Partially Obstructed Longitudinal Vaginal Septum Presenting in Adulthood With Complaint of Urinary Incontinence

Ariana L. Smith, Thomas F. Kolon, Douglas A. Canning, and Steven J. Weissbart

A longitudinal vaginal septum can be obstructive or nonobstructive. We report on an adult woman who presented with involuntary fluid loss per vagina and had a partially obstructive longitudinal vaginal septum. A 36-year-old nulliparous female presented with malodorous, clear, leakage per vagina that she described as "urinary incontinence." Examination revealed a fluid-filled fluctuant anterior vaginal wall with a draining sinus. Imaging revealed a solitary right kidney with duplicated ectopic fluid-filled ureters inserting into a partially obstructed left hemivagina with a longitudinal vaginal septum. A longitudinal vaginal septum may present in adulthood with the complaint of urinary incontinence. UROLOGY 124: 302–305, 2019. © 2018 Elsevier Inc.

A longitudinal vaginal septum is the result of incomplete fusion of the Müllerian ducts and may be obstructive or nonobstructive in nature. Müllerian anomalies can be associated with uterine and cervical septa, uterine didelphys and urinary tract anomalies.[1] Obstructed hemivagina and ipsilateral renal agenesis is a rare congenital anomaly that generally presents at menarche. Classically, obstructed hemivagina and ipsilateral renal agenesis involves renal agenesis but more recently it has been proposed that the condition also include ipsilateral renal anomalies such as dysplastic kidneys or ectopic ureters.[2]

CASE

A 36-year-old nulliparous female with known left renal agenesis presented with episodic malodorous, clear, thin drainage per vagina of 2 months duration. She reported regular monthly menstrual cycles since menarche, annual gynecologic examinations with Papanicolaou (PAP) smears and tampon use. She was sexually active and denied dyspareunia. She described the drainage as "urinary incontinence" but could not categorize it as either stress-related or urgency-related leakage. She was using 1 incontinence pad per day that ranged from damp to very wet. Pelvic examination demonstrated a 9 cm in length vagina of normal caliber with a fluid-filled fluctuant anterior vaginal wall. Clear vaginal drainage was expressed with compression of the anterior vaginal wall and seen emanating from a 1 mm sinus tract anterior to the cervical os in the proximal vagina. MRI of the pelvis demonstrated a solitary right kidney and 2 fluid-filled ectopic left ureteral remnants inserting into a cystic structure anterior to the vagina (Fig. 1).

The patient was taken for robotic-assisted laparoscopic removal of the fluid-filled left ectopic ureteral segments. Cystoscopy had confirmed absence of an orthotopic left ureteral orifice. Robotically, a single dilated ureteral structure was identified, and it was dissected from the vagina to superior to the iliac vessels where it ended blindly. Pathology of the resected ureter confirmed benign urothelial mucosa with a smooth muscle wall.

Post operatively the vaginal drainage was unchanged, and the patient was referred to a pelvic reconstructive specialist (A.L.S.) for further evaluation and treatment.

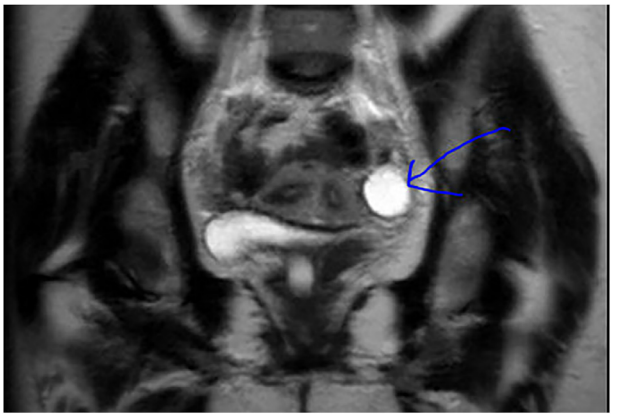

Figure 1. Pelvic MRI with fluid-filled ectopic ureteral segment. (Color version available online.)

Financial Disclosure: The authors declare no competing interests.

From the University of Pennsylvania, Division of Urology, Philadelphia, PA

Address correspondence to: Ariana L. Smith M.D., University of Pennsylvania, Division of Urology, 800 Walnut Street, 19th Floor, Philadelphia, PA 19107.
E-mail: Ariana.smith@uphs.upenn.edu

Submitted: September 28, 2018, accepted (with revisions): November 13, 2018

https://doi.org/10.1016/j.urology.2018.11.018
0090-4295

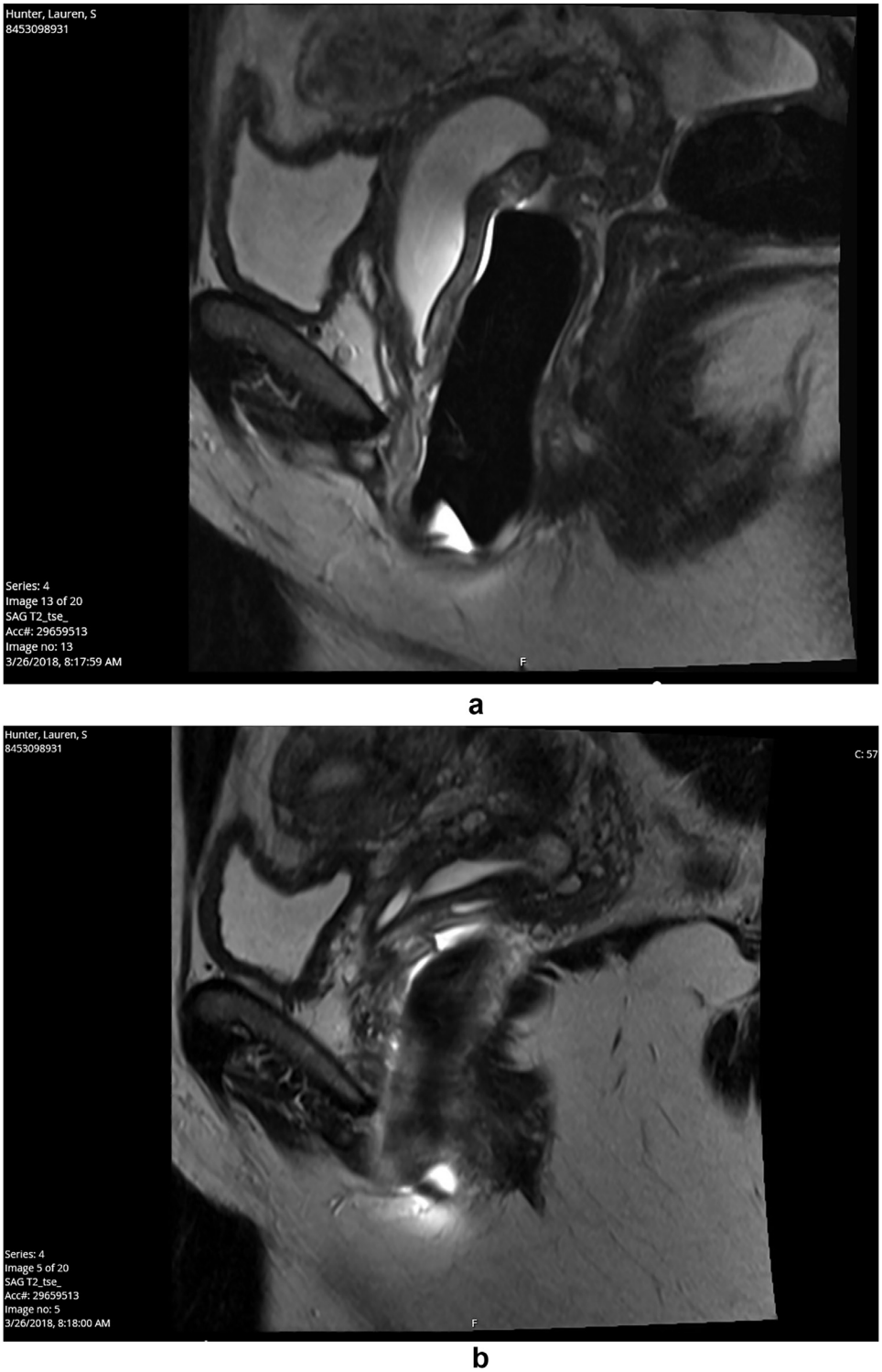

Figure 2. (**a**) Dilated and partially obstructed left hemivagina seen anterior to the right hemivagina. Distal ectopic ureteral segment seen between bladder and partially obstructed distal left hemivagina. (**b**) Fluid-filled ectopic left ureteral segment in the left hemipelvis.

Examination confirmed a fluctuant fluid-filled anterior vaginal wall with copious drainage of clear thin fluid from a 1 mm ostium located on the proximal anterior vaginal wall. An endovaginal coil MRI was obtained to define anatomy.

Imaging revealed a fluid-filled ectopic ureteral segment inserting into an obstructed left hemivagina anterior to the normal caliber right hemivagina (Fig. 2a, b). No left kidney or residual nephrons were visualized; a midline uterine and cervical septum was seen. Surgical intervention

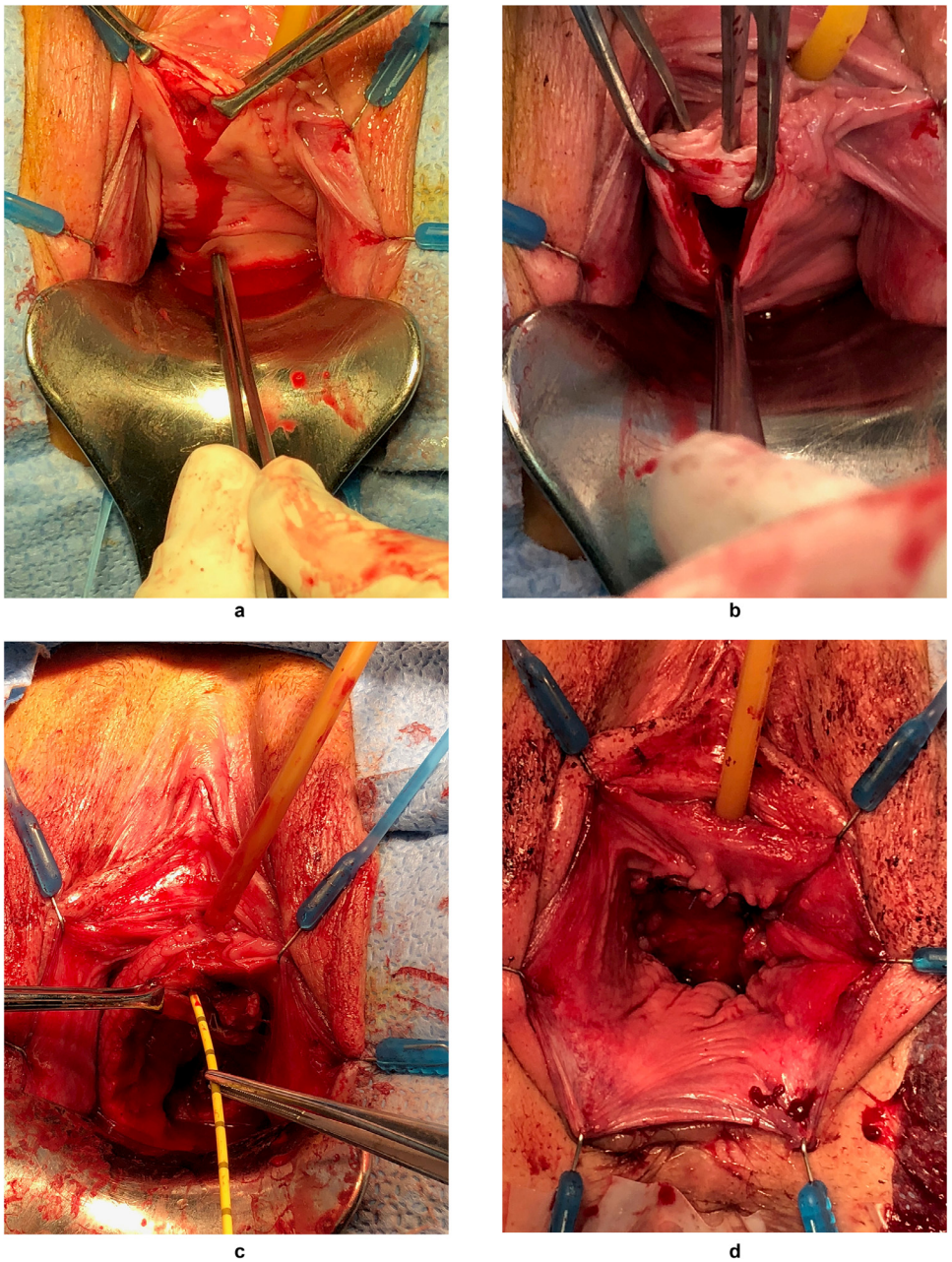

Figure 3. (**a**) Opening of the partially obstructed left hemivagina distally (at the Allis clamps, superiorly) and identification of the 1 mm draining sinus in the proximal vagina (at the Debakey forceps, inferiorly). (**b**) Opening of the fluid-filled left hemivagina corresponding to the fluid-filled cavity on MRI. (**c**) Yellow ureteral catheter entering the ectopic left ureteral segment and advancing 12 cm into the pelvis. (**d**) Complete takedown of the vaginal septum exposing the anterior vaginal wall and 2 cervices at the apex of the vagina. (Color version available online.)

included complete takedown of the vaginal septum, intubation of the left ectopic ureter in the left distal hemivaginal wall, and transvaginal dissection and removal of the left ectopic ureteral segment (Fig. 3). Postoperatively, the vaginal drainage ceased.

COMMENT

The adult presentation of a previously unrecognized and asymptomatic partially obstructed vaginal septum with new onset vaginal drainage is uncommon. This report describes an interesting case of duplicated, ectopic fluid-filled ureteral segments (with ipsilateral renal agenesis) inserting into a partially obstructed fluid-filled hemivagina. Development of a sinus tract from the obstructed vaginal unit produced drainage perceived by the patient as urinary incontinence.

In the absence of a renal unit, the source of the fluid is likely liquefied vaginal and cystic secretions refluxing into the retained ureteral segments.

Müllerian anomalies are rare and affect approximately 3%-6% of girls.[3] The female reproductive tract develops through a series of complex processes including fusion of the left and right Müllerian tracts. Congenital structural anomalies rarely occur but when present may be associated with congenital Wolffian anomalies.[1] Unilateral renal agenesis is associated with Müllerian anomalies up to 30% of the time yet no formal screening guidelines exist.[4] The issue of possible Müllerian anomalies is complicated by the fact that subtle uterine abnormalities are often not seen well by ultrasound in infancy. Furthermore, early evaluation of the pelvis by MRI usually requires general anesthesia. Thus, renal anomalies are frequently identified during routine antenatal ultrasound while Müllerian anomalies often go undetected until menarche or the development of obstructed menstrual symptoms. An early pelvic ultrasound in infancy is reasonable but the recommendation should be that patients with upper urinary tract anomalies, especially renal agenesis, require close follow-up and/or evaluation into puberty. In the presence of a solitary kidney, a high index of suspicion for congenital anomalies of Müllerian fusion should exist, especially when symptoms of vaginal drainage are present. As a field, urology has begun to appreciate a gap in care that patients with congenital urologic conditions experience as they transition from children to adults.[5] Although there is no high level evidence supporting how individuals with congenital upper tract anomalies should be monitored, our case serves as an important reminder that congenital anomalies can present in adulthood. Patients with congenital upper tract anomalies may be served by continued urologic evaluation into young adulthood and have their renal function, blood pressure, and urinary symptoms routinely assessed. As the field of transition urology and congenitalism develops, one may expect future research that identifies an optimal and cost-effect approach for monitoring individuals with congenital urologic conditions.

References

1. Breech LL, Laufer MR. Müllerian anomalies. *Obstet Gynecol Clin North Am*. 2009;36:47.
2. Schlomer B, Rodriguez E, Baskin L. Obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome should be redefined as ipsilateral renal anomalies: cases of symptomatic atrophic and dysplastic kidney with ectopic ureter to obstructed hemivagina. *J Pediatr Urol*. 2015;11:77e1–77e6.
3. Burgis J. Obstructive Mullerian anomalies: case report, diagnosis, and management. *Am J Obstet Gynecol*. 2001;185:338e44.
4. Friedman MA, Aguilar L, Heyward Q, Caldamone A. Screening for Mullerian anomalies in patients with unilateral renal agenesis: leveraging early detection to prevent complications. *J Pediatr Urol*. 2018;14:144–149.
5. Brandes SB. The practice of transition urology and congenitalism. *Curr Opin Urol*. 2016;26:342–343.