Urology Case Reports 38 (2021) 101686

Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: http://www.elsevier.com/locate/eucr

Pediatrics

Transvesical robotic excision of a Müllerian duct remnant

Laura C. Kidd [a], Chinonyerem Okoro [a], Michael Packer [b], Gregory Dean [b], Daniel D. Eun [a,*]

[a] Department of Urology, Lewis Katz School of Medicine at Temple University, 3401 N. Broad St., Philadelphia, PA, 19140, USA
[b] St. Christopher's Hospital for Children, 160E. Erie Ave., Philadelphia, PA, 19134, USA

ARTICLE INFO

Keywords:
Mullerian duct remnant
Robotics
Transvesical

ABSTRACT

Müllerian duct remnants are rare and found in patients with disorders of sexual development. Presenting symptoms vary and many parents opt for surgical management. Literature on robotic repair is limited to small series, single case reports and all were approached extravesically. We present our case of a unique transvesical approach. Perioperative parameters were favorable with no complications, suggesting robotic repair is a safe and effective treatment strategy for these unique patients.

Introduction

Persistent Müllerian duct syndrome (PMDS) is a disorder of sexual development (DSD) that affects virilized 46-XY males, and results from a mutation that either inactivates anti-Müllerian hormone (AMH) or its receptor. This allows persistence of the Müllerian derivatives (i.e. uterus, fallopian tubes, proximal vagina) that drain into a prostatic utricle. PMDS is associated with testicular abnormalities, usually bilateral or unilateral cryptorchidism or transverse testicular ectopia. Mixed gonadal dysgenesis (MGD) can present similarly, but involves genital abnormalities, such as hypospadias.[1]

These Müllerian duct remnants (MDR) can become symptomatic and prompt intervention, as was the case with the following patient, who underwent robotic excision of their MDR. Notably, this was done via a transvesical approach. To our knowledge, this is the first description of this technique in the literature.

Case presentation

Our patient was a 17-year old male with left renal agenesis, otherwise healthy, who was initially worked up for back pain and dysuria. Initial ultrasound suggested possible rhabdomyosarcoma of the prostate. Laboratory workup was negative and subsequent MRI showed a 4.3 cm MDR with no discernible plane between the lesion and the bladder (Fig. 1). Cystoscopy also confirmed a Müllerian duct remnant.

The patient and his parents were thoroughly counseled on management options, and they opted to proceed with robotic repair. His mother expressed significant concern regarding the patient's future erectile and fertility potential. For that reason, and based on the pre-operative imaging anatomy, we decided to attempt a transvesical approach, to minimize manipulation of the neurovascular bundle, seminal vesicles and vasa deferentia.

On the day of surgery, cystourethroscopy showed obvious indentation from the mass on the floor of the bladder. The urethral opening to the MDR was not cannulated, as decompressing the remnant might make dissection more difficult.

We then switched to the robotic portion of the case. Veress insufflation was achieved. After periumbilical camera port placement, the patient was placed in steep Trendelenburg and the remainder of the ports were placed as shown in Fig. 1. A 3cm midline full-thickness cystotomy was made over the bladder dome and retraction was provided bilaterally by two Keith needles, each passed in from the outside abdominal wall, through the lateral bladder leaflet, then passed back out again to the bedside assistant. The left ureteral orifice was congenitally absent. The right ureteral orifice was identified and avoided during the case. The mucosa was incised over the mass and a subcapsular dissection plane was developed and carried down distally between the MDR and the bladder. The neck was amputated, and the bladder floor defect was closed in a double layer with 3-0 barbed absorbable suture. The original bladder dome cystotomy was closed in a similar fashion, and a leak test was negative. EBL was 25 cc and total operative time was 125 min. There were no complications and the patient's catheter was removed three weeks post-operatively. Final pathology was a Müllerian duct cyst. At last follow-up, the patient was reported to have good erectile function.

* Corresponding author. 255 South 17th St., Medical Tower, 7th Floor, Philadelphia, PA, 19103, USA.
E-mail address: daniel.eun@tuhs.temple.edu (D.D. Eun).

https://doi.org/10.1016/j.eucr.2021.101686
Received 24 March 2021; Received in revised form 12 April 2021; Accepted 15 April 2021
Available online 19 April 2021

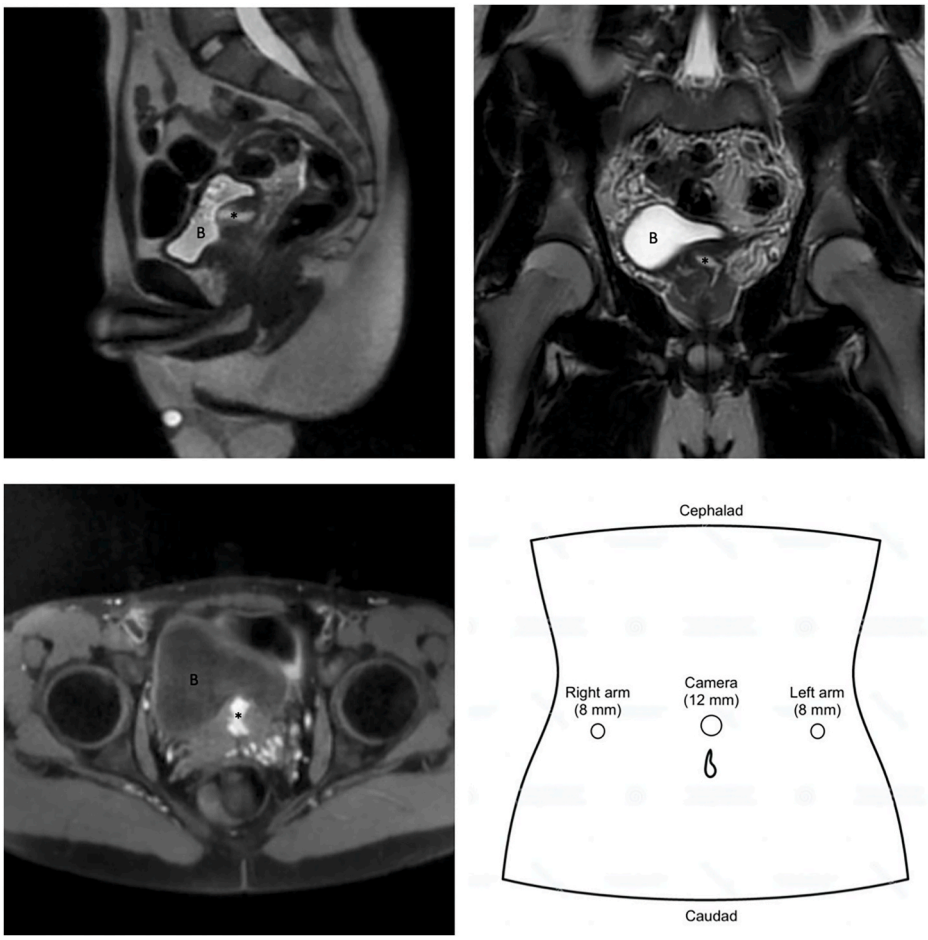

Fig. 1. Pre-operative T2-weighted sagittal (upper left) and coronal (upper right) MRI showing 2.4 × 2.8 × 4.3 cm Müllerian duct remnant (*) with no clear tissue plane between the margin of the lesion and the bladder (B); T1-weight axial MRI showing same lesion (lower left); Robotic port placement for case (lower right). Source image: www.dreamstime.com.

Discussion

Müllerian duct remnants are rare and presenting symptoms can vary. These are also frequently discovered in tandem with other stigmata of DSD, such as undescended testicle or hypospadias, for which providers should be vigilant. Surgical excision[2] is often elected for and this can be done safely with robotic assistance and minimally invasive techniques. Given the rarity of this condition, literature on the subject of robotic repair of MDR is limited to less than ten case series or single-patient reports.[3,4] All of these detail an extravesical approach. To our knowledge, there are no existing reports of a transvesical robotic MDR excision in the literature. However, we show that a transvesical approach may be ideal, depending on pre-operative anatomy, or above-average concern for fertility and erectile function (especially given the higher incidence of testicular anomalies in this patient population). In our experience, our transvesical approach was congruent with the existing published reports, which demonstrate favorable perioperative parameters and patient outcomes.

Of note, data is similarly limited on Müllerian Duct cysts, which can be variable in size depending on their fluid content. Theoretically if these were distended to the point of indenting the bladder, a precise dissection plane could be developed that might make them amenable to a transvesical approach. If not, they would likely be best served in a standard retrovesical fashion.

Conclusion

Müllerian duct remnants are associated with other disorders of sexual development and are rare. Presenting symptoms vary, and can include pain, hematuria, or lower urinary tract symptoms, among others. In the limited literature, only an extravesical approach has been published, but we present favorable outcomes with a unique transvesical approach that is safe, effective, and may be an ideal option for suitable candidates.

Acknowledgements

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

References

1. Picard JY, Cate RL, Racine C, Josso N. The persistent Müllerian duct syndrome: an update based upon a personal experience of 157 cases. *Sex Dev*. 2017. https://doi.org/10.1159/000475516.
2. Almousa S, Alqasem S, Albraidi H, Alfadhel S, Alfakhri A, Moazin M. Robot-assisted hysterectomy in a 41-year-old male: a rare case report. *Urol Case Rep*. 2020. https://doi.org/10.1016/j.eucr.2020.101121.
3. Khoder WY, Kretschmer A, Gratzke C, Becker A, Stief C. Robotic-assisted excision of giant prostatic utricular cysts: technique, outcomes and follow-up. *Surg Technol Int*. 2019.
4. Ferong K, Waterschoot M, Sinatti C, et al. Rare and special robotic surgery indications in the pediatric population: ectopic organs and differences of sexual development. *World J Urol*. 2020. https://doi.org/10.1007/s00345-019-02913-5.