A Curious Case of Continuous Incontinence

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Obstructed hemivagina, ipsilateral renal anomaly (OHVIRA) is a rare Müllerian duct abnormality with renal manifestations. Classical presentation is an adolescent female with abdominal pain following menarche. Because of its rarity, diagnosis is often delayed. There have been <20 reported cases of prepubertal OHVIRA. We present a case of missed OHVIRA diagnosis that presented with continuous incontinence following incision of the obstructed hemivagina and discuss the current medical literature on this subject. UROLOGY 92: 113–116, 2016. © 2016 Elsevier Inc.

Obstructed hemivagina, ipsilateral renal anomaly (OHVIRA), closely related to Herlyn-Werner-Wunderlich syndrome (uterine didelphys, obstructed hemivagina, ipsilateral renal agenesis), is a Müllerian duct abnormality first described in 1922.[1] Classically, this presents after menarche with severe dysmenorrhea and a pelvic mass due to hematocolpos.[1] Because of the rarity of this condition, there is often a delay in diagnosis.[1] There are very few reported cases of prepubertal OHVIRA. We outline a case of missed OHVIRA diagnosis in a 9-year-old girl, which presented after developing continuous incontinence after incision of the obstructed hemivagina.

CASE PRESENTATION

A 9-year-old female presented to an outside emergency room with abdominal pain. Abdominal ultrasound (US) showed hydrocolpos and an absent left kidney. The working diagnosis was imperforate hymen with blockage of menses, and she underwent an incision of the suspected obstructing hymen. Clear fluid was drained. After the procedure, she had continuous vaginal leakage and was referred to pediatric urology with a computed tomography (CT) urogram.

Upon referral, she was otherwise healthy with a normal prenatal and birth history. She weighed 30 kg. Genitourinary exam showed Tanner 1 external genitalia; a normal urethra; pooling of clear fluid within the vagina; an intact, although perforate hymenal ring; and a deeper area of tissue within the vagina that had previously been incised. There were no abdominal masses. Creatinine of the fluid pooled in the vagina was 2.5 mg/dL. Working diagnosis was either a postoperative fistula or an ectopic ureter.

The CT urogram demonstrated a normal right kidney, left hydroureter posterior to the bladder, and an atrophic ectopic left kidney. Delayed imaging demonstrated normal excretion of contrast on the right and filling of the left distal ureter with contrast. Müllerian anatomy consisted of uterine didelphys, with the left vagina and uterus showing moderate enlargement in the absence of hydrocolpos (Fig. 1). To better define her anatomy, a cystogram and genitogram and dimercaptosuccinic acid (DMSA) scan were performed. These tests demonstrated a normal bladder without vesicoureteral reflux or fistula and 2 vaginas and uteri without evidence of fistula or reflux into the urinary system. The DMSA scan demonstrated a minute focus of the tracer in the left pelvis.

Based on our clinical evaluation, we suspected the diagnosis of OHVIRA with urine now draining from the left kidney through an ectopic insertion into the recently unobstructed left hemivagina. Management options were discussed with the family, who expressed an interest in management that would eliminate her incontinence and obviate any long-term follow-up of the urinary system. Based on this discussion, we proceeded to the operating room for presumptive left nephroureterectomy.

On cystoscopy, there was an absence of the left hemitrigone and only the right ureteral orifice was identified (Fig. 2). On vaginoscopy, we identified 2 vaginas and 2 cervices. We could not identify an ectopic ureteral orifice within either vagina. Robotic-assisted laparoscopy was conducted via an 8.5 Fr camera port at the umbilicus and 8 Fr robotic trocars in the midclavicular line on each side of the umbilicus. A 5-mm assistant trocar was placed in the right upper quadrant. We identified the dilated left ureter within the pelvis with insertion into the left vagina (Fig. 3A). Proximally, there was an atrophic pelvic kidney with anomalous vessels (Fig. 3B). We performed a left nephroureterectomy. The distal ureter was amputated at its entry point into the vagina. The specimen was extracted through the umbilical camera port (Fig. 3C). The patient recovered uneventfully and went home on postoperative day 1. She was immediately dry after the procedure.

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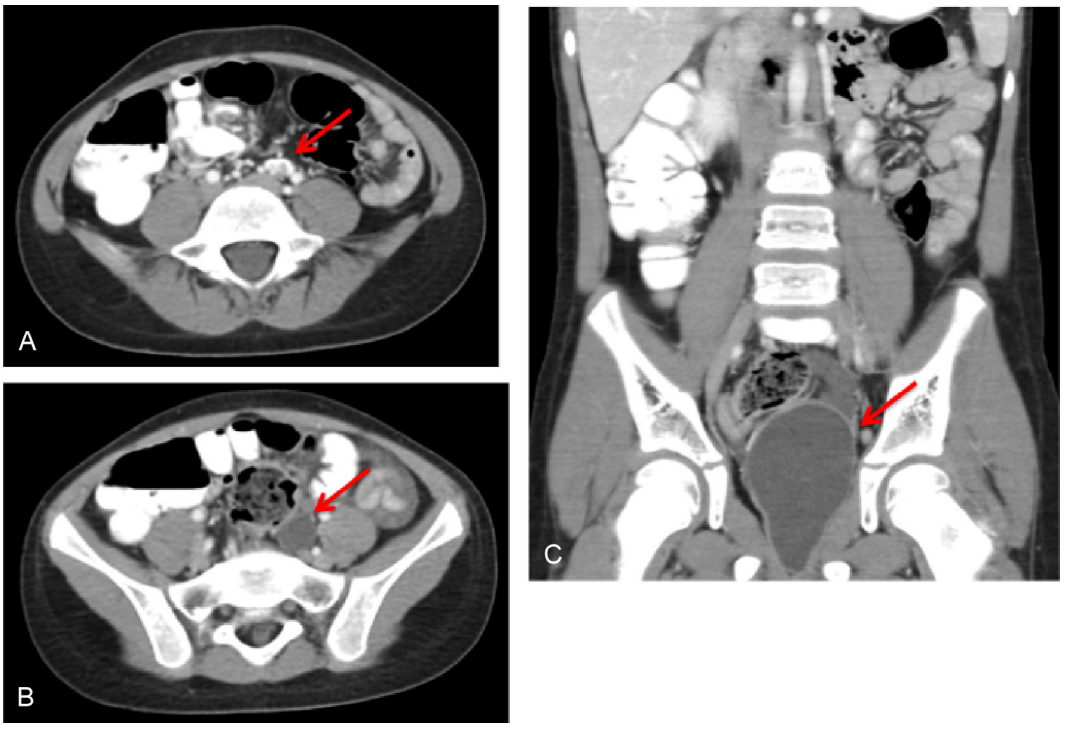

Figure 1. CT scan showing contrast enhancing left pelvic renal remnant (arrow, **A**), dilated left ureter (arrow, **B**), dilated left hemivagina (arrow, **C**). (Color version available online.)

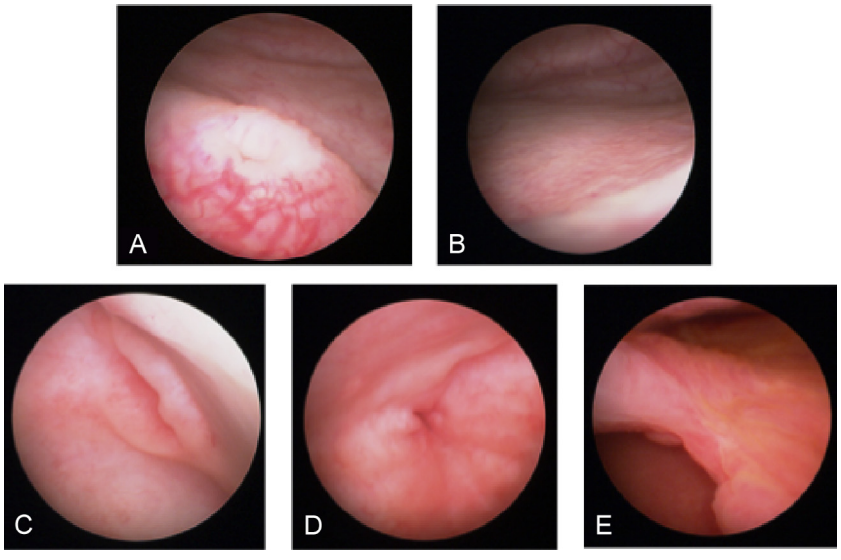

Figure 2. Cystoscopy revealed a normal right ureteral orifice **(A)** and absent left hemitrigone **(B)**, vaginoscopy revealed 2 cervices **(C,D)** and an incised vaginal septum **(E)**. (Color version available online.)

DISCUSSION

Obstructive Müllerian duct abnormalities have a prevalence of 2%-3%,[1] with 30%-43% of these having associated renal anomalies.[1,2] It is estimated that 1/20,000 females has OHVIRA.[3] Although rare, OHVIRA should be considered in any young female with a pelvic mass and dysmenorrhea[1] or a pelvic mass and any renal anomaly.[2] In the prepubertal female, diagnosis can sometimes be made perinatally with unilateral renal agenesis seen on prenatal imaging and vaginal bulge identified after birth.[4] Rarely do girls present between the newborn and pubertal period as our patient did.

There have been variations to the classic Herlyn-Werner-Wunderlich description, including a single uterus,[5] septate uterus, urogenital sinus,[6] multicystic dysplastic kidney with or without previous nephrectomy,[7] and ectopic remnant ureters.[8] Because of these variations, OHVIRA has been modified to include "anomaly" instead of "agenesis" as a