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Inflammation and infection

Open retropubic prostatectomy in a patient with bladder exstrophy for management of hematuria and urethral discharge – A case report

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ABSTRACT

Bladder exstrophy (BE) is a rare congenital disorder causing bladder and urethral malformation due to an abdominal wall embryological defect. Traditionally, BE had a poor life expectancy, but advances now offer a normal lifespan. A 57-year-old male with BE history and ureterosigmoidostomy repair presented with intractable hematuria, urethral discharge, and recurrent prostatic infections. He underwent retropubic subtotal prostatectomy without major complications, resolving urinary symptoms. Advances in BE management extend patient lifespans. This case demonstrates successful surgical management of hematuria and urethral discharge through radical prostatectomy in an adult with a history of BE.

1. Introduction

Bladder exstrophy (BE) is a congenital anomaly that is part of the bladder exstrophy-epispadias complex (BEEC). BE results from improper formation of the bladder and urethra during fetal development, which consequently presents as an open bladder and urethral plate at birth. The incidence of BE is 1:50,000 live births, making it a rare, but major congenital malformation, with serious implications on patients' quality of life.[1] Historically, BE carried a poor prognosis, with a study in 1926 estimating that more than half of patients died before the age of 10. However, given the advent of modern surgical techniques and improved neonatal intensive care, patients born with BE are now living longer into adulthood.[1] Here, we present our experience with a 57-year-old male with a history of BE in good general health treated with retropubic subtotal prostatectomy for continuous urethral discharge, and intractable hematuria syndrome. Adult reconstructive urologists have a longstanding history of employing prostatectomy and urethral closure techniques to address chronic urethral drainage in patients who have undergone diversion procedures due to conditions such as trauma, post-radiation complications, fistulae, and neurogenic bladder.[2–4] Notably, our case stands out due to the rarity of diversion via ureterosigmoidostomy in bladder exstrophy, making the inclusion of prostatectomy in this context particularly unique.

2. Case presentation

The patient in our report is a 57-year-old male with a history of BE, who previously underwent ureterosigmoidostomy. The patient presented for hematuria and urethral discharge for over 1 year, along with a history of recurrent UTIs. After thorough discussion with the patient, the decision was made to perform a retropubic prostatectomy to treat the persistent hematuria and recurrent UTI's.

2.1. Clinical findings

Imaging studies, including a pelvic magnetic resonance imaging (MRI) and computerized tomography (CT) scan of the pelvis, revealed notable findings. The MRI showed a 2.5 × 1.3 × 1.1 cm cystic structure connected to the prostate, containing prostatic calcifications (Fig. 1). This finding suggested a chronic collection of prostatic secretions at the site of the previous bladder neck transection. Additionally, the CT scan revealed thickening of the right renal pelvis, a calyceal diverticulum, and bilateral fat-containing inguinal hernias.

2.2. Diagnostic procedures

To evaluate the condition further, a cystoscopy with retrograde urethrogram was performed. The procedure accessed the patient's epispadic urethra and examined the urethra using fluoroscopic

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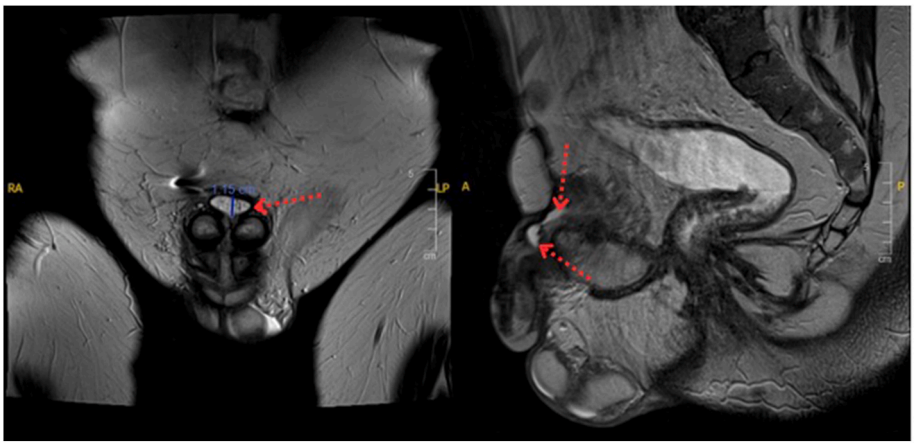

Fig. 1. Anterior to the prostate and superior to the penile base, there is a 2.5 × 1.3 × 1.1 cm peripherally enhancing cystic structure which contains a direct connection to the prostate. The lesion contains numerous T1/T2 hypointense layering stones likely corresponding to prostatic calcifications.

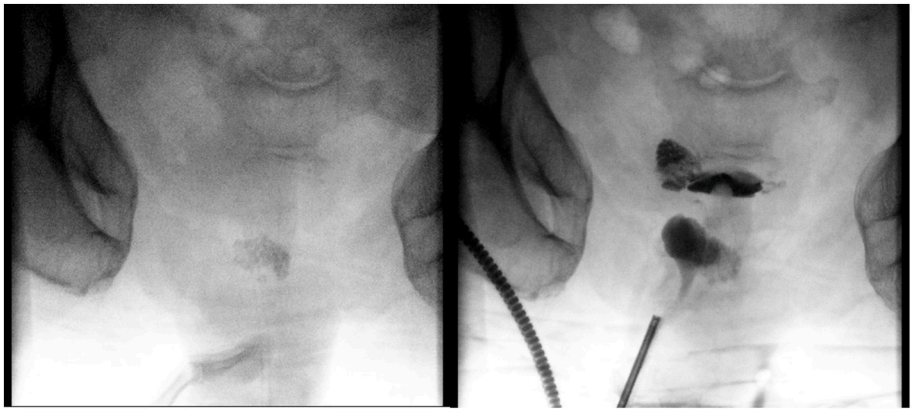

Fig. 2. Retrograde Urethrogram pre-contrast (left) and post-contrast (right).

guidance. The retrograde urethrogram showed no leakage of contrast, and minor passage of contrast into the seminal vesicles and possibly the prostate (Fig. 2). No bleeding source or prostatic calculi were identified.

Approximately a month after the consult following the cystoscopy, the patient presented once again with recurrent complaints of intractable bleeding, discharge, and recurrent prostatic infections. After thorough discussion of treatment options, the patient decided to undergo open retropubic radical prostatectomy for a more long-term solution.

2.3. Surgical management

The patient was positioned supine, and it was observed that there was pus-like fluid draining from the epispadic meatus. A lower midline incision was made from the base of the phallus up to the mid-lower portion of the abdomen. The incision was extended to the fascial layer, allowing identification of the prostatic tissue.

The linea alba was cut and with blunt dissection the retropubic space was entered. Surgical exposure of the prostate was performed by incising the endopelvic fascia, dissecting the lateral aspects of the puboprostatic ligaments and ligating the dorsal vein complex. The lateral prostatic pedicles were each controlled with figure of eight sutures.

Next a transverse capsulotomy was performed and blunt dissection of the ventral aspect of the adenoma was performed. The anterior commissure was sharply divided from the area of the bladder neck to the apex. This separated the lateral lobes of the prostate anteriorly and the prostatic urethra was visible. The mucosa over the lateral lobes was incised and the lobes were bluntly dissected and removed.

Subsequently, the prostatic urethra was entered and incised. No stones or other abnormalities were identified within the prostatic urethra. Once the prostate was completely free, it was carefully passed off the table as a separate specimen (Fig. 3). Hemostasis was achieved, and no obvious injury to the surrounding structures was observed.

The patient tolerated the procedure well, and the fascia was closed with simple interrupted #1 PDS sutures. The skin was closed using staples. The pathology report for the prostate specimen showed no evidence of cancer, with only areas of local inflammation identified in the prostate and seminal vesicles. The patient was discharged on the same day, and during the 10th-day post-surgery follow-up, he reported good appetite, regular bowel movements, well-healing incisions (Fig. 4), minimal pain at the incision sites, and comfortable rectal voiding. As of our most recent 90-day follow-up with the patient, there have been no reported episodes of urethral discharge, gross hematuria, or urinary tract infections.

3. Discussion

Bladder exstrophy (BE) is a rare congenital malformation that presents as an open bladder and urethral plate at birth. Given that BE is part of the bladder exstrophy-epispadias complex, patients will typically also present with defects in their abdominal wall and epispadias due to improper formation of the pelvic floor and genital organs, respectively.[5,6] The closure and management of BE was first described in 1906 by Trendelenburg.[7] During this era, BE patients experienced a profound decrease in life expectancy. A 1926 study by Mayo and Hendricks estimated that 50% of children died before reaching the age of 10. Today,

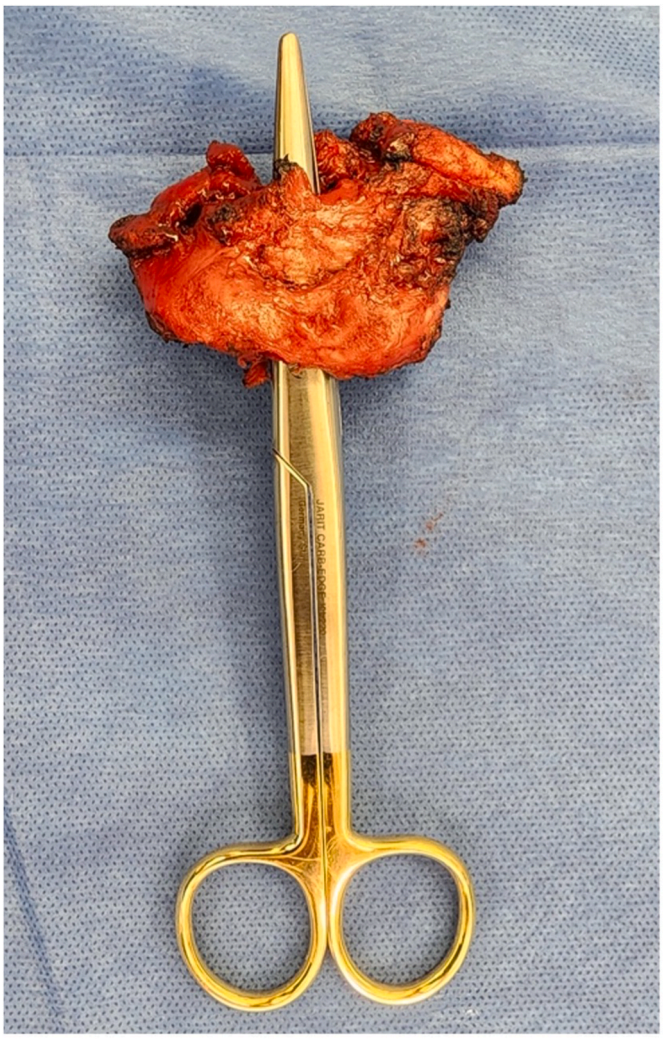

Fig. 3. Image of prostate after its removal.

however, advances in the medical and surgical management of BE have allowed for many patients to experience a normal life expectancy.[5,8]

Ureterosigmoidostomy was the first established urinary diversion surgery to provide continence to patients with BE. The ureters are tunneled into the tinea of the sigmoid colon, and the anus serves as the continence mechanism for both urine and feces. This approach is generally well tolerated, and anal continence is usually achieved.[9] However, urinary tract infections (UTI) remain a frequent complication for patients with BE. In a survey among U.S. bladder exstrophy patients 88% of respondents reported a history of UTIs. In addition, 71% of patients reported a history of recurrent UTIs, which the authors defined as $\geq$ two self-reported UTIs in the last six months or $\geq$ three self- reported UTIs in the past twelve months. The majority of patients reporting recurrent UTIs had undergone urinary tract reconstruction with an intestinal segment. The authors attribute this finding to continuous mucus production from intestinal segments, which may serve as a source for infection if not cleared from the urinary tract.[8]

Radical retropubic prostatectomy has been previously performed in a patient with BE and prostate cancer by Berkowitz et al., thereby demonstrating its safety and viability in this patient population.[6] Moreover, this prostatectomy technique is not unique to the management of the symptomatology that our patient presented with, but rather has a broader range of indications for its usage.[2–4] To the best our knowledge, there is no report of retropubic prostatectomy in the exstrophy-epispadias population for benign disease and the treatment of intractable hematuria and urethral discharge. Following retropubic prostatectomy, our patient's symptoms improved and there were no further reports of gross hematuria or discharge. These findings suggest that this surgical technique may be a viable option for management similar symptoms in BE patients in the setting of benign disease.

The size and appearance of the prostate gland among patients with BE was investigated by Hamper et al. The authors confirmed the presence of the prostate and seminal vesicles in this patient population through the use of transrectal ultrasonography. More importantly, the authors reported a range of prostate shapes, sizes, and configurations in patients with BE. They believe this variability may result from staged reconstruction surgery as well as inherent defects in patients' cloacal membrane.[10] Gearhart et al. performed a study using magnetic resonance imaging (MRI) to examine the location and size of the prostate in men with BE. MRI revealed that the urethra was located anteriorly to the prostate in these patients, and the prostate did not wrap

Fig. 4. Presurgical (left) and postsurgical results (Middle) and follow-up (right).

circumferentially around the urethra.[11]

4. Conclusion

Bladder exstrophy is a rare congenital malformation that is traditionally associated with poor life expectancy. However, advances in surgical repair now allow patients with BE to live into late adulthood. For this reason, adult urologists must be prepared to evaluate and manage urologic problems among these patients. Here, we present the successful management of hematuria, urethral discharge, and recurrent prostatic infection in an adult patient with BE through subtotal retropubic prostatectomy.

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Consent

Written informed consent was obtained from the patient.

Approval of the research protocol by an institutional reviewer board

Not applicable.

Informed consent

Informed consent for publication was obtained from the patient.

Registry and the registration no. of the study/trial

Not applicable.

Authors' contributions

MP evaluated and treated the case, collected all the clinical and radiological data. FL helped collect the clinical data and evaluated the patient. JSAV, DW, and CC all contributed to writing the manuscript. All authors read and approved the final manuscript.

Declaration of competing interest

The authors declare no conflicts of interest.

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None.

Abbreviations

BE	Bladder Exstrophy
BEEC	Bladder Exstrophy-Epispadias Complex
MRI	Magnetic Resonance Imaging
CT	Computerized Tomography

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