Laparoscopic Diagnosis and Management of Bilateral Uterine Remnants

Sahar M. Stephens, MD, Kristi A. K. Maas, MD, Ruben Alvero, MD, Samuel Chang, MD, Laxmi A. Kondapalli, MD, MSCE

Department of Obstetrics and Gynecology, University of Colorado Denver, Aurora, CO, USA (Drs. Stephens, Alvero, Kondapalli).

Department of Obstetrics and Gynecology, ME Medical Center, Tufts University School of Medicine, Portland, ME, USA (Dr. Maas).

Department of Radiology, University of Colorado Denver, Aurora, CO, USA (Dr. Chang).

ABSTRACT

Introduction: Müllerian anomaly is a result of abnormal elongation, fusion, canalization, or resorption of the paramesonephric ducts during organogenesis. An accurate diagnosis and appropriate treatment planning can be facilitated by imaging modalities; however, direct visualization of the pelvic organs may be necessary for an accurate diagnosis.

Case Description: A 14-year-old girl with primary amenorrhea presented with severe abdominal pain. Magnetic resonance imaging suggested a unicornuate uterus on the right side with a left-sided noncommunicating uterine horn, both with functional endometrium and likely high outflow obstruction. She was counseled on removal of the noncommunicating uterine horn and correction of the outflow obstruction; however, on laparoscopic and vaginoscopic exploration, she was found to have bilateral noncommunicating functional uterine remnants with a normal-length vagina and a septate hymen. She underwent laparoscopic removal of uterine remnants with complete symptom resolution.

Discussion: Preoperative imaging studies can help guide patient counseling and preoperative planning in cases of suspected müllerian anomaly; however, the final diagnosis may not be made until the time of surgery. The patient should be prepared for multiple possibilities during preoperative consultation.

Key Words: Müllerian aplasia, Mayer-Rokitansky-Küster-Hauser syndrome, Laparoscopy.

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Address correspondence to: Laxmi A. Kondapalli, MD, MSCE. 12631 East 17th Avenue B198-3 Aurora, CO 80045. Telephone: (303) 724-6139, Fax: (303) 724-2053, E-mail: laxmi.kondapalli@ucdenver.edu

INTRODUCTION

A defect or error in the elongation, fusion, canalization, or resorption of the paramesonephric ducts during organogenesis in the female fetus results in a müllerian anomaly. Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome affects 1 in 4500 women and is characterized by uterovaginal aplasia combined with normal functional ovaries; however, cases with rudimentary uterine structures have been described.[1–4] Accurate diagnosis of the type of müllerian anomaly present is integral to appropriate patient counseling and management. We describe a case in which magnetic resonance imaging (MRI) failed to accurately characterize the anomaly present highlight the importance of preoperative and intraoperative counseling in the management of these patients.

CASE

A 14-year-old girl was evaluated at our institution for primary amenorrhea, pelvic pain, and possible müllerian anomaly. One month before presentation, she was evaluated in the emergency department for severe pelvic pain. She was diagnosed with suspected transverse vaginal septum, given narcotic analgesics for pain management, and referred to the gynecology clinic for outpatient evaluation.

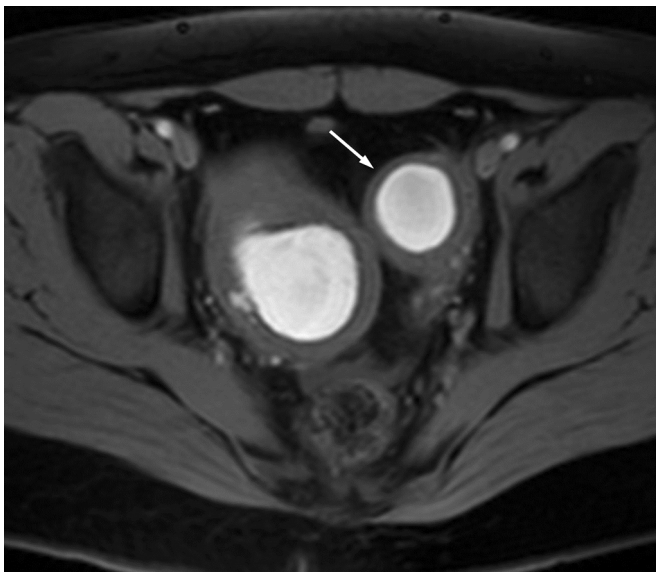

Figure 1. A fat-suppressed T1 Gradient Echo magnetic resonance image shows two separate dilated horns with hematometra seen as bright signal intensity. The smaller left horn (arrow) is not connected either to the right horn or to the vaginal pouch.

She was seen in the general gynecology clinic, where examination was notable for Tanner stage IV breast and pubic hair development. Pelvic examination at that time was limited; however, a transverse vaginal septum was again suspected. Continuous combined hormonal oral contraceptive pills were initiated, and pelvic MRI was ordered. The initial report indicated uterine didelphys with bilateral hematometra and a possible high transverse vaginal septum or cervical stenosis. The patient was subsequently referred to our clinic for further evaluation and treatment.

The patient's medical and surgical history was unremarkable. She reported the onset of breast and axillary hair development at 12 and 13 years of age, respectively. She had never had a menstrual period and had never been sexually active. She had been delivered at term by cesarean section for breech presentation. Her family history was notable for her mother having uterine didelphys and unilateral renal agenesis. Physical examination of our patient confirmed Tanner stage IV breast and pubic hair development; a pelvic examination was not performed.

The magnetic resonance images were reviewed independently and with the radiologist. The right uterine horn appeared to be continuous to the vaginal stump, and the caudal portion of the right horn showed low signal intensity on T2-weighted images that suggested cervical stroma. Imaging appeared most consistent with a unicornuate uterus on the right side with a left-sided noncommunicating uterine horn, and both showed hematometra. The lack of hematocolpos suggested either a proximal transverse vaginal septum or cervical stenosis with outflow obstruction (**Figures 1** and **2**).

Given the patient's significant pelvic pain and presence of hematometra, she and her parents were counseled on surgical management. A variety of surgical approaches were discussed because definitive anatomic diagnosis would be made intraoperatively. She and her parents consented to an examination under anesthesia, possible resection of the vaginal septum or cervical dilation for relief of the unicornuate hematometra, resection of the noncommunicating uterine horn, and other indicated procedures. Preoperatively, she underwent renal ultrasonography, which showed a horseshoe kidney with the connecting bridge lying over the abdominal aorta and measuring 8 mm in anterior-posterior thickness.

Intraoperatively, findings of the examination under anesthesia were notable for a septate hymen, which was surgically corrected. The vaginal canal was approximately 6 cm in length with normal caliber. A speculum examination showed a blind vaginal pouch with absence of a cervix. On bimanual examination, a firm pelvic mass was

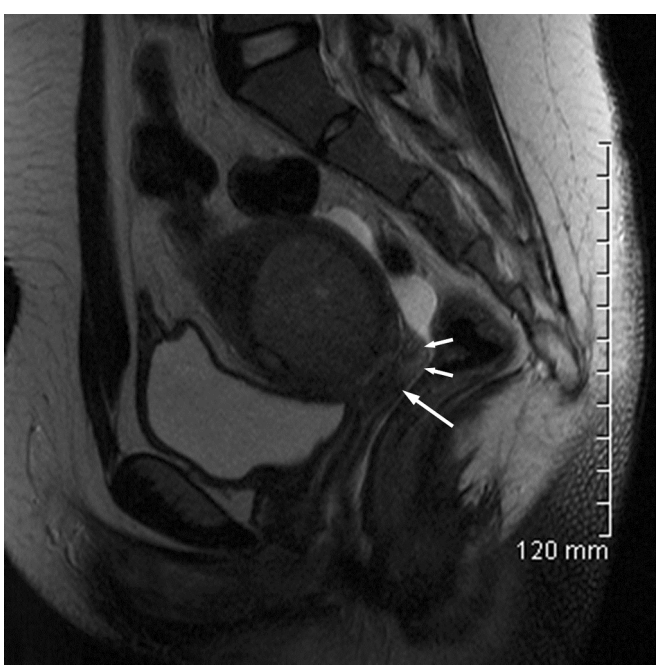

Figure 2. T2 Turbo Spin Echo sequence magnetic resonance image. The vaginal pouch (long arrow) is located relatively low, and the uppermost portion of the vagina is missing. Instead, an ill-defined connective tissue is present (short arrows).

palpable; however, a cervical os could not be felt through the apex of the vagina. Intraoperative transvaginal ultrasonography was performed, and two fluid-filled uterine horns were identified; however, no clear cervix was seen. At this point, a diagnostic laparoscopy was performed to better evaluate the pelvic anatomy.

On laparoscopy, two distinct, completely separate and dilated uterine structures were identified (**Figure 3**). The left uterine structure was spherical and approximately 4.5 cm in diameter; a left hydrosalpinx was also noted. The right-sided uterine structure measured approximately 8 × 6 cm and had a pear-shaped appearance, with the largest diameter at the most caudal portion. Both ovaries and the right fallopian tube were seen and appeared normal. Given the size of the right uterine structure and the MRI findings that suggested the presence of possible cervical stroma, the right uterine structure was thoroughly evaluated for the presence of a cervix to avoid unnecessary hysterectomy in this young patient.

Under direct visualization with the intra-abdominal laparoscope, a small incision was made in the vaginal apex and a hysteroscope was placed through the incision. Transillumination by the hysteroscope could be clearly seen through the peritoneum in the pelvis separate from the lower aspect of both uterine horns. A hysterotomy was made at the fundus of the right uterine horn, the hematometra was evacuated, and a blunt probe was passed into the uterine cavity. The blunt probe was advanced to the inferior border of the right uterine horn. As this was done, a vaginal examination was performed, confirming the absence of a connection between the lower uterine segment and the apex of the vagina. At this point, it was determined that a cervix was not present and that the right uterine structure did not represent a unicornuate uterus but rather a rudimentary horn with functional endometrium.

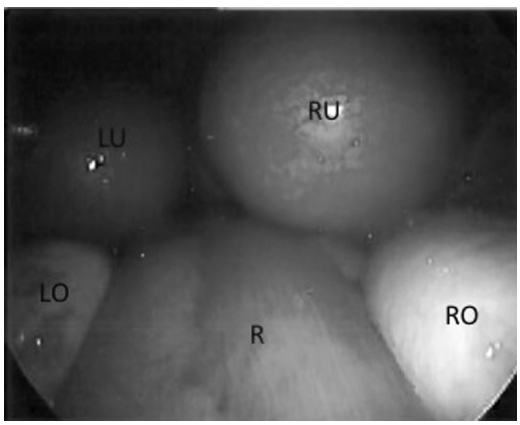

Figure 3. Laparoscopic view of normal ovaries and bilateral uterine horns. LO = left ovary, LU = left uterine remnant, R = rectum, RO = right ovary, RU = right uterine remnant.

While the patient was under anesthesia, her anatomic findings were discussed with her parents. Consent was obtained to perform removal of both uterine horns and the left hydrosalpinx. The uterine horns were surgically released from all attachments and underwent morcellation for removal through the laparoscopic port. The patient tolerated the surgery well and was discharged home the following day.

Pathologic evaluation confirmed the intraoperative diagnosis of bilateral uterine remnant and the absence of any cervical tissue. The final diagnosis was bilateral noncommunicating functional uterine horns, left hydrosalpinx, horseshoe kidney, and septate hymen.

At the patient's 2-week postoperative visit, she required only ibuprofen for pain management, had returned to school, and had resumed her normal activities. The müllerian anomaly and the indications for surgical excision were again discussed with the patient and her parents. Furthermore, we informed them that although the patient herself would be unable to carry a pregnancy, she could have a genetic child with the use of in vitro fertilization and a gestational carrier.

DISCUSSION

In the normally developing female fetus, the müllerian ducts differentiate to form the fallopian tubes, uterus, cervix, and upper vagina, whereas the gonadal ridge gives rise to the ovaries. The developing kidneys and urinary system emerge from the wolffian ducts, which run parallel to the reproductive tract. Between the ninth and 22nd week of gestation, the müllerian tract develops via elongation, fusion, canalization, and resorption.[5] The urogenital sinus develops into the external genitalia and the lower vagina, which must fuse with the caudal portion of the müllerian tract to create a complete outflow tract. Failure or errors in any of these steps may result in anatomic malformations broadly termed müllerian anomalies.

MRKH syndrome is the most common form of agenesis of the müllerian tract and occurs in 1 in 4500 women.[6] Type I MRKH syndrome is characterized by aplasia of the uterus and upper vagina. Although the incidence of rudimentary uterine horns among patients with MRKH syndrome is unknown, several case reports have identified uterine remnants in women with MRKH syndrome undergoing surgery for pelvic pain or mass, often involved with en-

dometriosis or leiomyoma,[1–4] and a recent small study of MRI in women in whom there was a clinical suspicion for MRKH syndrome identified uterine remnants in all patients evaluated.[7] Type 2 MRKH syndrome is associated with anomalies in other organ systems including the urinary tract, skeleton, central nervous system, and heart. Renal anomalies are most common, occurring in nearly 30% of women with MRKH syndrome in 1 study.[8] In that study horseshoe kidney accounted for 5% of the renal anomalies identified.

Our case represents multiple defects in embryologic development from hypoplasia of the müllerian ducts, horizontal fusion defect between the two müllerian ducts, vertical fusion defect between the upper genital tract and the urogenital sinus, and incomplete resorption of the vaginal canal leading to septate hymen.

The wide variety of müllerian anomalies that occur is highlighted by the various proposed classification schemes. Perhaps the most commonly used classification scheme is that of the American Fertility Society (AFS), which was revised in 1988 and is composed of 7 different broad categories encompassing 16 total variants.[9] The AFS classification scheme is inadequate in describing complex anomalies, as shown in our patient, who represents a combination of AFS class I (hypoplasia or agenesis) and class III (didelphys). In addition, the AFS classification system does not include abnormalities of the vagina (aside from vaginal agenesis) or the urogenital sinus.

Several alternative classification schemes have been proposed[10,11]; however, none have effectively replaced the AFS scheme in common practice. The Vagina, Cervix, Uterus, Adnex-associated Malformation classification scheme proposed by Oppelt et al[11] in 2005 uses a structure similar to the TNM classification for oncologic tumors to describe abnormalities of the vagina, cervix, uterus, adnexa, and associated systems. Unfortunately, the VCUAM system is complex, with over 30 unique categories, making it cumbersome to use. Perhaps the classification system published by Acién and Acién[10] in 2011 provides the best balance of complexity and ease of use with 6 general categories (including anomalies of the urogenital sinus) and 19 subcategories. The variety of classification schemes and variable terminology used by radiologists and gynecologists in describing their findings complicate the accurate description of the anomaly present in many patients.[12]

MRI has been found to be useful in the classification of and surgical planning for treatment of müllerian anomalies. Carrington et al[13] showed that 24% of patients evaluated with MRI preoperatively had specific surgical plans formulated as a result of the MRI findings. MRI is considered the gold standard for noninvasive evaluation of pediatric patients, with an 80% to 91% correlation between MRI findings and intraoperative findings.[14,15] In comparison, transabdominal ultrasonography has only a 59% correlation with operative findings.[15] Diagnosis by ultrasonography can be improved if 3-dimensional ultrasonography is used, with a 98% to 100% sensitivity and 100% specificity.[16]

One study found a sensitivity and specificity of MRI for the diagnosis of MRKH syndrome of 100% for both compared with the gold standard of laparoscopic examination. However, when evaluated for the ability to identify müllerian remnants, the sensitivity was only 81% and specificity was 91%.[17] Prior studies conducted at tertiary referral centers evaluating MRI in the diagnosis of suspected müllerian anomaly noted the development of a specific radiologic protocol as more experience was gained, typically including T2-weighted sagittal, coronal, and transverse images and T1-weighted axial images.[12,18] Having a radiologist experienced in interpreting abnormal pelvic anatomy is also important to the accurate characterization of müllerian anomaly.

Although MRI and 3-dimensional ultrasonography are useful for preoperative planning, definitive diagnosis may not be possible until the time of surgical exploration. Even in cases in which the anatomic abnormalities are well demonstrated, the complexity of anomaly may preclude classification into a single category. As shown in our case, it is important to discuss potential limitations of preoperative imaging as well as multiple surgical approaches. Great effort was made to keep the family informed as the true anatomy was elucidated intraoperatively and to review the diagnosis and reasoning for removal of the uterine structures postoperatively with the patient and her family.

Hysterectomy has long been the recommended treatment for noncommunicating uterine remnants with functional endometrium.[19] Uterovaginal anastomosis, a treatment option for women with cervical agenesis, has been associated with high rates of reoperation and complications.[20] Given the rarity of the disorder, evidence for uterovaginal anastomosis is limited to case series and reports, with the largest series involving 30 patients.[20] Recently, a few reports have been published with lower rates of complications and a few successful pregnancies in women with cervical agenesis undergoing uterovaginal anastomosis.[21,22] Although some reports include uterovaginal anastomosis to a unilateral uterine structure, most reported cases involve women with a normal uterine body. Uterovaginal anastomosis was not attempted in our patient because of the absence of a normal uterine body, high

rate of reoperation and complications, and unclear potential for successful pregnancy in a hypoplastic uterine horn. If uterovaginal anastomosis is to be attempted in cases of cervical agenesis, it is recommended that this be performed by a surgeon with prior experience to avoid multiple surgical procedures.

CONCLUSION

The accurate diagnosis and characterization of müllerian anomaly are important for surgical planning if surgery is indicated and can be facilitated by pelvic MRI. However, preoperative imaging has limits, especially in cases of complex müllerian anomaly, and true understanding of the pelvic anatomy may not be obtained until the time of surgery. Patients need to be appropriately counseled regarding the limitations of preoperative assessment and the many possibilities of surgical interventions before surgery.

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