Zinner syndrome: 
a case report and literature review

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Abstract

Introduction and objective: Zinner syndrome is a rare congenital abnormality of the mesonephric duct (Wolffian) consisting of unilateral renal agenesis, seminal vesicle cyst and ipsilateral ejaculatory duct obstruction. Abnormalities involving the contralateral seminal vesicle or ejaculatory duct are extremely rare. We present the therapeutic approach in a patient with renal agenesis and large contralateral seminal vesicle cyst.

Patient, Method and Results: A 24 years old patient known with right congenital solitary kidney presents with dysuria, pollakiuria, nocturia, hypospermia, ejaculatory pain and diffuse hypogastric and perineal pain. The imaging exams (abdominal ultrasound, contrast-enhanced computed tomography) reveal a cystic mass of the lower mid-abdomen of 14/11 cm with mass-effect, the absence of the left kidney, grade II right hydronephrosis and right megaureter. Preoperatively, the transonic mass has been interpreted as a seminal vesicle cyst. We performed the robotic-assisted laparoscopic cyst resection by using an approach similar as for radical prostatectomy. The intraoperative assessment revealed a cystic mass with a point of origin in the right seminal vesicle, contralateral to the renal agenesis, with an aspect suggestive for Zinner syndrome.

Conclusions: Zinner syndrome is a rare urological condition that must be suspected in the young adult with recurrent irritative-obstructive lower urinary tract symptoms associated with pelvic pain syndrome, pelvic cystic mass, unilateral renal agenesis and significant alteration in sperm parameters. The conservative approach is limited to asymptomatic cases. The robotic-assisted minimally invasive approach facilitates the accurate delineation and isolation of anatomical structures, in a territory where subsequent to pericystic inflammation and changes in surgical plans, the classic surgical approach to adjacent structures proves difficult.

Keywords: contralateral renal agenesis, dysuria, hypospermia, seminal vesicle cyst, Zinner syndrome
**Clinical cases**

**Introduction**

Zinner syndrome is a congenital urological abnormality of the Wolffian distal mesonephric duct that appears between weeks 4 and 13 of gestation and is considered to be the male counterpart of Mayer-Rokitansky-Küster-Hauser syndrome seen in women. The seminal vesicle cyst incidence is 0.005% and in two thirds of cases is associated with ipsilateral renal agenesis [1-2].

Its particularity consists in the association of unilateral renal agenesis, seminal vesicle cyst and ipsilateral ejaculatory duct agenesis or atresia [3-4].

The cystic mass starts to expand once with the installment of sexual life, with a right/left side incidence of 2:1. In the case of asymptomatic patients the diagnosis is established after the age of 30, when assessing the fertility related issues [5-6]. The clinical manifestations are atypical in most of the symptomatic patients – dysuria, urgency, polakiuria, ejaculatory or pelvic pain, occurring in the context of a large cyst or stenotic malformations [7].

In most of the reported cases, seminal vesicle cysts do not exceed 5 cm in diameter; however, gigantic cysts measuring up to 12 cm in diameter have been also reported in the literature. In this case, the cyst has a diameter of nearly 14 cm, with adjacent mass-effect, causing ureteral obstruction with secondary hydronephrosis and impaired renal function as well as irritative-obstructive symptoms of the lower urinary tract (LUTS).

The therapeutic decision is influenced by the size of the cyst and its systemic impact. Treatment consists of antibiotherapy and transurethral drainage, transurethral incision of the ejaculatory duct and cyst resection by a classical surgical, laparoscopic or robotic-assisted approach [8].

**Case report**

A 24 years old patient presents with dysuria, polakiuria, dribbling, nocturia, hyposepermia, oligoasthenoteratozoospermia, ejaculatory pain, chronic constipation and diffuse perineal and hypogastric pain, that started 9 weeks prior to admission. A diagnosis of congenital solitary right kidney was made in 2009, upon a routine ultrasound scan, but no pelvic cystic mass or hydronephrosis had been observed at that time.
We decided in favor of a robotic-assisted laparoscopic cyst resection procedure and adopted an approach similar to that used for radical prostatectomy, using 5 trocars (3 for the robotic arms and 2 for the assistant). The optical trocar (1) have been positioned 2 cm above the umbilicus, the two robot working ports (2, 3) at 10 cm each from the optical trocar (1) on a transverse line 2 cm below the umbilicus, and two assistant ports [12 mm (5) and 5 mm (4)] (Fig. 2). The surgical table was set in Trendelemburg position, with the patient tilted at 30 degrees. In order to avoid bowel lesions or cyst perforation secondary to its mass-effect, pneumoperitoneum was obtained with the Hasson technique (port 1). Circumferential cyst isolation, advanced slowly due to its high volume, the pericystic tissue inflammation and the multiple vascular variants. Intraoperative cyst drainage was required with the externalization of approximately 2300 ml of sero-citrin liquid. No neoplastic cells were found in the liquid cytology specimen.

Vasa deferentia have been identified and isolated from among the cystic neovascular emergences. The cyst was in intimate contact with the left seminal vesicle, with normal macroscopic-anatomic aspect. The cystic wall was extracted using a specimen bag. A Jackson-Pratt drain was used for the pouch of Douglas, with the exteriorization of about 60 ml of serosanguinous liquid, which was removed after 24 hours. The actual console time was 85 minutes, estimated blood loss during procedure 50 ml, and the patient was discharged day four postoperatively.

The histopathology exam revealed the presence of cylindrical epithelial tissue, with smooth muscle fibers with seminal vesicle components (Fig. 3).

**Fig. 3**
Histologic analysis of excised specimen

**Discussions**

The Zinner syndrome is defined by the triad consisting of renal agenesis, seminal vesicle cyst and ipsilateral ejaculatory duct obstruction [4]. It is a rare congenital condition and the uniqueness of the involved abnormalities, as in this case, is provided by the presence of renal agenesis and the contralateral seminal vesicle impairment. A review of the available literature revealed only four similar cases being reported to date [9,10].

An ultrasound scan may provide high accuracy imaging in many cases [11], but may be of limited use in obese patients or in patients with high-volume tumors. Both abdominopelvic and endorectal ultrasound scans were performed, but failed to identify the point of origin of the cyst due to its mass-effect onto the adjacent anatomical structures.

In selected cases, where standard imaging is inconclusive, a vasoseminal vesiculography is performed with cyst drainage and subsequent contrast media injection for diagnosis purposes [12].

We performed a contrast-enhanced computed tomography (CT) of the abdominopelvic segment in order to enhance diagnosis. However the origin of the cystic mass could not be observed and remained to be determined intraoperatively.

Recent data have shown that magnetic resonance imaging (MRI) is more effective in assessing sperm ducts and seminal vesicles due to high resolution, superior soft-tissue contrast and multiplanar imaging capabilities that can accurately define the anatomic relationships of the examined tissues [13]. These results are consistent with our experience. In this case the CT evaluation proved to be of limited value.

There are numerous therapeutic alternatives, from antibiotherapy and percutaneous or transurethral drainage with subsequent sclerosing, to transurethral incision of the ejaculatory duct and classic surgical, laparoscopic or robotic-assisted cyst resection. Various approaches to treatment exist, with a tendency towards confining surgery to symptomatic patients [8]. Experience with robotic-assisted seminal vesicle cyst resection is limited, and even more in the case of high-volume cysts. According to published data, there are only four cases of robotic-assisted laparoscopic cyst resection for tumors with a diameter exceeding 12 cm.

Issues regarding surgical treatment have been pointed considering the importance of avoiding vicinity organ injury and preservation or regaining of fertility after the procedure. Due to changes in anatomical structures and surgical planes there is a heightened risk of damaging the ureter, pelvic vessels, nerve fibers, urethra and the ipsilateral or contralateral seminal vesicle and spermatic duct.

The high-accuracy magnification and the degrees of freedom of the robotic arms in robotic-assisted surgery offer a high degree of safety in the delimitation of...
adjacent structures, which is not found in classic surgery. [14]

**Conclusions**

Zinner syndrome is a rare urological condition with a significant impact on the quality of life of the individual as well as the couple, secondary to the impaired fertility.

Conservative treatment is limited to asymptomatic patients.

The robotic-assisted minimally invasive approach facilitates the precise delimitation and isolation of adjacent anatomical structures, in a territory where due to pericystic inflammation, numerous vascular variants and changes in dissection planes, the classic surgical approach proves very difficult.

**References**