Original Study

Surgical Pearls: Laparoscopic Removal of Uterine Remnants in Patients with Mayer-Rokitansky-Küster-Hauser Syndrome

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A B S T R A C T

Background: Females with Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome may require surgical removal of uterine remnant(s) which can be accomplished with a laparoscopic approach, described in this case series.
Cases: Nine females with MRKH and pelvic pain were treated with laparoscopic resection of uterine remnants without major complication. The following management recommendations are offered: (1) preoperative evaluation for urinary tract anomalies and postoperative cystoscopy; (2) medial traction of the remnant to allow adequate exposure of the pelvic sidewall; (3) awareness of possible anomalous vascular supply to uterine remnant; (4) individualized management of associated endometriosis; (5) careful use of surgical terminology, avoiding use of the word hysterectomy.
Summary and Conclusion: Laparoscopic removal of uterine remnant(s) is safe and effective.
Key Words: Laparoscopy, MRKH, Uterine remnant, Pelvic pain

Introduction

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome, characterized by vaginal agenesis and varying degrees of cervical and uterine agenesis, occurs in approximately 1 in 4000 to 1 in 5000 female births.[1] This syndrome is diagnosed mainly in teenagers (15-18 years old) who present with primary amenorrhea. In those with pelvic pain, magnetic resonance imaging (MRI) is the gold standard to evaluate anatomic variants.[2,3] Recently we identified a 2.3-fold increased risk of pelvic pain in MRKH patients with uterine remnants containing endometrium.[4] If removal of obstructed remnants is planned for surgical therapy of the pain, a laparoscopic approach can be considered.

Laparoscopic resection of uterine remnant(s) poses unique challenges due to the aberrant anatomy, associated urinary tract anomalies, and possible endometriosis with resulting adhesions. The laparoscopic approach, allowing for short recovery and improved postoperative pain and cosmesis, has been used for this surgical procedure, although only case reports have been published.[5–9] The current case series discusses select clinical aspects and surgical findings in 9 patients undergoing laparoscopic resection of uterine remnants, and reviews the unique surgical aspects of this procedure.

Methods

Using current diagnosis codes for “other anomalies of uterus” and “other anomalies of cervix, vagina and external female genitalia” (752.3, 752.0-752.9), we identified all females with MRKH seen in the outpatient setting in the Department of Obstetrics and Gynecology at the University of Michigan Health System from January 1, 2004, to December 31, 2011, when laparoscopy was commonly performed. Surgical and clinic schedules were used to identify MRKH females who might have been missed by the initial search criteria. All patients who underwent surgery for resection of uterine remnants were included. Their electronic medical records were retrospectively reviewed, and information regarding patients’ demographics, clinical presentation, imaging studies, surgical findings, surgical procedures, pathology, and postoperative follow-up was abstracted. The study was approved by the Institutional Review Boards at the University of Michigan.

In our practice, all MRKH females are evaluated with physical exam and renal imaging. Patients with pelvic pain also undergo a pelvic MRI. Their anomalies were classified according to the American Fertility Association (AFS) classification system.[10] Endometriosis was staged intraoperatively according to the AFS revised classification.[11]

Standard laparoscopic technique was employed for surgical removal of the uterine remnants. Pneumoperitoneum was initiated by placement of the Veress insufflation needle in the umbilicus. Additionally, 2 or 3 5-mm accessory laparoscopic ports were placed in the lower abdomen. The vascular pedicles of the uterine remnants were controlled with electrosurgical instruments, using either the PKS cutting forceps (Gyrus ACMI, Southborough, MA), the Enseal tissue sealer, or the Harmonic scalpel (both by Ethicon Endo-Surgery, Cincinnati, OH). In cases where morcellation of the uterine remnant was performed, one of the accessory ports was extended to 12 mm.

Statistical analysis included descriptive analysis. Continuous variables are expressed as median (range).

The authors indicate no conflicts of interest.

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Results

During the study period, we evaluated 33 females with MRKH in the Pediatric and Adolescent Gynecology clinic.[4] Of those 33, 9 (27%) patients subsequently underwent surgery for removal of uterine remnant(s) for pelvic pain. The median age of the patients at the time of surgery was 17 (range, 10-23 y), and the median body-mass index was 25.6 kg/m^2 (range, 20.3-31.3). All patients presented with cyclic (59%) or acyclic (41%) pelvic pain; an extensive review of systems, as well as a complete physical exam, was performed to rule out other sources of pelvic pain. Pelvic MRI studies were obtained in all patients to assess pelvic and abdominal anatomy and to rule out other sources of pelvic pain. Based on the AFS Classification system, all patients had combined agenesis with bilateral hypoplastic uterine remnants (Class Ie). See Fig. 1. Enhancement of the uterine remnant cavity on MRI, suggestive of functional endometrium, was found in 8/9 (89%) females who underwent surgical resection of uterine remnants. The ninth patient, with no endometrium in her remnants on MRI, had severe cyclical pelvic pain and no other cause could be determined. Her pain completely disappeared with administration of 3 months of a GnRH agonist, so the patient was given the option of a surgical removal, which led to the alleviation of her pain.

Prior to surgery, imaging of the urinary structures with MRI, computed tomography, or renal ultrasound was performed in all patients. Three (33%) patients were found to have renal anomalies, including 1 patient with unilateral absent ureter and kidney, 1 with a pelvic horseshoe kidney, and 1 with mild bilateral ureteroceles.

In all patients, the initial surgical approach was laparoscopic. A careful survey of the abdominal and pelvic structures was performed with the laparoscope, evaluating for adnexal structures, pelvic sidewall regions (including identification of the uterine remnant, round ligaments, inguinal rings, vascular structures, and ureters), endometriosis lesions, and adhesions. All 9 patients were confirmed to have bilateral uterine remnants (Fig. 2) and absent cervix. The vaginal length ranged from 1 to 9 cm as determined by pelvic examination, and dependent on stages of dilation or sexual activity.

Five (56%) patients had endometriosis upon visual inspection of the pelvis (2 with stage I, 1 with stage II, and 2 with stage III), with the left side of the pelvis more affected than the right side.[11] Four (44%) patients were found to have significant pelvic adhesions (in 3 of those, concurrent endometriosis and pelvic adhesions were observed).

On inspection of the vascular supply of the uterine remnants, all remnants had identifiable utero-ovarian vessels. However, low lying main inferior vascular pedicles (corresponding to the uterine vessels) were clearly identified in only 6 (67%) cases. In the 3 remaining cases, no discernible main vessel or only small collaterals were observed.

After thorough survey of the pelvic anatomy was completed, the procedure for removal of the uterine remnant was performed (Table 1). The first step was applying medial traction on the uterine remnant using a grasper or tenaculum through the contralateral or the suprapubic port. The round ligament, fallopian tube, and utero-ovarian ligament were then cauterized and transected, similar to the surgical steps undertaken for laparoscopic hysterectomy. The peritoneum was incised and developed anteriorly and posteriorly, allowing for the identification of the course of the ureter and the vascular supply to the remnant. In those 6 patients where a low lying main vascular supply to the remnant was seen, these blood vessels were then skeletonized, cauterized, and transected. Aberrant blood vessels were typically noted in the inferior edge of the remnant consistent with the absence of the cervix, rather than at the lateral aspect. In those cases where the lower edge of the remnant was not clearly identified, the bladder flap was further developed to cross the median

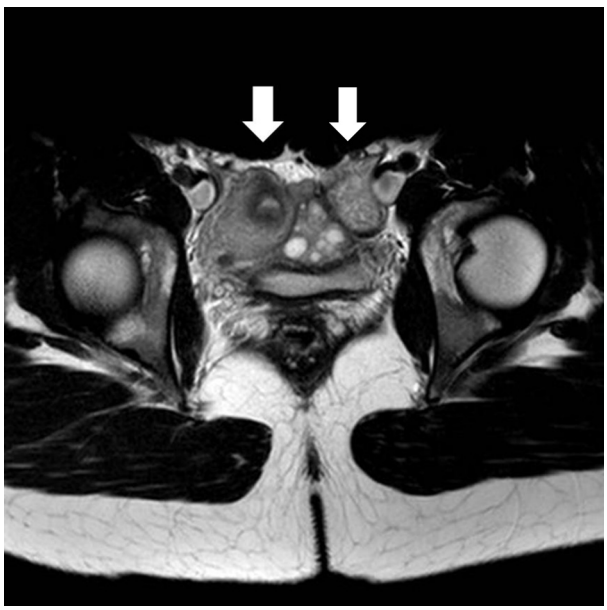

Fig. 1. Pelvic MRI image showing a patient with MRKH syndrome, vaginal agenesis, and hypoplastic bilateral uterine remnants. Arrows point to the uterine remnants. Enhancement of the cavity on T2 indicated functional endometrium, confirmed on pathology.

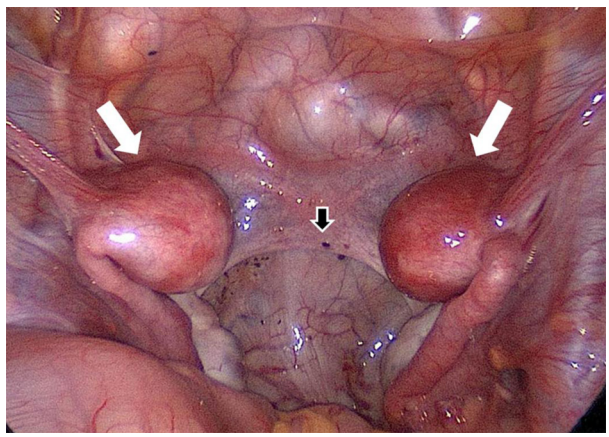

Fig. 2. Laparoscopic image of an adolescent with MRKH syndrome, bilateral uterine cavitated remnants (*large white arrows*), and endometriosis (*black arrow*). An adolescent with MRKH syndrome and bilateral uterine cavitated remnants (*white arrows*), illustrating the lateral displacement of the remnants. The black arrow points to "powder-burn" spots, consistent with endometriosis.

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