Zinner syndrome published in Romania. In 2008 a group of physicians in Bucharest reported a malformation that could be classified as a particular form of Zinner syndrome: single seminal vesicle, which suffered a cystic transformation and enlargement, associated with unilateral kidney agenesia [7].

Zinner syndrome was first described by the author who gave its name in 1914. It is a rare disease, and it is not easy to have a correct estimation of its prevalence. In an ultrasonographic screening performed on 280,000 children over 2 and a half years in Taiwan in 1990, 13 cases of Zinner syndrome were identified, thus estimating a frequency of 0.00464% [8]. In a systematic review of the publications on Zinner syndrome between 1999 and 2020, a Chinese group of researchers counted 214 cases.

The exact cause of the syndrome is unknown. Its pathogenetic mechanism consists in a failure of the mesonephric duct to develop, between the 4th and the 13th week of intrauterine life, in both directions: upwards, where there is no fusion between the ureteric bud and the metanephric blastema, which is hence not induced to form the kidney, and downwards, where the permeability of the ejaculatory duct is affected [1].

The age at diagnosis varies between newborn and 76 years [1]. In the adult population the most frequent clinical presentation is that of our patient, i.e., urinary symptoms like dysuria, painful micturition, urgency, incontinence, hematuria. The next group of symptoms as frequency of appearance is local pain: perineal, lower abdominal, pelvic, and scrotal pain. Disturbances in the reproductive function are also described: infertility, painful ejaculation, decrease of the volume of the semen. Hematospermia may be the presenting symptom [1,9]. About 20% of the patients are asymptomatic and the diagnosis is set thanks to the imaging workup for other diseases [1,10-12].

In the pediatric population the presentation depends on the age. While in teenagers the presentation is similar to that of adults, very small children have particular presentations: urinary infections, pelvic pain, or the discovery of the syndrome is incidental. Some of the very small children have the diagnosis through pre-natal ultrasonographic screening [13,14].

According to the most complete study, the pooled analysis of the Chinese group on 214 cases, most of the patients (65.8%) underwent surgical interventions (seminal vesiculectomy, unroofing of cysts etc.). It is worth noting that most of the interventions were minimally invasive, using the laparoscopic or even robotic approach [1,4-6].

The outcome of most of the patients is favorable. Out of the 214 patients in the pooled database of the Chinese researchers, 52 patients had complications (mainly infections, even sepsis) and comorbidities. Among the comorbidities the authors name the tumors, mostly adenoma and adenocarcinoma of the seminal vesicle. There were 3 deaths [1].

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