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Is MRI Sufficient to Diagnose Rudimentary Uterine Horn?: A case report and review of the literature

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Introduction

Mullerian anomalies have varying presentations, some of which overlap with more common diagnoses. Pelvic pain represents one such presenting symptom and imposes considerable distress. Its broad range of etiologies, both gynecologic and non-gynecologic, makes pelvic pain a frustrating symptom for patients and physicians alike. Proper evaluation often may involve one or more diagnostic imaging modalities, and treatment may be medical or surgical. Though current imaging modalities such as MRI and 3D ultrasound are highly predictive of Mullerian anomalies, it is important to bear uncommon presentations of common pathologies in mind. Herein we report the case of a 21 year old woman with unilateral dysmenorrhea whose 3D ultrasound and MR imaging suggested a Mullerian anomaly. However, at laparoscopy a necrotic fibroid was ultimately diagnosed.

Case Report

A 21 year old, nulligravid, morbidly obese female (BMI of 40) was referred to an academic center for further evaluation and treatment of a possible non-communicating uterine horn identified on ultrasound performed for evaluation of dysmenorrhea. The young woman reported intermittent left-sided cramping and pelvic pain since menarche. Though most often associated with menses, the pain occasionally occurred at other times during the menstrual cycle. The pain had been satisfactorily managed by oral contraceptives from menarche to the current presentation. When the patient discontinued oral contraceptives to

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attempt pregnancy, disabling pain resumed. Her symptoms caused her to miss school and work, and she required narcotics for pain control. She had no significant medical history and no previous surgeries.

On physical exam abdomen was non-tender, and external genitalia was normal. She had a single cervix apparent via speculum. On bimanual exam, no tenderness, fullness, or discrete masses were appreciated; however it was difficult to palpate her uterus and ovaries secondary to body habitus. 2-D and 3-D transvaginal ultrasound revealed rightward-deviated uterus with an adjacent walled structure. The walls of the structure were isoechoic to myometrium and contained hyperechoic material. Endometrium distinct from echogenic contents was not visualized. The right uterine cavity continued to the endocervix and did not communicate with the contents of the left uterine structure.

Subsequent magnetic resonance imaging (MRI) of the pelvis demonstrated a marked deformity of the uterus suggesting partial duplication. Interpreting radiologist had expertise in gynecologic MRI, and the study was reviewed by radiology and gynecology staff during formal interdepartmental conference. A right uterine horn was suggested, leading to a single cervix, with a distorted left horn remnant that appeared to be dilated by hydro-/hematometra (Fig 1). No renal abnormality was noted. Both ovaries appeared within normal limits. The presumed diagnosis of non-communicating horn with hematometra was consistent with the patient's history of longstanding cyclic left-sided pain that resolved with hormonal suppression via oral contraceptive.

The patient was counseled for resection of left uterine horn based upon the radiographic findings. A robotic-assisted laparoscopic approach was planned. The patient underwent diagnostic laparoscopy prior to engaging the robot to confirm the diagnosis and assess the feasibility of excising the horn. Laparoscopy revealed a bulbous contour of the left uterus, but failed to show the pronounced convexity expected from a unicornuate uterus with a rudimentary horn (Fig. 2A). No endometriotic implants were seen. Furthermore, chromopertubation through the patient's single cervix resulted in bilateral spill of methylene blue from the fallopian tubes, which was contrary to the preoperative diagnosis of a left, non-communicating horn. Incision via harmonic scalpel into the serosa overlying the mass revealed underlying intact myometrium superficial to degraded myometrial tissue, from which arose an efflux of chocolate-colored viscous fluid. The mass was drained and tissue was removed and sent to pathology. Chromopertubation was again performed to confirm that the endometrial cavity had not been entered, and the defect was repaired (Fig. 2B). The patient recovered well from laparoscopic myomectomy and was pain free through her first postoperative menses. Histopathologic diagnosis was consistent with leiomyoma.

Discussion

The prevalence of Mullerian duct anomalies in the general population has been reported as 0.1%–3.8%. However, since symptoms vary and are often nonspecific or absent, many women go undiagnosed, and prevalence is likely underestimated (1). Still, identification of these anomalies has improved. In 1988 the American Fertility Society produced a standard form for classification of Mullerian defects (2). Through the use of standardized nomenclature and improved radiologic investigations, physicians are better equipped to accurately identify pelvic anomalies and therefore, to manage their gynecologic and obstetric sequelae.

This young, nulliparous patient reported predominantly cyclic left-sided pelvic pain that initiated at menarche and abated with combined oral contraceptives. Her gynecologic history and imaging suggested a Mullerian anomaly. This clinical presentation was consistent with a

non-communicating/obstructed hemi-uteri containing hormonally responsive endometrium. Such patients typically become symptomatic at menarche and are diagnosed in their teens or early twenties. Pelvic pain and dysmenorrhea manifest due to obstruction and/or endometriosis. Such patients may also seek evaluation due to infertility or a history of recurrent obstetrical complications (3,4).

Traditionally MRI has been considered the best noninvasive means of diagnosing anomalies of the reproductive tract. MRI provides excellent delineation of both internal and external uterine contour and enables measurement of intercornual diameter, visualization the endometrial contour, and identification of a uterine horn, should one be present. MRI has been quoted as having an accuracy of up to 100% in correctly identifying Mullerian anomalies (5). Cases of complex gynecologic pathology can prove challenging, however. In this case, the patient's leiomyoma was notably atypical in signal characteristics. It displayed T1 hyperintensity and T2 intermediate signal intensity, rather than the T1 and T2 hypointensity classically described. These imaging characteristics were later explained by proteinaceous fluid encountered at surgery but are not typical MRI findings of leiomyoma uteri. This case demonstrates that in the setting of necrosis, even MRI can mislead.

Like MRI, 3D ultrasound allows assessment of both internal and external uterine contour. Addition of the coronal plane enables visualization of the cavity and fundus. 3D ultrasound further enables study of vascularization, calculation of cavity volume(s), and measurements of septum length and thickness. The superior diagnostic capabilities of 3D ultrasound in comparison to 2D ultrasound and hysterosalpingogram have been documented. In recent years, with the increasing availability of 3D ultrasound, several small studies have evaluated its sensitivity and specificity in evaluation of Mullerian anomalies. All studies demonstrated high accuracy of 3D ultrasound in the diagnosis of uterine malformations, reporting sensitivity and specificity as 98% and 100% respectively (6).

There remains, however, a dearth of studies comparing MRI with 3D ultrasound in the diagnosis and classification of uterine anomalies. One such study, performed by Bermejo et al in 2010, reported a high degree of concordance between 3D ultrasound and MRI, stating that images obtained from each modality were practically equivalent. The authors argued 3D ultrasound to be a valid alternative to MRI, considering its lower cost and improved patient tolerability. The study was limited by the fact that MRI was only carried out after a uterine anomaly was identified on 3D ultrasound. Thus, the ability of MRI to identify cases of uterine anomalies not identified on 3D ultrasound could not be assessed (7). In this case, the 3D ultrasound findings were consistent with MRI in suggesting a right uterine horn. Future studies comparing 3D ultrasound and MRI are needed to further determine whether 3D ultrasound represents an equally efficacious modality to MRI in the diagnosis of Mullerian anomalies.

Historically, hysterosalpingogram (HSG) was first line in evaluating abnormalities of the uterine cavity, particularly in the setting of infertility, recurrent miscarriage, or obstetric sequelae. Subsequently, 2D pelvic ultrasound predominated, and more recently this modality has largely been unseated by the modalities discussed above. Though specificity of 2D ultrasound for diagnosis of Mullerian anomalies was reported to be 100%, with no noted false positives, sensitivity may be as low as 44% (8). HSG remains an important tool in the evaluation of uterine and tubal anatomy, but its routine use is no longer recommended in evaluation of Mullerian anomalies, especially when MRI and 3D ultrasound are readily available. Because definitive diagnosis and classification of Mullerian anomalies requires evaluation of the external uterine contour, MRI, 3-D ultrasound, and laparoscopy have supplanted HSG in this domain (5, 9). It is noted, however, that here, preoperative HSG would likely have demonstrated fill of and spill from the bilateral fallopian tubes with

injection of radio-opaque contrast through the patient's single cervix, as chromoperturbation did intra-operatively.

This case raises the question of what should constitute the standard pre-operative protocol for a patient whose history, examination and imaging studies suggest a rudimentary horn. In this instance, a degenerating fibroid with atypical MRI signal characteristics mimicked this Mullerian anomaly. In 2011, Ludwin et al. compared diagnostic accuracy of sonohysterography, hysterosalpingography, and hysteroscopy, three modalities that were not used in this case. The diagnostic accuracy varied both by modality and by type of anomaly; however, the authors concluded that sonohysterography had the highest overall diagnostic accuracy (95.2%) and sensitivity (95.9%) of the three (10). In addition, the authors indicated that combining imaging modalities improved accuracy of detection. It should be noted, however, that a prior study by Mazouni et al. found that adding any of these modalities to MRI alone did not improve sensitivity in diagnosing Mullerian defects. Therefore, these authors deemed MRI the gold-standard non-invasive diagnostic test, short of laparoscopy (11).

It is difficult to anticipate unusual presentations of leiomyomata, and their imaging features may be highly variable, making interpretation challenging. When fibroid imaging characteristics are classic, they are easily identified. However, necrotic and degenerating fibroids have been reported to mimic other diagnoses such as hydrometra and pelvic malignancy (12). In these cases, as in ours, only diagnostic laparoscopy was definitively diagnostic of leiomyoma, with the imaging features otherwise suggesting more unusual pelvic pathology. It is the opinion of these authors that given MRI findings suggesting non-communicating horn with hematometra (a diagnosis less common than fibroids but by no means rare), further preoperative diagnostic testing would not have changed the recommendation for minimally invasive surgical treatment in this symptomatic patient.

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Figure 1.

(A) Axial T1-weighted axial image of the pelvis and (B) axial T2-weighted image with fat suppression suggest two separate uterine cavities (arrows) with thick intervening myometrium (arrowheads). The T1 hyperintense, T2 intermediate signal of the presumptive left cavity are notably atypical of leiomyoma and suggest necrosis and/or hemorrhage. (C) Sagittal T2-weighted image with fat suppression suggests continuation of right uterine cavity to cervix (arrow) (not shown: left “cavity” does not communicate with right cavity or cervix). (D) Coronal inversion recovery images suggest two separate cavities with only right cavity continuous with the endocervix (arrow).

Figure 2A

Figure 2B

Figure 2.

Figure 2A: Bulbous contour of left anterior corpus, containing involuting intramural leiomyoma.

Figure 2B: Restoration of normal uterine contour following laparoscopic resection of degrading myoma and repair of defect.