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Robotic management of painful Zinner syndrome, case report and review of literature

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ABSTRACT

BACKGROUND: Zinner Syndrome is a congenital pathology due to an embryologic anomaly occurring between the 4th and 13th gestational week. This embryologic defect leads to unilateral renal agenesis, ipsilateral seminal vesicle cyst and ejaculatory duct obstruction. Most of the time patients are asymptomatic and do not need any treatment but for symptomatic cases, only surgical removal of the cyst and seminal vesicle are 100% effective.

CASE: The case presented here is that of a healthy 33-year old man with symptomatic right seminal vesicle cyst and ipsilateral renal agenesis. First a conservative approach was attempted but each time the symptoms ended up reappearing. We decided to use robot-assisted laparoscopy to completely resect the cyst and the right seminal vesicle. There was no postoperative complication and the patient's symptoms improved immediately. After a 6 months follow-up the patient remains completely asymptomatic.

CONCLUSION: Complete excision of the seminal vesicle cyst is the only 100% effective treatment option for symptomatic patients with Zinner syndrome. Minimally invasive approaches like conventional laparoscopy or robotic assisted laparoscopy are safe and effective and should currently be considered as the surgical gold standard.

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1. Introduction and background

Zinner Syndrome is a congenital triad of abnormalities, comprising unilateral renal agenesis, ipsilateral seminal vesicle cyst and ejaculatory duct obstruction [1]. The origin of this syndrome comes from an embryologic anomaly occurring between the 4th and the 13th gestational week. Normally, the ureteric bud develops from the distal mesonephric duct and extends in a dorsocranial way to meet the metanephric blastema which will differentiate to form the kidney. The mesonephric duct also differentiates later into epididymis, vas deferens, ejaculatory duct, seminal vesicle and hemitrigone [3]. The Zinner Syndrome results from an anomaly of development of the distal portion of the mesonephric duct.

The occurrence of symptoms are related to the growth of the cyst during the sexual active life [2]. The most common symptoms are: dysuria, frequency, perineal pain, epididymitis, pain following ejaculation and scrotal pain [1].

Currently, the treatment for symptomatic men is a surgical resection of the cyst and the seminal vesicle because of the high rate of recurrence after conservative treatment. This case report has been reported in line with the SCARE criteria [4].

2. Presentation of case

The case presented here is that of a 33-year old man complaining of dysuria, burning during micturition and painful ejaculation.

Physical examination was unremarkable except for a sensitive right testis.

Urinalysis and blood tests were unremarkable. Scrotal ultrasound found a cyst of the epididymis and the patient was first treated with antibiotics and surgical removal of the cyst was performed because of persisting symptoms.

Anatomopathological analysis of the cyst showed benign cells.

Initially symptoms improved but reappeared one year later.

The digital rectal examination was still normal but the transrectal ultrasound revealed a large cyst next to the bladder floor.

Magnetic resonance imaging (MRI) showed a 49 × 33 × 42 mm cyst of the right seminal vesicle (Fig. 1).

A conservative approach was adopted and a transrectal punction and aspiration of the cyst was performed. The cytological analysis of the liquid revealed no evidence of malignancy. The symptoms totally disappeared after the aspiration but they eventually reappeared after a year.

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Fig. 1. Axial T2 weighted MRI image showing a large (49 × 33 × 42 mm) cyst of the right seminal vesicle.

The patient underwent iterative punctures over 4 years but every time the complains reappeared after approximately a year. A CT scan was performed and showed total agenesis of the right kidney, which established the diagnosis of Zinner Syndrome. A more radical approach was therefore necessary. A minimal invasive laparoscopic robot assisted approach was performed with surgical excision of the cyst.

The patient was installed as for radical prostatectomy (i.e. supine position, legs spread, after placement of a Foley catheter). Four transperitoneal robot trocars (placed in an arc around the umbilicus) and two assistants trocars (one 5 mm under the xyphoïd appendix and one 12 mm above the right antero-superior iliac spine) were used.

The patient was put on a 28 degree Trendelenbourg position and a 30 degree camera was used (Fig. 2).

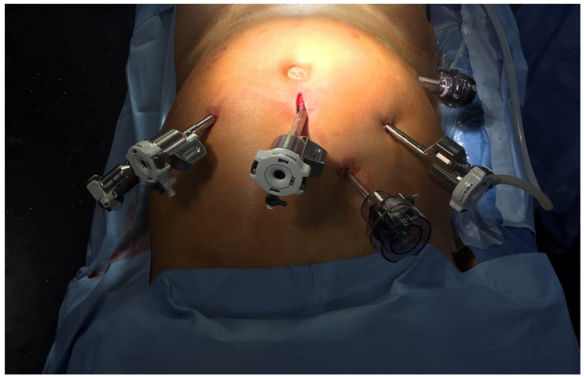

Fig. 2. Position of the six trocars.

The peritoneum overlying the Douglas pouch was incised over the bulge formed by the cyst. The two seminal vesicles and the two vas deferens were identified and were pushed back to the left by the cyst. The cyst was completely adherent to the right posterior wall of the bladder and potentially to the right ureter (Fig. 3).

The dissection of the right ureter started from top on the crossing over the iliac vessels downwards to its insertion into the bladderwall. An ultrasound probe and a flexible cystoscope were used to ensure of the absence of communication between the cyst, the bladder and the ureter.

After ensuring no ectopic ureteral insertion, the cyst was incised and a citrine fluid was suctioned.

Blunt dissection was used to peel the walls of the cyst away from the bladder and the right ureter. The right seminal vesicle and the distal part of the right vas deferens were resected with extreme caution due to the proximity of the neurovascular bundles. The specimen was removed as a whole through the 12 mm assistant port using an Endobag (Fig. 4).

Intravenous Indigo Blue injection was used to exclude an injury of the right ureter. A portion of the bladder seemed dangerously thin, therefore a 2-0 Vicryl cross stich was placed.

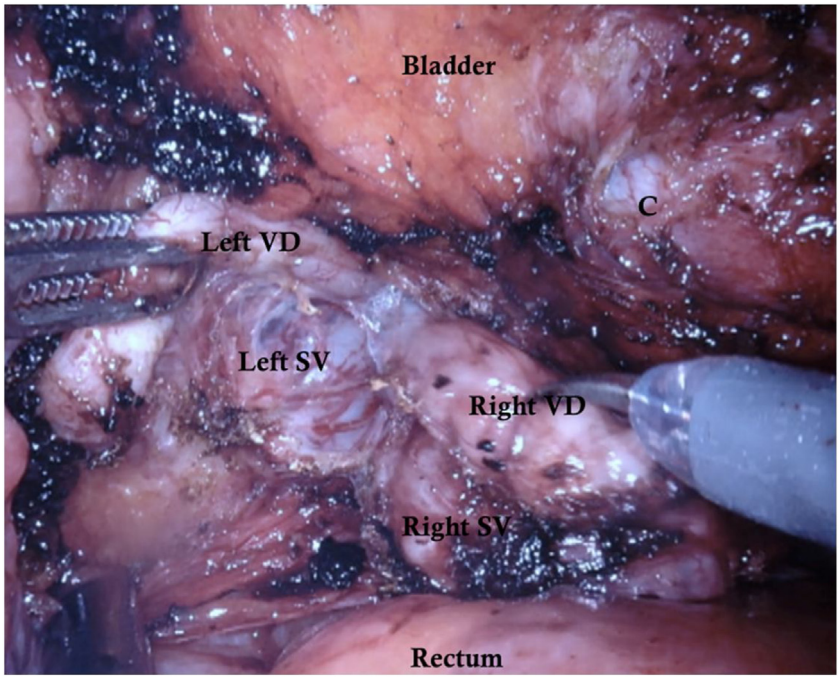

Fig. 3. View of the disposition of the vas deferens (VD) and the seminal vesicle (SV) in relation to the cyst (C).

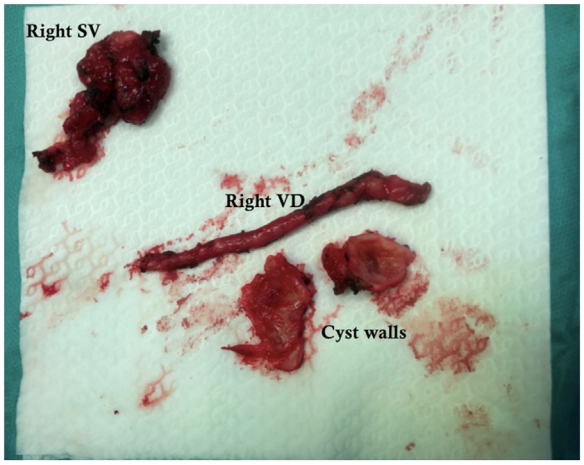

Fig. 4. Macroscopic view of the right seminal vesicle – the right vas deferens and the wall of the cyst.

After the hemostase check, surgisnow was put in the Douglas pouch and a Jackson-Pratt drain was placed.

Operative time was 120 min. Blood loss estimate was less than 50 mL.

The drain was removed and the patient was discharged on post-operative day 2. The Foley catheter was removed on postoperative day 4.

Anatomopathological analysis showed benign cells.

There was no postoperative complication and the patient's symptoms improved immediately. After 6 months follow-up the patient remains completely asymptomatic.

3. Discussion

Zinner syndrome is a rare congenital pathology. At present, there are only 200 cases reported in the literature. The diagnosis should be evoked in young, sexually active man, with vesicle cyst causing dysuria, frequency, epididymitis, scrotal and ejaculatory complains [1,2].

Asymptomatic cases, which are the vast majority do not need any treatment [1–3]. For symptomatic cases different treatment options can be considered.

A conservative transrectal aspiration, is easy to perform and worth a try, but as it carries a high risk of recurrence and infection, the procedure should probably not be repeated after failure [1,2]. Only surgical removal of the cyst and seminal vesicle are 100% effective [1,2].

Historically, open vesiculectomy via transabdominal or transperineal approach were the only surgical options. Currently, in the area of minimal invasive surgery, laparoscopic and robot assisted surgery should definitely be considered. This minimally invasive approaches offer better vision, reduced blood loss, shorter hospital stay and faster recovery [5].

Robot assisted approach improves conventional laparoscopy advantage even further. It offers a better 3D visualization, enhanced dexterity and precision, improved surgeon comfort and suture quality and reduced blood loss [6,7].

To the best of our knowledge, only 6 cases of Zinner syndrome that have been managed by robotic surgical resection of the seminal vesicle cyst, have been described in the literature [5–10].

One case was managed by simple resection of the cyst without performing a vesiculectomy. Good early postoperative results are described but follow up data are lacking [8]. The 4 other cases associated a vesiculectomy, which should guarantee the absence of long term recurrence. In one of the cases managed by vesiculectomy, per-operative discovery of atrophic ipsilateral kidney resulted in change of operative plan and surgery was completed by concomitant removal of atrophic kidney [5].

In our case, the kidney was known to be atrophic pre-operatively but it was decided not to perform a concomitant nephrectomy. First of all because we found no recommendations justifying nephrectomy in the literature [11,12]. Also a concomitant nephrectomy would have implied having to undock the robot and change the patient's position on the table, which would have increased the surgical complexity and operating time.

4. Conclusion

Zinner syndrome is a rare congenital pathology comprising unilateral renal agenesis, ipsilateral seminal vesicle cyst and ejaculatory duct obstruction. Resection of the cyst is only mandatory for symptomatic patients.

Minimally invasive approach like conventional laparoscopy or robotic assisted laparoscopy should currently be considered as the surgical gold standard.

Conflicts of interest

None.

Sources of funding

None.

Ethical approval

None. Because this was a report of an interesting case, and not a trial or observational research, there was no need for ethical approval.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying image.

Author contribution

Laura Demaeyer : Conceptualization, Writing and Original Draft.
Serge Holz : Supervision.
Didier Pamart : Resources and Investigation.
Steven Taylor : Resources and Investigation.
Michel Naudin : Supervision.
All authors read and approved the final manuscript.

Registration of research studies

None. Because this was a report of an interesting case, and not a trial or observational research.

Guarantor

Dr Michel Naudin, CHU Ambroise Paré, Mons, Belgium.

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