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MINIMALLY INVASIVE MANAGEMENT OF ZINNER'S SYNDROME

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Abstract Zinner's syndrome (ZS) is a rare congenital malformation of the seminal vesicles and ipsilateral upper urinary tract which is characterized by the triad of ipsilateral ejaculatory duct obstruction, seminal vesicle cysts, and renal agenesis. We report an 18-year-old male who presented intermittent gross hematuria and hematospermia and ejaculation volume diminution. ZS was diagnosed with magnetic resonance imaging (MRI) of the prostate, among other complementary studies requested. We performed a minimally laparoscopic resection of the left seminal vesicle. The postoperative was uneventful symptoms resolved, and 3 months later semen analysis showed increase in ejaculation volume. Currently, the minimally invasive approach is feasible and effective for the treatment of the seminal vesicle cysts excision in the ZS. We presented a symptomatic patient treated by a laparoscopic approach with successful middle follow-up results.

Key words: laparoscopy, Zinner syndrome

Resumen ***Manejo mínimamente invasivo del síndrome de Zinner.*** El síndrome de Zinner (ZS) es una malformación congénita rara de las vesículas seminales y del tracto urinario superior ipsolateral que se caracteriza por la tríada de obstrucción del conducto eyaculatorio ipsolateral, quistes de vesículas seminales y agenesia renal. Presentamos el caso de un varón de 18 años que presentó hematuria macroscópica intermitente y hemospermia y disminución del volumen de eyaculación. El diagnóstico de ZS se realizó mediante resonancia magnética (RM) de próstata, entre otros estudios complementarios solicitados. Realizamos una resección mínimamente laparoscópica de la vesícula seminal izquierda. El postoperatorio se resolvió sin complicaciones y 3 meses después el análisis de semen mostró un aumento en el volumen de eyaculación. Actualmente, el abordaje mínimamente invasivo es factible y efectivo para el tratamiento de la escisión de quistes de vesículas seminales en el ZS. Presentamos un paciente sintomático tratado por vía laparoscópica con seguimiento medio exitoso.

Palabras clave: laparoscopia, síndrome Zinner

Zinner's syndrome (ZS), is an infrequent congenital malformation of the seminal vesicles and ipsilateral upper urinary tract[1]. It is a developmental arrest during the embryogenesis between 4 and 13 weeks that affect the Mullerian duct. Up to now, less than 300 cases were reported[2]. ZS is characterized by a phenotypic triad of ejaculatory duct obstruction, seminal vesicle malformation, and unilateral kidney agenesis. Diagnose is commonly made in older adolescence due to lower tract urinary symptoms such as dysuria, urinary frequency, perineal pain and epididymitis, and more rarely ejaculatory symptoms. Diagnosis is confirmed by imaging[3].

We present a symptomatic patient with ZS which was laparoscopically treated.

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Clinical case

An 18-year-old male presented with intermittent gross hematuria and hematospermia in the last three months. The patient did not have relevant clinical or surgical history. Creatinine levels and cultures (urine and semen) were normal and negatives. Ultrasonography revealed the absence of the left kidney and no other pathological result. The semen analysis was normal but the volume of the ejaculation was less than the standard results. We performed a flexible cystoscopy that showed a cyst-like lesion that expanded over the bladder trigone. Magnetic resonance imaging (MRI) of the prostate showed a cystic structure from the left seminal vesicle with blood or high-protein content, which has a size of 39 × 19 mm (Fig. 1). Due to the patient continued with hematospermia and hematuria, we performed a laparoscopic approach with the four-trocar technique. The peritoneum was incised (overlying the dilated left seminal vesicle) and the left seminal vesicle was transected using a cold-scissor with prior placement a Hem-o-Lok clip to not to injure the pararectal plexus located lateral to the tip of the vesicle while dissecting it off. Special efforts were taken with the right *vas deferens*, seminal vesicle, and neurovascular bundle during the dissection to preserve fertility (Fig. 2). The specimen was removed via the 12-mm trocar using a recovery bag. Surgical time was 45 minutes, without bleeding or the placement of an abdominal drain. On

Fig.1.– MRI images A) sagittal, and B) axial, showing the tubular structure arising from seminal vesicle. Coronal MRI images C) and D), show a tubular structure from the left seminal vesicle with high-protein content (39 mm × 19 mm).

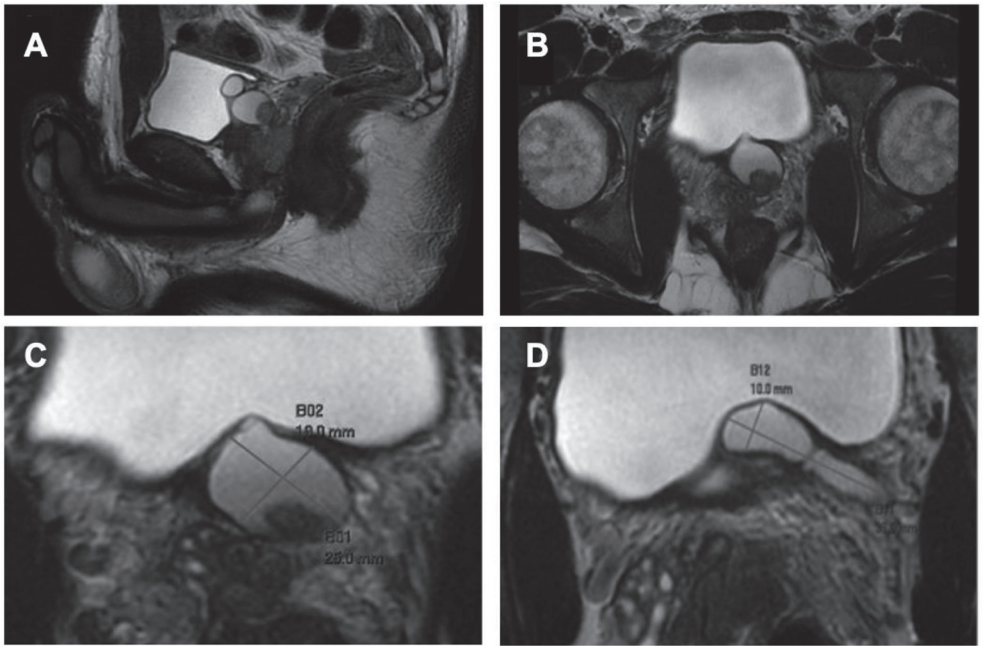
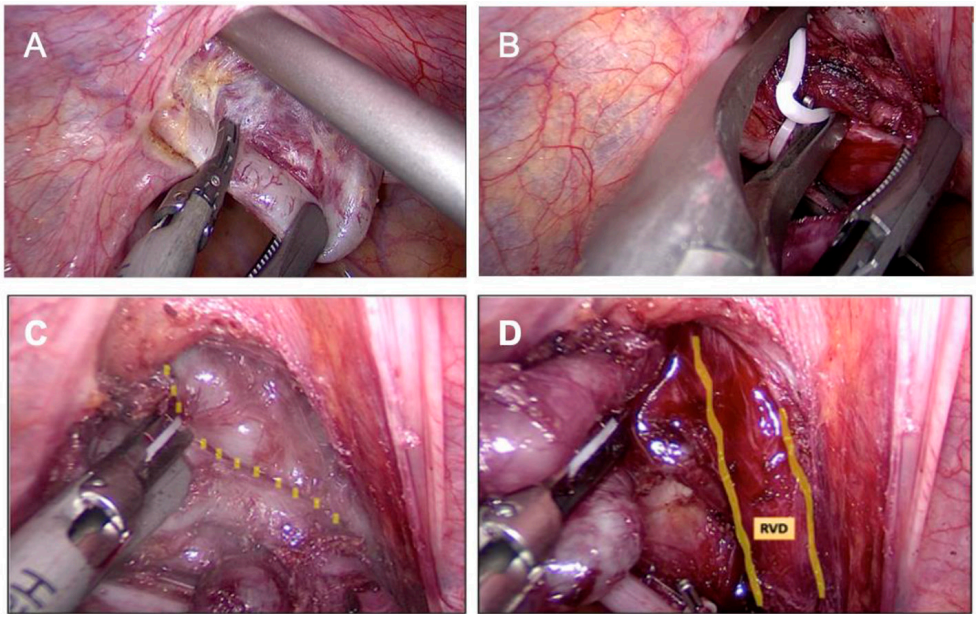


Fig. 2.– A) Dissection of left vas deferens to visualize left seminal vesicle. B) Hem-o-Lok clip placement to seminal pedicle's dissection. C) and D) show neurovascular bundle preservation and separation of the right vas deferens during the dissection.



RVD: right vas deferens

the first postoperative day, the Foley catheter was removed and the patient was discharged without complications. Pathology did not reveal any malignant elements. Hematospermia and hematuria resolved, and three months postoperative semen analysis showed increase of ejaculation volume. Currently, the patient is 39 months after the surgery and continue asymptomatic.

Discussion

ZS is a disease that has an embryological origin. The mesonephric duct forms the reproductive male system (seminal vesicles) and the ureteric bud which form the renal system. At some point of mesonephric duct development, its malfunction leads to the phenotypic triad of ejaculatory duct obstruction, seminal vesicle malformation, and unilateral kidney agenesis. Patients with small seminal vesicle cyst remain asymptomatic and could be managed by conservative treatment. Larger cysts usually present symptoms in late adolescence such as urgency, frequency, dysuria, hematuria due to bladder irritation. However, other patients present hematospermia, painful ejaculation, or infertility, with a later oligozoospermia and even azoospermia. Usually, these symptoms occur when the cyst is larger than 8-10 cm. There are also reported cases with very large cysts that can resulting in the bladder or colonic obstruction. Nevertheless, in this case the cyst's size was smaller than previous reports and it even produced symptoms. Ultrasound can be helpful as initial approach showing ipsilateral renal unit is absent and transrectal ultrasound the obstructed ejaculatory ducts could be identified. MRI is very accurate for delineation of the prostate and seminal vesicles and confirming that the cystic periprostatic structures are indeed within the seminal vesicles, and also identifying ectopic ureteric orifices. Historically, the gold-standard treatment was open surgery, but currently, the minimally-invasive approaches as laparoscopic and robotic are the practice. Since Kavoussi et al[4] described using laparoscopy for retro vesical lesions it became the preferred method for treating these patients. These approaches present some advantages as of direct access to the pelvis and the seminal vesicle, such as good visualization and easy access to the deep pelvis. Kord et al[5] reported 5 patients' series that were surgically treated. Four of them laparoscopically and one robotic-assisted. They reported a long hospital stay (mean = 7.4 days) but presented a 3-year long-term follow-up without symptoms recurrences.

Currently, the minimally invasive approach is feasible and effective for the treatment of the seminal vesicle cysts excision in the ZS. We presented a symptomatic patient treated by a laparoscopic approach with successful middle follow-up results.

Conflict of interest: None to declare

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