VIDEO ARTICLE
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Zinner Syndrome: A Rare Case Report

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Abstract

Over 200 cases of seminal vesicle cysts linked to ipsilateral renal agenesis have been reported in the literature, indicating Zinner syndrome. This condition occurs when the ureteric buds fail to meet the metanephros, leading to cystic dilatation in the ipsilateral seminal vesicle along with unilateral renal agenesis. Here, we are discussing a 17-year-old boy who presented with lower urinary tract symptoms predominantly flow symptoms, was evaluated thoroughly, and robotic surgery was selected as the best minimally invasive treatment.

Keywords: Zinner syndrome, general urology, radiology, seminal vesicle cyst

Introduction

More than 200 cases of seminal vesicle cysts associated with ipsilateral renal agenesis have been reported. These cases are indicative of Zinner syndrome (ZS), a rare congenital condition characterized by cystic seminal vesicles and ejaculatory duct obstruction in association with ipsilateral renal agenesis (1).

Embryology

Due to inadequate migration, the ureteric bud emerging from the proximal section of the Wolffian duct cannot join the metanephros (1).

The inability of the ureteric buds to migrate from the mesonephric duct is the underlying reason for the failure to meet metanephros. As a result, cystic dilatation develops in the ipsilateral seminal vesicle due to ejaculatory duct blockage and ipsilateral renal agenesis caused by the failure of metanephric blastoma to differentiate (2).

Case Presentation

A 17-year-old boy presented with chief complaints of voiding lower urinary tract symptoms for 3 months. Local examination of the genitalia and abdomen was normal, and the digital rectal examination (DRE) was also normal.

Ultrasound revealed left atrophic kidney (5 cm), left dilated ureter, and the remaining anatomy was normal.

Computed tomography (CT) scan abdomen findings are depicted in Figures 1, 2 and 3. Semen analysis revealed oligospermia. All blood tests were normal.

Patients and relatives were counseled regarding left nephroureterectomy with cystic lesion excision robotically.

Materials and Methods

Surgery (Voice Over Included with Video)

Initially in the lithotomy position, cytoscopy showed normal urethra, right ureteric orifice normal, left ureteric orifice could not be localized, left poster-lateral bladder wall bulging toward the lumen, possibly due to extraluminal compression by the cystic structure. The bladder mucosa was normal.

Position changed to right lateral position: atrophic kidney dissected with vasculature clipped and cut, and ureter dissected until bladder.

Position changed to Lithotomy with head low position-the ureter was found to be ending in the left cystic structure, possibly the left seminal vesicle, dissected and cut at the base. An abdominal drain was placed. Specimens were retrieved in bags by Pfannenstiel incision. The postoperative period was uneventful.

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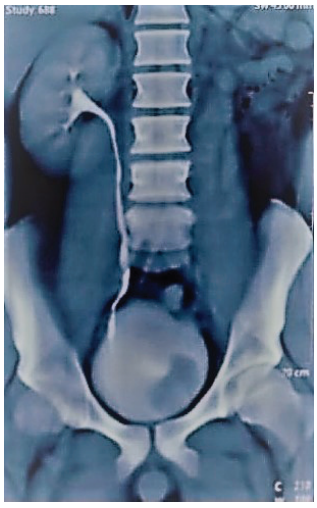

Figure 1. Non-functional left kidney

Figure 2. Atrophic left kidney with dilated ureter opening in the cystic leaves seminal vesicle

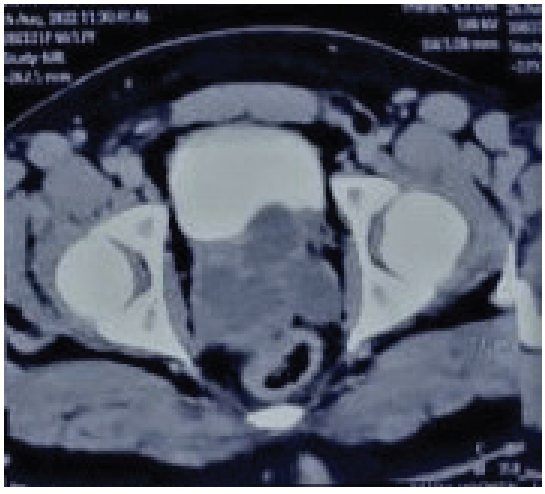

Figure 3. Left seminal vesicles could not be identified separately from the cystic structure

Discussion

Zinner's syndrome is a rare congenital disease characterized by the association of ipsilateral renal agenesis or dysplasia, ectopic uretera, and ipsilateral seminal vesicle cysts, first described by Zinner in 1914 (3).

It is thought that Mayer-Rokitansky-Küster-Hauser syndrome affects females and ZS affects males. An ectopic ureter may develop when the ureteric bud emerges distally from the urogenital sinus. It might empty into the seminal vesicle cyst, vas deferens, ejaculatory duct, or bladder neck. Seminal vesicles and dilated tubules may become cysts as a result of ejaculatory duct occlusion (2).

Therefore, aberrant ureteric bud development is a cause of ZS. Abdominal pain, fullness, and micturition-related symptoms, such as dysuria, hematuria, urgency, and obstructed urination are typical presenting features. Signs become more noticeable during peak sexual or reproductive activity, particularly in the second and third decades (4), although they can sometimes appear without symptoms (2).

The ipsilateral ureter and kidney are confirmed to be absent on CT urogram, but the origin of the cyst is typically not confirmed. The best method is magnetic resonance imaging, which also allows analysis of the cyst contents, which are typically pure liquid and appear hypointense on T1 and hyperintense on T2 (5). DRE may or may not reveal a clinically significant finding. In our case, there was no clinically aberrant finding however (6) had shown a cystic mass palpable per rectally.

Clinical assessment determines how ZS should be managed (3). The patient's symptoms, cyst size, and presence of comorbidities all influence how the patient is managed. It is possible to use observation management in patients who are asymptomatic or have minimal symptoms (6). For patients with minor symptoms, conservative treatment with antibiotics, transurethral needle aspiration of the cyst, or transurethral aspiration combined with substance instillation (alcohol and minocycline) is appropriate (7). Despite being simple to perform, conservative transrectal aspiration carries a significant risk of infection and recurrence; if it proves fruitless, it should not be repeated (6). The cornerstone of treatment for symptomatic patients is surgery (2). The condition can be treated surgically using an open, laparoscopic, or robotic technique.

The advantages of a robotic approach over conventional laparoscopy include the ability to manipulate instruments more easily in a small working space and to move with greater precision and degrees of freedom. This allows the calibrated use of thermal energy, which lowers the risk of blood loss and nerve injury. Because seminal vesicles are located deep in the pelvis,

better vision with higher magnification and three-dimensional imaging is key to vesiculectomy (3).

Given its positive outcomes, robotic surgery can be regarded as the gold standard for surgical therapy in these patients (3). The surgeon can simultaneously manage the upper and lower which significantly lowers the morbidity associated with making two separate incisions (3).

Prolonged follow-up is required because seminal vesicle diseases are prone to recurrence due to potential coexisting ejaculatory duct abnormalities or its development following surgery. Because of this, transurethral resection of the ejaculatory duct should be taken into consideration (8).

Conclusion

This case presents an adult seminal vesicle cyst, renal dysplasia linked to an ipsilateral incomplete duplicated ectopic ureter, and an uncommon congenital defect of the genitourinary tract. With good surgical and cosmetic outcomes, the robotic-assisted laparoscopic approach was selected as the best minimally invasive method for treating this patient's unusual congenital cystic malformation.

Video 1.

Ethics

Informed Consent: Written informed consent was obtained from the patient and parents regarding the surgery and publication of information on this case in scientific meetings/journals.

Footnotes

Authorship Contributions

Surgical and Medical Practices: G.R.S., A.B., Concept: G.R.S., Design: A.B., Data Collection or Processing: A.B., H.B., Analysis or Interpretation A.B., Literature Search H.B., Writing: A.B.

Conflict of Interest: No conflict of interest was declared by the authors.

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