Young or old, this condition may unfold: The use of ultrasound in the diagnosis of a wide spectrum of Herlyn Werner Wunderlich Syndrome, a case series*

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ABSTRACT

Herlyn-Werner-Wunderlich syndrome, characterized by uterus didelphys with blind hemivagina and ipsilateral renal agenesis, is a rare Mullerian duct anomaly. This case series shows a wide spectrum of the condition, one presenting in an adolescent, managed conservatively and the other in the perimenopausal age group given a more definitive management. The first case is an 18-year-old nulligravid who manifested with progressive dysmenorrhea and foul smelling vaginal discharge a few years after menarche. She subsequently underwent vaginal septotomy followed by diagnostic hysteroscopy. On the other hand, the second case is a 46-year-old nulligravid whose chief complaint is a foul smelling vaginal discharge and consequently went through a total abdominal hysterectomy with salpingo-oophorectomy. To our knowledge, the second case is the only patient diagnosed in the perimenopausal stage and underwent a total hysterectomy. Ultrasound is the first-line imaging modality used in both cases and its merits are highlighted to prove its importance and diagnostic value in the workup of this condition.

Keywords: Uterine didelphys, obstructed hemivagina, renal agenesis, Herlyn-Werner-Wunderlich Syndrome, Mullerian duct anomaly, 2D Ultrasound, 3D Ultrasound

INTRODUCTION

Herlyn-Werner-Wunderlich syndrome, characterized by uterus didelphys with blind hemivagina and ipsilateral renal agenesis, is a rare Mullerian duct anomaly.[1] Its estimated occurrence is 0.1%–3.8%.[2] Though its incidence in the Philippines is not yet established, a few local reports are available for review. At East Avenue Medical Center, a tertiary government institution, 4 cases were reported in the last 5 years[3]. There were 7 cases published in local literature in 2015 by Sucayan-Sta. Ana and Gorgonio[4]. Morante and Alensuela[5] also in 2015 reported 2 cases. The first case in this report, an adolescent, fits the typical presentation, course of work-up and management as in the cases reviewed in the available local reports. However, the second case, a perimenopausal woman, is uncommon as to the age of diagnosis, and subsequent management. Ultrasound is the first-line imaging modality used in all of the cases reviewed and has proven its importance and diagnostic value in the workup of this condition.

We aim to present a series of 2 cases of this rare syndrome, within a span of 3 years at our tertiary private hospital. The objectives of this report are the following: 1) To compare the clinical signs and symptoms presented by the 2 patients belonging to different age groups, adolescent and perimenopausal period, 2) Discuss the imaging work up of this rare syndrome and highlight the merits of ultrasound in making the diagnosis, 3) Discuss the different management that can be done in patients with this condition. The 2 cases had different presentations as well as management revealing a gamut of how the syndrome may appear and be treated. The patient profile, history and physical examination, imaging and the management of both cases are summarized in Table1.

CASE SERIES

CASE 1

An 18-year-old nulligravid, had new onset dysmenorrhea three years after menarche (at the age of 13) during day 1 of menses. There was progression of dysmenorrhea 3 months prior to admission, which was accompanied by foul smelling brownish vaginal discharge prompting consult with an OB-GYN in Oriental Mindoro. Physical examination showed a bulging mass on the right vaginal wall and direct tenderness on the right adnexal region. A whole abdominal ultrasound was requested which showed a non-visualized right kidney and a suspicious hypoechoic ovoid structure in the cervical area. A transvaginal ultrasound revealed a bicornuate uterus

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Table 1. Summary of the patient profile, history and physical examination, imaging and management of both cases

Case No.	1	2
Age at Diagnosis	18	46
Clinical Presentation	Progressive dysmenorrhea and Foul smelling vaginal discharge	Foul smelling vaginal discharge
Pertinent Physical Examination	4 x 3 cm cystic, tender, bulging mass on the right vaginal wall, 3 cm from introitus; cervix firm, short, closed, uterus normal-sized, movable, nontender; left adnexa no mass or tenderness; right adnexa not fully assessed due to tenderness on palpation	3 x 3 cm smooth bulging mass noted on the right side of cervix; uterus normal in size, anteverted, slightly movable, non-tender, (+) bilateral adnexal tenderness but adnexa could not be fully evaluated due to voluntary guarding
Imaging modality TVS	(2D) Consider uterine didelphys with blind hemivaginal pouch on the right, proliferative endometrium, normal-sized ovaries (3D) confirmed the presence of 2 uteri and 2 cervices – Uterine Didelphys; blind hemi-vaginal pouch on the right	(2D) Globular, anteverted uterus, questionable bicornuate, secretory endometrium, right adnexal mass probably cystic right ovary, left ovary not seen, consider cervical mass - fluid collection in the right pelvic cavity (3D) confirmed the presence of 2 separate uteri and 2 cervices - uterine didelphys
CT scan/MRI	(MRI) absent right kidney and ureter, uterine didelphys (both uteri have separate cervices and vaginal canals sharing a common midline septum), hematocolpos, right	(CT scan) uterine didelphys, probably obstructed right hemivagina and absent ipsilateral kidney
Management	Vaginoscopy, Diagnostic hysteroscopy followed by vaginal septotomy and drainage of hematocolpos	Pelvic laparotomy; Total abdominal hysterectomy with right salpingo-oophorectomy and left salpingectomy

and a probable cervical mass. The assessment at that time was Mullerian duct anomaly hence she was referred to a tertiary hospital for further evaluation.

Upon consult at our institution, physical examination showed a flat abdomen with direct tenderness on the right lower quadrant and no mass was palpated. On inspection of external genitalia, Tanner stage 4 pubic hair distribution was noted and there were no gross lesions. On speculum examination, there was a 4 x 3 cm cystic bulging mass over the right vaginal wall, 3 cm from the introitus; cervix was pink, smooth with minimal whitish, mucoid, non-foul smelling discharge. On internal examination, about 3 cm from the introitus was a 4 x 3 cm cystic, tender, bulging mass on the right vaginal wall; the cervix was firm, short, closed, uterus was normal-sized, retroverted, movable, nontender; left adnexa had no mass or tenderness; right adnexa cannot be fully assessed due to tenderness on palpation. Transvaginal 2D ultrasound on transverse view revealed two separate corpuses: left hemiuterus measures 1.9 x 2.13 x 2.43 cm, retroverted homogenous, with an endometrial lining of 0.56 cm and cervix of which measures 1.57 x 2.21, 2. 1.28 x 1.55 cm while the right hemiuterus measures 2.44 x 1.81 x 2.95 cm, anteverted, homogenous, with an endometrial lining of 0.84 cm and cervix of which ends into a cystic mass with low level echoes, 3.29 x 2.49 x 3.09 cm appearing in the vagina. Ultrasound impression at that time was consider uterine didelphys with blind hemi-vaginal pouch on the right, proliferative endometrium and normal-sized ovaries (Figure 1). Three-dimensional ultrasound confirmed the presence of 2 separate uteri and 2 cervices (Figure 2A) with the right cervix ending into a cystic mass with low level echoes (Figure 2B). MRI showed absent right kidney and ureter, uterine didelphys with 2 separate uteri with widely divergent fundi and empty uterine canals. Both uteri have separate cervices and vaginal canals sharing a common midline septum. The right vaginal canal is dilated and appears to end blindly with intraluminal T1W intermediately dark and T2W intermediately bright signals suggesting possible hematocolpos. The left vaginal canal is slightly

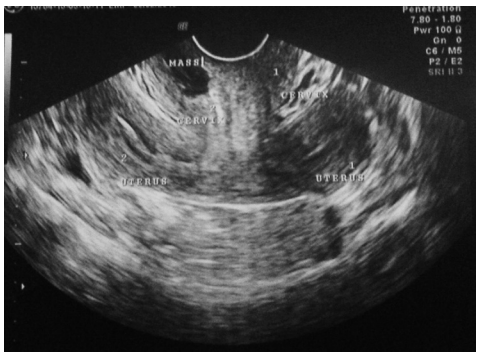

Figure 1. *Case1: Transvaginal 2D Ultrasound* (transverse view) - Uterine didelphys, Two separate corpuses: Left hemiuterus measures 1.9 x 2.13 x 2.43 cm, retroverted homogenous, with an endometrial lining of 0.56 cm and cervix 1.57 x 2.21, 2. 1.28 x 1.55 cm right hemiuterus measures 2.44 x 1.81 x 2.95 cm, anteverted, homogenous, with an endometrial lining of 0.84 cm and cervix of which ends into a cystic mass with low level echoes, 3.29 x 2.49x 3.09 cm appearing in the vagina.

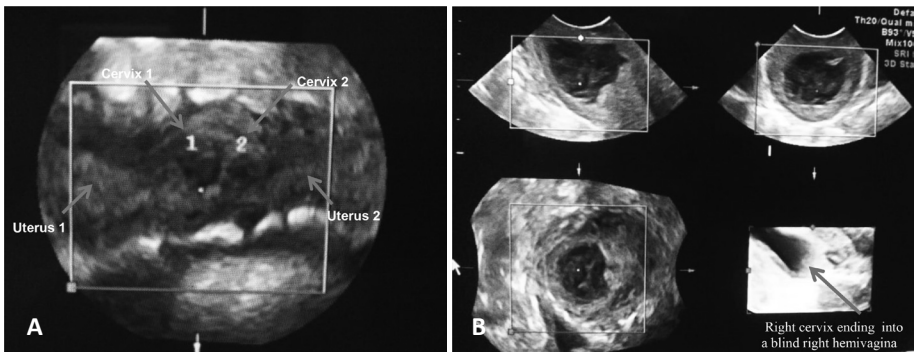

Figure 2. *Case1 3D Three-dimensional ultrasound* **(A)** confirmed the presence of 2 separate uteri and 2 cervices **(B)** with the right cervix ending into a cystic mass with low level echoes.S

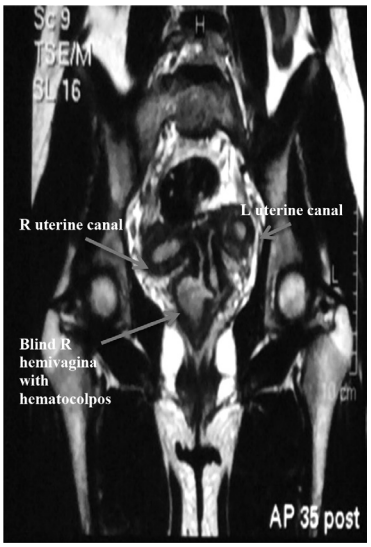

Figure 3. *Case 1 MRI of the abdomen*. Absent right kidney and ureter, uterine didelphys with 2 separate uteri with widely divergent fundi and empty uterine canals. Both uteri have separate cervices and vaginal canals sharing a common midline septum. The right vaginal canal is dilated and appears to end blindly with intraluminal T1W intermediately dark and T2W intermediately bright signals suggesting possible hematocolpos. The left vaginal canal is slightly compressed by the dilated right side and courses towards the perineal region.

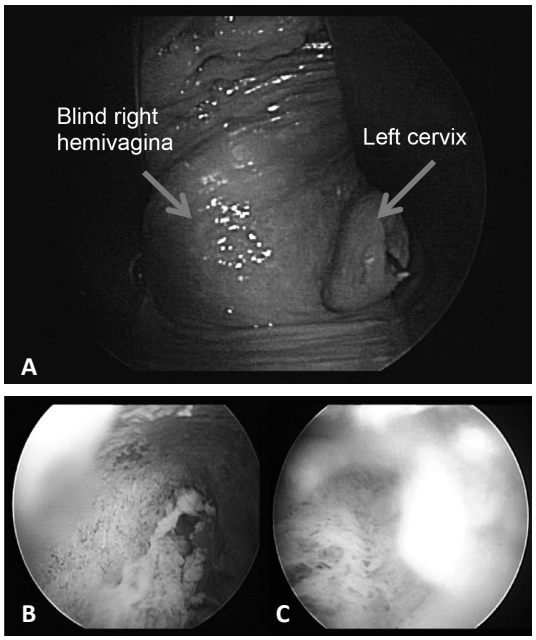

Figure 4. Case 1 (A) Diagnostic vaginoscopy showing a blind right hemivagina and left cervix. **(B)** Diagnostic hysteroscopy of the left hemiuterus, the fundus and a single ostium were visualized. The endometrial lining was thin **(C)** Diagnostic hysteroscopy of the right hemiuterus, where the fundus and a single ostium were visualized.

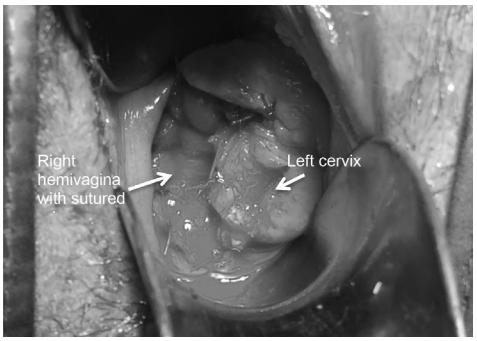

Figure 5. Left cervix and right hemivagina after septotomy showing the sutured edges of the newly-formed opening

compressed by the dilated right side and courses towards the perineal region. Both ovaries appear normal (Figure 3). Diagnosis of Herlyn-Werner-Wunderlich Syndrome was made based on clinical presentation and imaging studies. Patient was subsequently advised admission for vaginal septotomy with drainage of hematocolpos and diagnostic hysteroscopy.

Upon diagnostic vaginoscopy, a blind right hemivagina and the left cervix were appreciated (Figure 4A). Upon diagnostic hysteroscopy on the left hemiuterus, the fundus and a single ostium were visualized (Figure 4B). The endometrial lining was thin. Proceeded with fine needle aspiration of the right bulging vaginal mass obtaining brownish fluid. This was followed by vaginal septotomy with drainage of 200 cc foul smelling chocolate brown fluid admixed with purulent material. Proceeded with diagnostic hysteroscopy of the right hemiuterus, where the fundus and a single ostium were visualized (Figure 4C). The endometrial lining was thin. There was no connection seen between the two uterine cavities. After septotomy, the newly formed opening of the right hemivagina had sutured edges (Figure 5) and post-operatively, a vaginal mold (a plastic syringe covered by condom) was placed for at least one week.

Menses resumed as expected with no accompanying dysmenorrhea.

CASE 2

A 46-year-old, nulligravid, came in due to a 5 year history of recurrent yellow-greenish foul-smelling vaginal discharge. She consulted two months PTA, when aside from the discharge, she had hypogastric pain graded 7/10, non-radiating. She was seen by an internist at a tertiary hospital in Parañaque. Complete blood count showed leukocytosis with neutrophilic predominance (WBC 21.4, Segmenters 0.79). She was advised to undergo CT scan of the whole abdomen to determine the cause of the abdominal pain, however the patient refused and decided to seek second opinion at our institution. Upon assessment by the gynecology service, the abdomen was noted to be soft with tenderness on both lower quadrants. On speculum exam, cervix was pink smooth and deviated to the left with yellow-greenish foul discharge, with a pinkish smooth bulging mass on the right of the cervix. On internal examination, the cervix was firm, short and closed, with a 3 x 3 cm smooth bulging mass noted on the right side of cervix; uterus was normal in size, anteverted, slightly movable, non-tender, there was bilateral adnexal tenderness however adnexa could not be fully evaluated due to voluntary guarding. On rectovaginal exam, there was a bulging cystic non-tender mass at the right anterior rectal wall. Assessment at that time was Pelvic Inflammatory Disease. She was admitted and was started on Cefoxitin 2g/IV every 6 hours, Metronidazole 500 mg/ tablet 1 tablet every 12 hours and Doxycycline 100 mg/ tab every 12 hours. At that time, the patient disclosed that she underwent left oophorectomy for endometrial cyst in 2008, however, she was not told of any uterine anomaly when intraoperative findings were discussed with her. No records were available for reference. A transvaginal 2D ultrasound was done to further evaluate both adnexa which showed a globular uterus, questionable bicornuate (5.59 x 8.57 x 5.03 cm); secretory endometrium; right adnexal mass probably cystic right ovary measuring 4.03 x 3.51 x 3.81 cm; left ovary not seen, consider cervical mass - a fluid collection in the right pelvic cavity (5.89 x 5.38 x 5.24 cm) (Figure 6). At that time, an endophytic cervical mass was considered hence a CT scan of the whole abdomen was done. CT scan showed absent right kidney and ureter with slightly enlarged left kidney; consider uterine didelphys, with a fluid-filled structure inferiorly showing connection with the right uterine cavity which may represent an obstructed and/or incompletely formed hemivagina causing hemato- and/or hydrocolpos; round hypodense structure with thin enhancing wall at right adnexal region may be ovarian in origin with associated hydrosalpinx; findings of uterine didelphys, probably obstructed right hemivagina and absent ipsilateral kidney (renal agenesis) seen in Herlyn-Werner-Wunderlich syndrome (Figure 7). A transvaginal 3D ultrasound was performed to diagnose the mullerian duct anomaly present in the patient and confirmed the presence of 2 separate uteri and 2 cervices making the diagnosis of uterine didelphys (Figure 8). Pertinent also in the history was that her menarche was at 12 years old, with regular intervals, lasting 6 to 7 days amounting to 5 moderately soaked pads per day with dysmenorrhea on days 2 to 3 of her cycle. Patient took Mefenamic Acid as needed which afforded relief of the dysmenorrhea. She had 1 same-sex partner with no dyspareunia.

After completion of antibiotics, patient was re-admitted with a diagnosis of Herlyn-Werner-Wunderlich Syndrome, ovarian new growth, right, probably benign. She underwent pelvic laparotomy and upon opening of the abdomen, there were dense adhesions between the omentum and the anterior abdominal wall. There was a 6 x 6 cm multiloculated mass containing clear serous fluid, encysting the right ovary and fallopian tube. The right ovary was cystically enlarged to 5 x 4 cm and the right fallopian tube was bulbuous measuring 8 x 1.5 x 1.5 cm (Figure 9a). This encapsulated mass had flimsy adhesions on the right lateral wall. The uterus was heart-shaped measuring 6 x 10 x 3.5 cm on its entirety (Figure 9b). It was densely adherent to the rectosigmoid colon obliterating the cul de sac. The right kidney and ureter were absent. The left ovary was surgically absent and the left fallopian tube was unremarkable. Parts of the omentum and pelvic sidewalls were densely adherent to the anterior and lateral surface of the uterus. Total abdominal hysterectomy with right salpingooophorectomy and left salpingectomy was done and upon circumferential incision at the cervicovaginal junction, there were 2 vaginal lumen noted (Figure 9c). The right vaginal lumen exuded yellow-greenish non-foul fluid and was noted to have a blind end. The left vaginal lumen was patent. On cut section of the uterus, there were two separate uterine horns and two cervices. The right uterine horn measured 5 x 5 x 3.5 cm and the left uterine horn measured 5 x 6 x 3.5 cm. The two horns were separated by a septum with 0.5 cm thickness. The endometrial lining of both were smooth. The right cervix measured 3 x 2x 2 cm and left cervix measured 4 x 2 x 2 cm. Both endocervical linings were smooth (Figure 10). On cut section of the right ovarian cyst, it was unilocular and the cyst wall was smooth with no nodularities and papillarities. The specimen was sent for histopathology which revealed uterus didelphys with proliferative endometrium, bilateral cervix with chronic inflammation, nabothian cyst and squamous metaplasia, cystic follicle in the right ovary, chorinic salpingitis right and unremarkable left fallopian

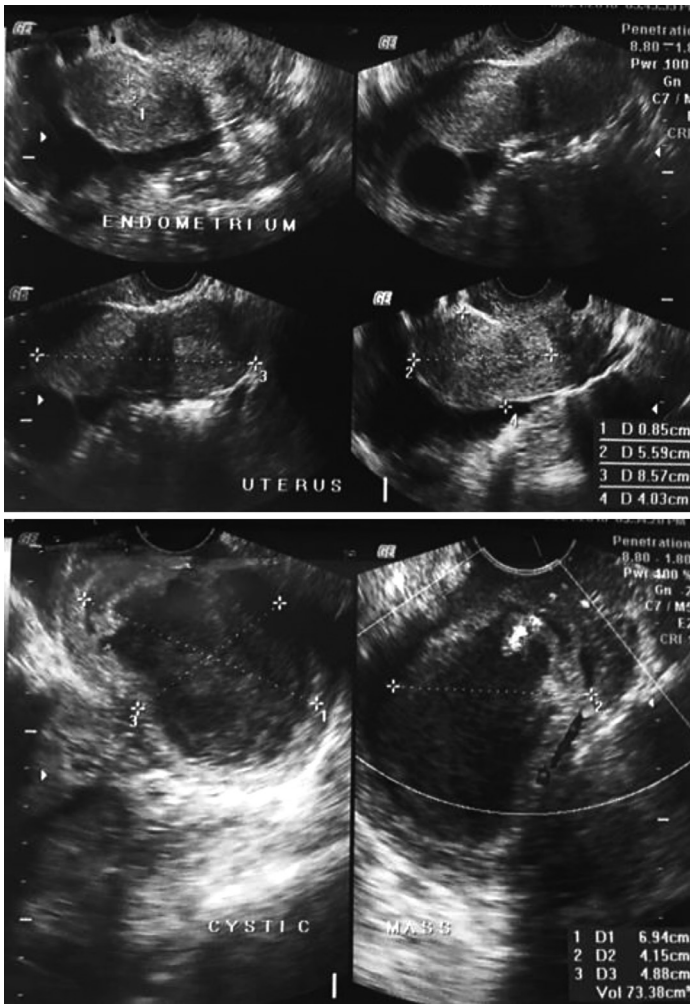

Figure 6. *Case 2 Transvaginal 2D ultrasound.* Globular uterus, questionable bicornuate, secretory endometrium; right adnexal mass probably cystic right ovary, left ovary not seen, consider cervical mass - a fluid collection in the right pelvic cavity

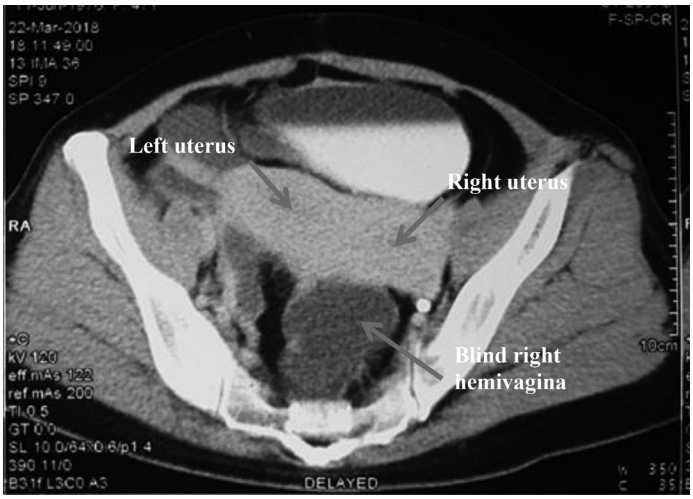

Figure 7. *Case 2 CT scan of the abdomen*. Showed 2 uterine horns with a fluid-filled structure inferiorly showing connection with the right uterine cavity which may represent a blind right hemivagina with hemato/hydrocolpos

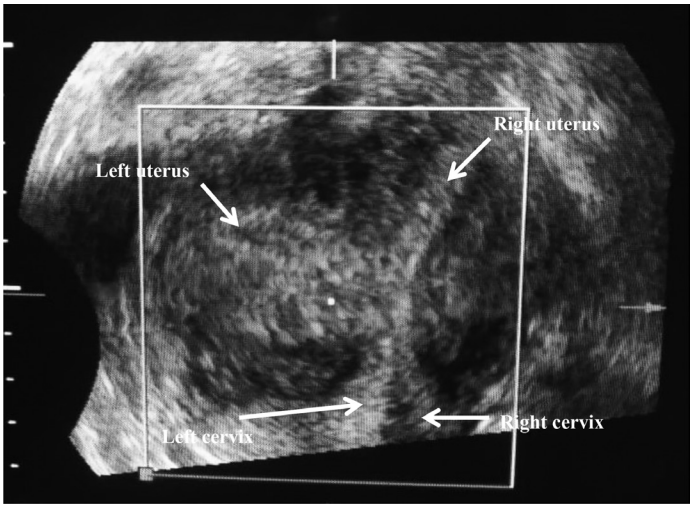

Figure 8. *Case 2 Transvaginal 3D ultrasound* confirmed the presence of 2 separate uteri and 2 cervices making the diagnosis of uterine didelphys

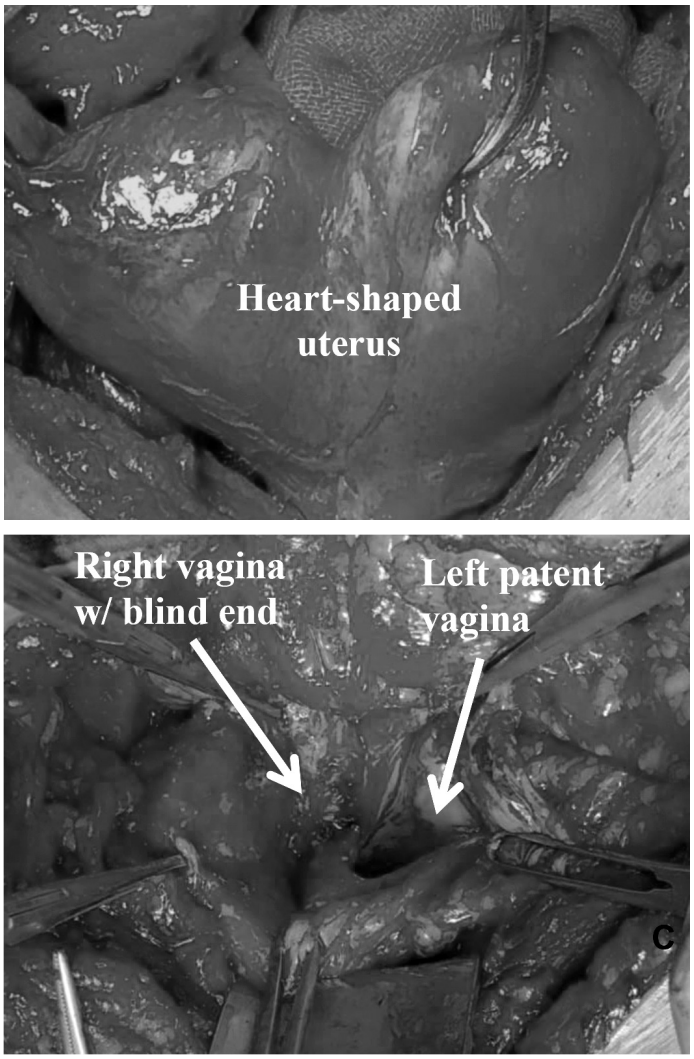

Figure 9. *Case 2 Intraoperative findings*. **(A)** The uterus was heart-shaped measuring 6 x 10 x 3.5 cm on its entirety with dense adhesions to the rectosigmoid colon obliterating the cul de sac. **(B)** Upon circumferential incision at the cervicovaginal junction, there were 2 vaginal lumen noted.

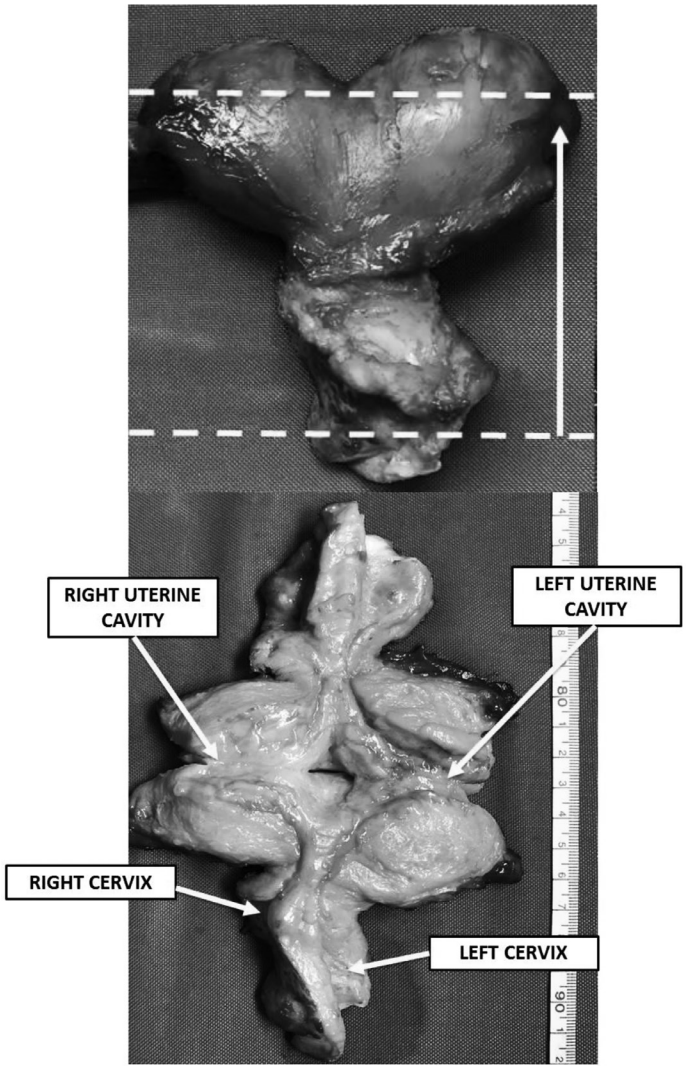

Figure 10. Case 2 Cut-section of specimen. On cut section of the uterus, there were two separate uterine horns and two cervices. The right uterine horn measured 5 x 5 x 3.5 cm and the left uterine horn measured 5 x 6 x 3.5 cm. The two horns were separated by a septum with 0.5 cm thickness. The endometrial lining of both were smooth. The right cervix measured 3 x 2x 2 cm and left cervix measured 4 x 2 x 2 cm. Both endocervical linings were smooth.

tube. Final diagnosis was Herlyn-Werner-Wunderlich Syndrome, cystic follicle, right, pelvic endometriosis stage IV, s/p left oophorectomy for endometrial cyst (2008).

CASE DISCUSSION

Herlyn-Werner-Wunderlich Syndrome (HWWS) is an uncommon congenital disorder characterized by Mullerian and mesonephric duct anomalies with an obstructed hemivagina. The female reproductive tract develops from a pair of Mullerian ducts that form the fallopian tube, uterus, cervix, and the upper two-thirds of the vagina. Normal development of the Mullerian ducts depends on the completion of three phases: organogenesis, fusion, and septal resorption.[6] The simultaneous insult to both Mullerian and mesonephric ducts at 6 to 9 weeks of fetal life gives rise to HWWS. However, the exact etiology of the insult and pathogenesis of the syndrome is still not clear.[7] The mesonephric ducts give rise to kidneys and also act as inductor elements for adequate Mullerian duct fusion. On the side where the caudal portion of the mesonephric duct is defective, the Mullerian duct is displaced laterally so it cannot fuse with the contralateral duct, resulting in didelphic uterus, and cannot come into contact with the urogenital sinus centrally. The contralateral Mullerian duct gives rise to a vagina while the displaced component forms a blind sac— an imperforate or obstructed hemivagina[8]. However, the distal part of the vagina is not affected because it originates from the urogenital sinus. According to the American Society for Reproductive Medicine (ASRM), HWWS represents a class III uterine anomaly and a vaginal class IIa anomaly. This applies to both of our cases[9].

The estimated overall prevalence of Mullerian duct anomalies (MDA) is 2–3% of women[10]. Uterus didelphys constitutes approximately 11% of MDAs while hypoplasia or agenesis of the uterus and proximal vagina represent 5–10%. Renal anomalies in association with MDAs are present in up to 43% of patients[10]. Approximately 75% of patients with didelphys uterus have a complete or partial vaginal septum[11], which is most commonly longitudinal in HWWS. The estimated occurrence of HWWS is 0.1%–3.8%. [2]

The presentation of HWWS is varied as evident in our case series. The primary and most common presenting symptom (90%) is progressive pelvic pain[12], usually diagnosed 2 months to a year after menarche.[13] The first case experienced her symptoms of dysmenorrhea accompanied by dyspareunia, 5 years after menarche while the second case had dysmenorrhea but was not bothersome enough to seek consult. The two patients in the case series may have incomplete obstruction, causing the delay in the diagnosis, as there is one hemivagina, which is patent, allowing for menstrual blood to exit, while the other side, being obstructed; eventually leads to a large hematocolpos[14]. Delays in diagnosis may also be due to a number of factors, such as in patients presenting with cyclical dysmenorrhea, the patient's primary care provider often prescribes non-steroidal anti-inflammatory drugs and oral contraceptives, which suppresses or eliminates menses and further delays recognition of the condition[14] such as in our second case. To our knowledge, the second case has the most advanced age at diagnosis, being in the perimenopausal age group. The second case's primary symptom is foul smelling vaginal discharge. Tong et al.,[15] reviewed the clinical characteristics of 70 patients with HWWS and found that 20% presented with foul smelling

vaginal discharge and pelvic inflammation. Retention of menstrual blood in the obstructed hemivagina leads to the formation of a hematocolpos as seen in both of our patients. Physical examination of both our patients revealed a bulging mass in the right side of the pelvic cavity which represents hematocolpos which is usually detected as pelvic mass in 40% of patients.[16]

Lan Zhu et al[2] in 2015 suggested a new classification as follows: Classification 1, a completely obstructed hemivagina (1.1 with blind hemivagina or 1.2 cervicovaginal atresia without communicating uteri) and Classification 2, an incompletely obstructed hemivagina (2.1 partial reabsorption of the vaginal septum or 2.2 with communicating uteri) (Figure 11). This new classification is formed because the clinical characteristics differed significantly between the completely and incompletely obstructed vaginal septum. Using this classification, the 2 patients in this case series may belong to classification 2.1, an incompletely obstructed hemivagina with partial reabsorption of the vaginal septum which may account for the later age of onset. Purulent or bloody vaginal discharge can also be the chief compliant such as in both of our cases since they are susceptible to ascending genital infection. However, the presence of the small communication between the 2 vaginas in both of our cases are not established.

Ultrasound is a very useful modality for diagnosing and classifying MDAs.[17] It is the initial imaging used to evaluate the uterus and other pelvic structures because it is the least invasive and most cost effective. In both of the cases presented in this case series, ultrasound was the initial diagnostic imaging tool used and triggered further workups to confirm the diagnosis and detect concomitant anomalies. However, two-dimensional ultrasonography has low sensitivity of 44% in evaluating MDAs[18]. In the advent of 3D/4D transvaginal ultrasound technology, through volume acquisition from any 2D images and displayed in any multiplanar views – coronal being the best and important, it can reproduce the clearer image of the triangular shape of the endometrial cavity and the fundal contour of the serosal surface thereby improving its diagnostic value. 3D ultrasound's sensitivity is 100%-98% and specificity is 100% in correctly categorizing mullerian duct anomalies.[19] This is considered valuable in discerning the different types of MDAs. Assessing the effectiveness of 3D ultrasound against laparoscopy and hysteroscopy for the diagnosis of uterine congenital defects, Mohamed etal.[20] reported a sensitivity of 97% and a specificity of 96%. Ghi etal.[21] reported a sensitivity and a specificity of 100% for the detection of uterine malformations and 96% concordance between ultrasound and endoscopy with respect to the type of anomaly diagnosed. Bermejo etal[22] compared magnetic resonance imaging with 3D-US

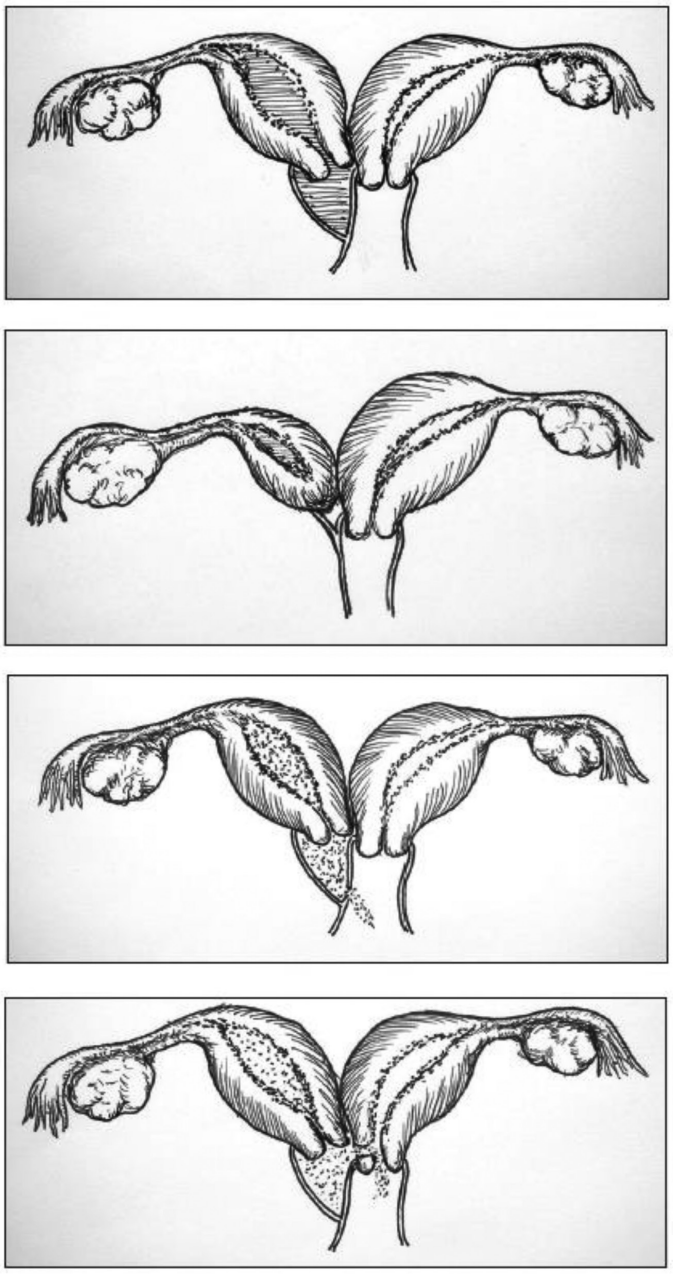

Figure 11. On cut section of the uterus is an empty endometrial cavity with decidualized endometrium (EM). A gestational sac with fetus (F) is seen occupying the anterior lower uterine segment pushing down to the proximal half of the cervix (CX).

in 286 women with Mullerian anomalies and concluded that if a bimanual pelvic examination and careful cervical imaging is performed, the two imaging techniques are similar, with a high degree of concordance. Overall, when performed by trained operators, 3D-US is a less invasive and cheaper method for the diagnosis of simple uterine malformations than laparoscopy or MRI, respectively. In both of our patients, the use of 3D ultrasound aided in the better diagnosis of the Mullerian duct anomaly present.

Although CT scan, as used in the second case, may offer visualization of uterine morphology as well as

adjacent pelvo-abdominal structures to make the diagnosis of HWWS, magnetic resonance imaging (MRI) is still considered the gold-standard imaging technique as it has a 100% sensitivity and 96 to 100% specificity.[18] It provides more detailed information regarding uterine morphology like uterine contour, the shape of the intrauterine cavity, the character of the septum, the continuity with each vaginal (obstructed and non-obstructed) lumen as well as associated aspects such as endometriosis, pelvic inflammation and adhesions[23].

Resection of the vaginal septum is the treatment of choice for obstructed hemivagina and resultant hematocolpos.[10] Vaginal septotomy is accomplished most commonly through a hysteroscopic approach.[24] The first case underwent vaginal septotomy followed by hysteroscopy. Since she had delayed presentation of cyclic pelvic pain and there was no hematometra seen on the side of the obstructed hemivagina, this suggests a possible connection between the two uterine cavities by which blood may have been draining to the unobstructed side. Hence diagnostic hysteroscopy was performed to establish presence of a connection, however, no connection was seen. Laparoscopy adds the theoretical benefit of further delineating exact uterine and pelvic anatomy prior to vaginal septotomy. Patients with recurrent stenosis of the vaginal septum after surgery can safely undergo re-resection of the septum with preserved ability to conceive and maintain pregnancy[7]. This is a very important aspect to consider since the first case is an adolescent with plans of future fertility. On the other hand, the second case underwent total hysterectomy. Either total or unilateral hysterectomy may be required in cases in which septal resection is not possible[24] and may also be considered in patients with recurrent stenosis and severe endometriosis or uterine infection or in patients who do not wish further pregnancies.[7] The second case, having severe pelvic endometriosis and pelvic inflammatory disease, with consideration that she is in the perimenopausal age group with no plans of childbearing, a definitive management in the form of a total hysterectomy with salpingo-oophorectomy was carried out. In our review of literature, none of the cases of Herlyn Werner Wunderlich Syndrome were managed by a total abdominal hysterectomy with salpingooophorectomy, primarily because only our second case was diagnosed in the perimenopausal stage and thus was offered a definitive management.

CONCLUSION

Mullerian duct anomalies such as Herlyn-Werner-Wunderlich Syndrome are rare. It classically presents in adolescent girls, soon after menarche but cases have been reported even years after menarche such as in our first case. The diagnosis, however, should not be disregarded even in perimenopausal patients as it can still be present such as in the second case. Mullerian duct anomalies often times have complex presentations and problems of diagnosis arise hence in 62% of these cases, the use of more than one imaging method is necessary. Ultrasound has proven to be a very useful modality for the diagnosis of Mullerian duct anomalies and may be used as an initial imaging technique to evaluate the uterus and other pelvic structures being the least invasive and most cost effective. 2D Ultrasonography has advantage of low-cost, accessibility and real-time imaging. 3D ultrasound has multiplanar views – coronal being the best and valuable in diffentiating the different types of MDAs and may be comparable with MRI if a bimanual pelvic examination and careful cervical imaging is performed. However, MRI is still considered a modality of choice for both diagnosis and accurate surgical planning of Mullerian duct anomalies. Surgical excision of the obstructing septum and the drainage of retained blood is the definitive treatment especially in young patients such as in our first case. The resolution of obstruction gives immediate relief of acute symptoms and would prevent long-term complications like endometriosis, which would affect the chances of fertility in the future. Either total or unilateral hysterectomy may be required in cases in which septal resection is not possible and may also be considered in patients with severe endometriosis, uterine infection or in patients who do not wish further pregnancies such as in our second case. Herlyn Werner Wunderlich Syndrome may have different presentations, diagnostic approach as well as management revealing a gamut of how the syndrome may appear and be treated. ■

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