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Double the trouble: A case report on uterine didelphys with unilateral cervical atresia

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Abstract:

Genetic errors and teratogenic events during embryonic development can lead to congenital abnormalities of the female reproductive tract. Many patients are asymptomatic, while some have major abnormalities that can cause severe impairment of menstrual and reproductive functions. This case report focuses on Müllerian anomaly Class III, uterine didelphys, along with obstructed hemivagina and ipsilateral renal agenesis (the Herlyn–Werner–Wunderlich syndrome [HWWs]). The patient presented with cyclic pelvic pain due to the hematometra and hematocolpos. She initially underwent resection of the vaginal septum but had a recurrence of obstructive symptoms more than a year after vaginal surgery was performed. This prompted further evaluation with magnetic resonance imaging revealing cystic dilatation of the right uterine horn and a hypoplastic right vagina appearing to end blindly. The patient subsequently underwent hemihysterectomy. The right hemiuterus was noted to have a depression on its thickened inferior aspect, but no cervix was identified. This case is a variant of the classic HWWS, as there was cervical atresia on the right uterine corpus. A thorough preoperative evaluation and accurate intraoperative assessment of patients with Müllerian anomalies can decrease misdiagnoses, guide appropriate intervention, and decrease the risk of future reproductive complications.

Keywords:

Cervical atresia, Müllerian anomaly, uterine didelphys

Introduction

Genetic errors and teratogenic events during embryonic development can lead to congenital abnormalities of the female reproductive tract. These can affect the external genitalia or Müllerian structures. Many patients are asymptomatic, while others present symptoms at different times in a woman's life. Some major abnormalities may even cause severe impairment of menstrual and reproductive functions.[1]

This case report focuses on a particular Müllerian anomaly, uterine didelphys (Class III), characterized by a complete lack of fusion of the Müllerian ducts leading to the two separate uterine cavities and the two cervices.[2] This subclass has an estimated overall prevalence of 11% and among the general population, it is the second-least common congenital uterine malformation next to a unicornuate uterus.[3] Most reports include this anomaly along with a syndrome of obstructed hemivagina, ipsilateral renal anomaly (OHVIRA), otherwise known as the Herlyn–Werner–Wunderlich syndrome (HWWS). The largest institutional study available in literature documented 87 varied double uteri with OHVIRA. In that study, 72.4% presented as HWWS, a classic anatomic variant, but the remaining 27.6% were rare variants. Included in the latter subset is the 4.5% (four cases) who presented with unilateral cervical atresia, similar to the case presented in this paper.[4]

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Submitted: 02-Mar-2023
Revised: 20-Apr-2023
Accepted: 25-Apr-2023
Published: 30-May-2023

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How to cite this article: Loquero CG, Yiu-Senolos ME. Double the trouble: A case report on uterine didelphys with unilateral cervical atresia. Philipp J Obstet Gynecol 2023;47:37-41.

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The index patient presented with cyclic pelvic pain, a classic symptom of obstructive Müllerian anomalies causing hematometra and hematocolpos. Early series of cases reported excellent recovery of the obstructed uterus after vaginal septum resection with no recurrence of hematometra.[5] It is well documented that resection of an obstructing vaginal septum is the treatment of choice for such cases. This conservative approach provides drainage of retained blood in order to relieve symptoms as well as to preserve fertility. However, the patient, in this case, had a recurrence of hematometra more than a year after vaginal surgery was performed. Thus, one can surmise that a more thorough preoperative and intraoperative investigation may be needed when conservative management fails.

Case Report

An 18-year-old female was admitted for the second time to our institution due to hypogastric pain.

The patient had menarche at 13 years old with regular monthly cycles lasting for 5 days, consuming 2–3 moderately soaked pads per day associated with tolerable dysmenorrhea, which was relieved by rest and intake of mefenamic acid. The patient denies any history of sexual contact. She has no known comorbidities with no family history of congenital anomalies.

Three years before admission, the patient was admitted in another institution due to complaints of sharp hypogastric pain radiating to the right inguinal area. She was managed as a case of recurrent urinary tract infection (UTI). Ultrasound of the kidneys–ureters–bladder revealed an absence of the right kidney with compensatory enlargement of the left kidney. Magnetic resonance imaging (MRI) was advised but was not done due to financial constraints. Thereafter, the patient had no recurrence of UTI.

Two years before admission, the patient noted worsening dysmenorrhea. Pelvic ultrasound done showed uterine didelphys with considerations of right hematometra, dilated cervix, hematocolpos, and hematosalpinx versus dilated ureter. She was advised of surgery but sought admission to our institution, where a transrectal ultrasound done during the 1st day of her menses showed uterine didelphys with an intact 0.8 cm-thick endometrium on the left hemiuterus. The endometrium of the right hemiuterus was 2.25 cm in thickness and the cavity was dilated with heterogenous fluid, probably hematometra [Figure 1]. She was diagnosed to have obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) and underwent excision of transverse vaginal septum. She was discharged, improved, and was prescribed

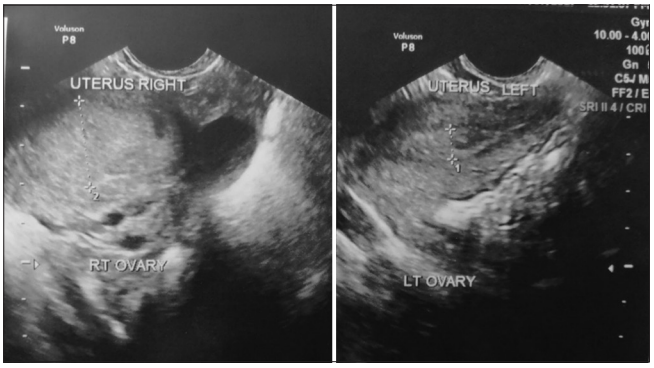

Figure 1: Right uterus with hematometra and hematocolpos (left image) Left uterus (right image)

cytoproterone acetate + ethinyl estradiol to stop her monthly bleeding. The patient had a complete resolution of pain after that.

The patient continued to take combined oral contraceptive (COC) pills and was amenorrheic. A transrectal ultrasound done 2 months after surgery revealed two normal-sized anteverted uteri with thick endometria and two closed cervices. The left cervix was distinctly connected to the vagina. The right cervix appeared short. Both ovaries were unremarkable.

Five days before admission, the patient discontinued taking the COC pills. She noted heavy bleeding, consuming four fully soaked pads per day. On the 3rd menstrual day, she had vaginal spotting associated with severe hypogastric pain radiating to the right inguinal area. She took tramadol + paracetamol 37.5/325 mg one tablet as needed every 8 h and mefenamic acid 500 mg one tablet as needed every 8 h, which offered temporary relief. The persistence of the pain prompted admission.

On admission, her vital signs were stable. Physical examination revealed the abdomen to be soft with direct tenderness on the right lower quadrant and hypogastric area. On rectal examination, a 4 cm × 3 cm tender, mobile mass was noted on the right adnexal region. The rest of the physical examination findings were unremarkable.

An obstructed hemivagina was considered and the patient was scheduled for an excision of the vaginal septum. On vaginal exploration under spinal anesthesia, the left cervix was seen with minimal bleeding from its os [Figure 2]. However, the right cervix was not identified. Further exploration was hindered by the protrusion of the urinary bladder in that area. The planned procedure was abandoned, and further imaging tests were done.

An MRI done revealed duplication of the uterine horns with cystic dilatation of the right and a hypoplastic right vagina that appears to end blindly. Retained hemorrhage

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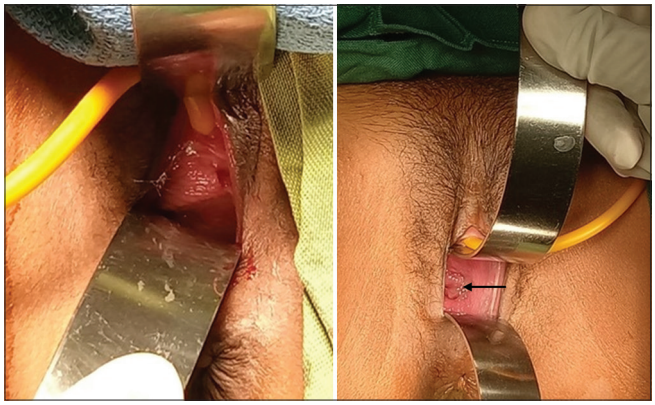

Figure 2: Vaginal exploration findings: Protruding bladder (left image) & left cervical os (arrow in right image) were visualized

was seen within the right uterine horn, while the left was not enlarged with an unremarkable endometrium. The internal and external os and a single vaginal canal were well-developed, which appeared to exit through a single introitus. There was no intravaginal septum formation noted. Müllerian duct anomaly type III (uterine didelphys) was considered.

The patient subsequently underwent a pelvic laparotomy. Two uteri were seen: The right hemiuterus measured 7 cm × 7 cm with endometriotic implants noted on its posterior wall, while the left measured 5 cm × 3 cm with a distinct connection to the vagina. The right fallopian tube was dilated to 4 cm × 2 cm, filled with blood clots. Both ovaries and the left fallopian appeared grossly normal. A right hemihysterectomy and ipsilateral salpingectomy were done. On sectioning of the right uterus, approximately 100 ml of clotted blood was noted with a depression seen on its thickened inferior aspect, but no cervix was identified [Figure 3].

The postoperative course was unremarkable and the patient was discharged improved. On the final histopathology report, the right hemiuterus showed basal endometrium and myometrium with no significant histopathology findings. Grossly, there was an abnormal-shaped cervix with no cervical os. Microscopically, basal endometrium with deep underlying adenomyosis was seen. The right fallopian tube showed serosal endometriosis. The final diagnoses were hematometra secondary to uterine didelphys with right cervical agenesis and pelvic endometriosis American Fertility Society Stage I.

Discussion

The embryology of the female genital tract is derived from the Müllerian ducts, which form the fallopian tubes, uterus, and upper vagina. Initially, these ducts extend downward from the level of the mesonephros (mesonephric kidney), and they turn

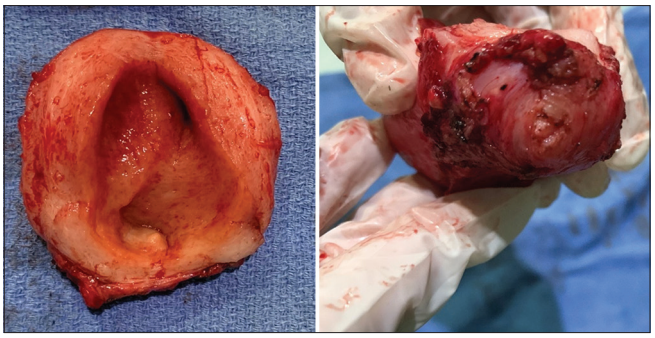

Figure 3: Longitudinal cut section of right uterine corpus post right hemihysterectomy (left image) obliterated lower uterine cavity (right image)

medially to meet and fuse together in the midline. When both Müllerian ducts develop but fail to fuse at approximately 8th–10th week of gestation, a duplicated system results. This is class III of the American Society of Reproductive Medicine (ASRM) classification of Müllerian anomalies, termed uterus didelphys. A more recent classification system is provided by the European Society of Human Reproduction and Embryology, which further provides subclasses of cervical and vaginal anomalies with clinical significance.[6] However, the classification of the ASRM is still the most commonly used classification for Müllerian duct anomalies.

The classic HWWS is a syndrome characterized by the triad of: (1) uterine didelphys; (2) obstructed hemivagina, where a partially formed vaginal septum results in one unobstructed and one obstructed vagina; and (3) ipsilateral renal agenesis, due to the close relationship between the development of urinary and reproductive systems. This syndrome is also commonly reported in literature as obstructed hemivagina and ipsilateral renal anomaly (OHVIRA), which includes two of the three defects of classic HWWS. Aside from renal agenesis, other malformations, including multicystic dysplastic kidney and renal duplication, have also been reported.[7]

Patients affected by HWWS have no specific symptoms until puberty or a few years after the menarche, as was experienced by the index patient. They typically present with acute pelvic pain, dysmenorrhea, presence of pelvic mass, recurrent UTIs, and urinary retention.[8] Due to obstruction, these young patients usually suffer from similar and sometimes severe symptoms. There is a higher risk of endometriosis, hematosalpinx, and pelvic inflammatory disease, which, together with obstruction, could jeopardize the future fertility of the patients.[9]

Anatomic congenital cervical malformations can be classified as agenesis (no cervix) or dysgenesis. The latter form has three distinct variations: (1) cervical obstruction – A well-formed cervix exists, but a portion of the endocervical lumen is obliterated; (2) cervical fibrous

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cord – A cervical cord of variable length and diameter is noted with a completely obliterated endocervical canal; and (3) cervical fragmentation – Portions of the cervix are noted with no connection to the uterine body and sometimes palpable below the fundus. Absence or fragmentation of the uterine cervix was the most frequently observed malformation.[10] In 30 women examined and treated for cervical malformation, 19 of them underwent primary hysterectomy, while 11 underwent a more conservative procedure with the aim of conserving reproductive capacity. This conservative procedure included uterovaginal anastomosis for patients with cervical agenesis and cervicovaginal reconstruction, including the creation of a neocervical canal for the different forms of cervical dysgenesis. In those where cervical reconstruction was completed, 6 of the 11 cases ultimately underwent hysterectomy after obstruction of the neocervical canal. One patient with cervical obstruction, on the other hand, had a successful pregnancy and subsequently delivered.[10]

Previous case reports suggest that treatment for OHVIRA or HWWS entails simple resection of the vaginal septum. However, some variants of this anomaly, such as the presence of cervical atresia seen in the case presented, may warrant a more extensive approach in the form of a hemihysterectomy. Thus, it is prudent to accurately assess anatomical structures preoperatively and intraoperatively in order to provide definitive and adequate treatment.

Ultrasonography is a logical initial imaging tool because of its low cost and ability to provide real-time imaging in a less invasive way. Traditional two-dimensional ultrasonography has a sensitivity of around 44% and specificity of 85%–92% in evaluating Müllerian anomalies. However, with the advent of three-dimensional (3D) ultrasonography, higher sensitivity of 93% and specificity of 100% may be achieved.[11] However, MRI is considered the gold standard as it can provide clear delineation of both the internal and external uterine anatomy, including cervical and vaginal morphology. It has a reported accuracy of up to 100% for Müllerian anomaly evaluation.[12] Images obtained with 3D ultrasound have been shown to be equivalent to that of MRI and both have similar diagnostic accuracy. Few differences were observed when only the lower part of the uterus was studied. Thus, it was proposed that MRI should be performed particularly for the assessment of the cervix and vagina.[12] In a case series of pediatric patients with congenital reproductive anomalies, MRI diagnosis of obstruction sites correlated completely with those seen during surgery. On the other hand, ultrasound findings were consistent in the diagnosis of typical HWWS but inconsistent or indeterminate in cases complicated by cervicovaginal agenesis.[9] In the case presented, MRI was done and the diagnosis of OHVIRA/HWWS was made. At that time, the cervical atresia was not identified. However, hypoplastic vagina was noted, seen as a fibrous band-like structure inferiorly from the right hemiuterus. This may signify the overlooked cervical dysgenesis in the patient. When a hematometrocolpos with minimal upper vagina and a hematometra with cervical atresia is difficult to distinguish definitively, vaginoscopy with diagnostic laparoscopy can further help with anatomic mapping.[13]

Treatment options target at avoiding complications and restoring the functionality of the genital system to help achieve fertility potential. This can be accomplished by early surgical intervention with resection of the vaginal septum and hematometra drainage in classic HWWS. However, variations associated with cervicovaginal atresia are often difficult to treat, and surgical interventions such as utero-vaginal canalization are recently gaining momentum. This procedure has a low risk of postoperative complications and offers normal menstrual and fertility outcomes. However, fertility may be affected if a patient had endometriosis before presentation, postoperative adhesions, restenosis of the new cervix, or changes in the cervical microenvironment.[14] As the diagnosis of cervical atresia is often made for the first time intraoperatively, attempts at fistula formation and simple septum resection have been described with the accumulation of menstrual egress, eventually leading to hysterectomy. Hysterectomy is the most commonly reported surgical outcome in literature with this condition. Being unaware of the presence of cervical atresia preoperatively precludes effective counseling of the patient and her family and delays the formulation of the most appropriate surgical plan, possibly leading to additional procedures.[15] A study reports two cases of uterine didelphys with cervicovaginal agenesis who subsequently underwent hemihysterectomy. Intraoperatively, the unilateral cervicovaginal agenesis was noted with no connection between the obstructed hemiuterus and the vaginal vault. This necessitated the removal of the obstructed hemiuterus in both patients.[13] In our case, the patient initially underwent excision of the vaginal septum. However, the persistence of obstructive symptoms prompted further evaluation and the patient subsequently underwent hemihysterectomy.

Women affected by Müllerian duct anomalies are at increased risk of obstetric complications such as recurrent miscarriage, abnormal fetal presentation, postpartum hemorrhage, retained placenta, fetal mortality, fetal growth restriction, and premature rupture of membranes.[8] HWWS has a good obstetric prognosis: Pregnancy rate of 87%, with approximately

62% having positive obstetric outcomes without complications during delivery. In a case series of 42 patients with obstructed hemivagina who underwent resection of the vaginal septum, nine had 20 pregnancies, with live births of 69%.[16] Reproductive performance and obstetric complications of 49 patients affected by didelphys uterus were also reported. In this study, the incidence of primary infertility was not significantly increased in these women. The rate of spontaneous miscarriage was 21%, with preterm birth in 24%, and cesarean section rate at 84%, which is reflective of the high incidence of a breech presentation at 51%.[17] In another study, one successful pregnancy was carried to term in four women with a double uterus who subsequently underwent hemihysterectomy. In the same subset of the patients, one pregnancy resulted in spontaneous abortion.[18]

Summary

We present a case of a variant of the classic HWWS syndrome with cervical atresia on the right hemiuterus. A thorough preoperative evaluation and accurate intraoperative assessment of patients with Müllerian anomalies can decrease misdiagnoses, guide appropriate intervention, and decrease the risk of future reproductive complications.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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