Herlyn-Werner-Wunderlich Syndrome (Obstructed Hemivagina and Ipsilateral Renal Anomaly) presenting with acute abdomen: A case report

Ricah Mae A. Capitania MD
Department of Obstetrics and Gynecology, Northern Mindanao Medical Center, Cagayan de Oro, Philippines

ABSTRACT

Herlyn-Werner-Wunderlich (HWW) syndrome, denotes the presence of a double uterus with an obstructed hemivagina and ipsilateral renal agenesis (OHVIRA). It is a rare complex of structural abnormalities of the female urogenital tract.

We are presenting a case of 15-year old G0, with this syndrome. She presented with acute abdomen. On transvaginal ultrasound, there was presence of a huge bulbous cystic structure that displaces the uterus towards the left side, with a biloculated cystic mass in the right ovary. The patient was immediately scheduled for emergency exploratory laparotomy which revealed bulbous mass noted at the cervical area of a uterine didelphis, a distended hemivagina and right hematosalpinx with grossly normal ovaries. She subsequently underwent hemivaginal septal resection and drainage of the hematometrocolpos and hematosalpinx. Patient was discharged on the 4th hospital day and was advised vaginal dilatation with the use of a candle covered with a condom for 1 week to prevent vaginal narrowing.

Keywords: **OHVIRA, Herlyn-Werner-Wunderlich syndrome, uterine anomaly**

INTRODUCTION

Herlyn-Werner-Wunderlich syndrome that shows an obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome is a rare complex of structural abnormalities of the female urogenital tract. Common presenting symptoms include recurrent pelvic pain, dysmenorrhea, an enlarging pelvic mass and abnormal vaginal discharge. Occasionally, patients may present at menarche with pelvic pain secondary to hematocolpos, while others may present with pain, fever and abscess formation. The diagnosis is delayed sometimes.

In this study we outline the clinical course and the management of a patient with HWW-OHVIRA.

Objectives:

1. To present a case of Herlyn-Werner-Wunderlich Syndrome (Obstructed Hemivagina and Ipsilateral Renal Anomaly) in a 15-year-old; and
2. To discuss the etiology, clinical manifestations, diagnosis, and management of Herlyn-Werner-Wunderlich Syndrome.

CASE PRESENTATION

This is a case of LR, a 15-year-old single, Filipino, Roman Catholic from Talakag, Bukidnon, Philippines admitted for the first time in this institution on June 25, 2019, with a chief complaint of Hypogastric pain, with an unremarkable past medical or surgical history.

Patient claims to have a family history Hypertension and Diabetes Mellitus in the maternal side. She is the first child of a brood of 3, and is a grade 10 student. She claims no suicidal ideations and depression. She does not smoke or drink alcoholic beverages, and denies use of illicit drugs.

Two months prior to admission (PTA), patient had onset of hypogastric pain, mainly on the right lower quadrant area, characterized as crampy, localized, non-radiating, on and off, pain score of 2/10, spontaneously resolved without medications. No consult done.

Two days PTA, hypogastric pain recurred now characterized sharp, localized at the hypogastric area, persistent with increasing, pain score 7-10/10, this prompted consultation at local hospital, she was advised to have transvaginal ultrasound and she was prescribed with Mefenamic Acid 500 mg tab every 8 hours for pain which temporarily relieved the pain.

On the morning prior to admission, still with persistent pain, transvaginal ultrasound was done with results of: There is a huge bulbous cystic structure measuring around 19.1 x 7.62 x 7.3 cm seen occupying the pelvic region. It displaces the uterus towards the left side. 'The uterus measures around 4.21 x 2.05 x 2.46 cm with distinct endometrial stripe measuring around 0.43 cm. A biloculated cystic mass measuring around 6.8 x 5.5 x 3.7 cm is seen lateral to the aforementioned bulbous mass and inseparable from the right ovary. The left ovary is not visualized. No free fluid seen. Impression: Normal sized uterus with thin endometrium. Huge Bulbous cystic pelvic mass as described above may present hydrosalphinx. Cannot rule out ovarian origin of the mass. Biloculated cystic right adnexal mass likely right ovarian in origin. May do CT Scan or MRI for further evaluation.

The patient subsequently went to the OB-ER due to the persistent pain.

Pertinent physical examination findings were unremarkable. Speculum examination showed a cystic mass bulging at the right lateral wall of the vagina measuring 4x4 cm. Internal examination showed presence of right adnexal mass that is 17 x 10 cm in size with tenderness on palpation.

The patient was then admitted as a case of Acute abdomen secondary to Ovarian New Growth, Probably Ruptured, Gravida 0.

On the day of admission, the laboratory work-up was done with the following result: leukocytosis with 10.83x103/uL with neutrophilic predominance, decreased hematocrit at 35.60%, decreased MCV at 81.30 fL, increased MCHC at 35.4 g/dL. The patient was immediately scheduled for emergency exploratory laparotomy.

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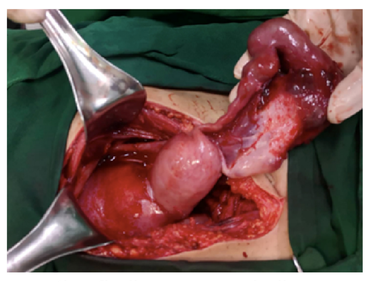

Figure 1. Intraoperative findings were a bulbous mass noted at the cervical area of a uterine didelphis, measuring 15 x 8 cm, a distended hemivagina and right hematosalpinx with grossly normal ovaries.

At this point a Reproductive, Endocrinology and Infertility Specialist was called for intraoperative evaluation and a needle was inserted in the hemivagina, which revealed a bloody aspirate. Patient then underwent hemivaginal septal resection and drainage of the hematometrocolpos and hematosalpinx amounting to around 500 cc.

Postoperatively, she was given Clindamycin 300 mg tab TID per orem. A whole abdomen ultrasound showed ipsilateral renal agenesis. Patient was discharged on the 4th hospital day and was advised vaginal dilatation with the use of a candle covered with a condom for 1 week to prevent vaginal narrowing.

The final diagnosis for this patient is Gravida 0, Herlyn-Werner-Wunderlich Syndrome (Obstructed Hemivagina Ipsilateral Renal Agenesis).

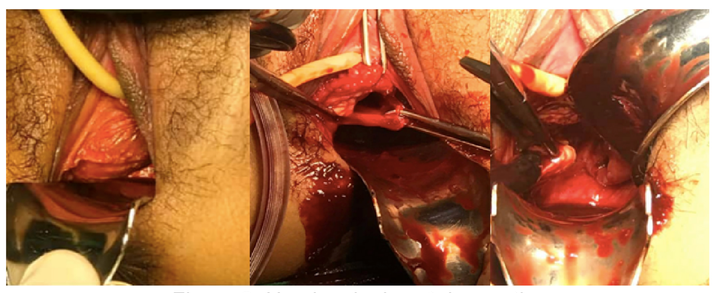

Figure 2. Hemivaginal septal resection.

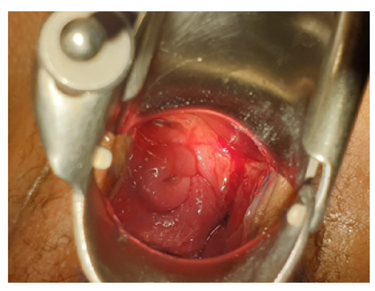

Figure 3. Cervix, 1 week, post operative.

CASE DISCUSSION

Abnormalities of the uterus are categorized as lateral fusion defects and occur due to disordered duct fusion and septal resorption. The presence of a double uterus with an obstructed hemivagina and ipsilateral renal agenesis (OHVIRA), also referred to as Herlyn-Werner-Wunderlich (HWW) syndrome, is a complex anomaly associated with changes in the fusion of the mullerian ducts and a concomitant Wolffian abnormality. (3)

Clinical symptoms vary depending on the ureterovaginal relations in individual cases, but the syndrome can be described generally in three groups. Group 1 patients have complete unilateral vaginal obstruction without uterine communication, resulting in a paravaginal mass and symptoms of severe dysmenorrhea and lower abdominal pain. Menses are regular. Group 2 patients have an incomplete unilateral vaginal obstruction without uterine communication, the presenting symptoms are lower abdominal pain, severe dysmenorrhea, excessive foul mucopurulent discharge, and, in some instances, intermenstrual bleeding. Group 3 patients have complete vaginal obstruction with a laterally communicating double uterus. They have a paravaginal mas, lower abdominal pain, and dysmenorrhea. Menses are regular. Because menses in this syndrome are rarely irregular, the possibility of this syndrome as a diagnosis can be easily overlooked. A careful pelvic examination is necessary to make the correct diagnosis. (1)

Magnetic Resonance Imaging (MRI) is the gold standard for diagnosis. However, three-dimensional ultrasound is also beneficial and has been considered to be equivalent in most circumstances (3). MRI is more sensitive in detecting the uterine contour, the shape of the intrauterine cavity, and the character of the septum compared to other imaging modalities, but it is less adequate in diagnosing endometrioisis, pelvic inflammations, and adhesions, so it has been suggested that the gold standard of diagnosis should be laparoscopy (4).

Careful excision of the vaginal septum is the treatment of choice for a unilateral vaginal obstruction. Prophylactic antibiotics should be administered before surgery. After opening the vaginal pouch, the surgeon should use suction and lavage to remove the pooled blood and mucus. Because the obstructing septum is usually thick, removal can be difficult. Clamps should be used to isolate a generous vaginal pedicle while the suture is being tied in place to prevent slippage of tissue. Such pedicles retract during healing, and formation of vaginal stenosis is avoided. In most instances, surgery is restricted to excision of the septum, and abdominal exploration is unnecessary. In addition to relief of pain due to obstruction, surgery also reduces the chance of pelvic endometriosis due to retrograde menstrual seeding (1).

Reproductive performance for patients with this disorder is usually consistent with that of patients with a double uterus unless the delay in diagnosis and resection of the obstructing septum has been sufficient to destroy tubal function or to cause the development of endometriosis (1).

CONCLUSION

Awareness of these syndrome variants is important for health-care providers for both diagnostic and therapeutic perspectives so unnecessary laparotomies may be avoided. For inexperienced surgeons and sonographers, the diagnosis is likely to be missed because of the presence of normal menstruation and non-specific abdominal pain. Accurate diagnosis and immediate surgical intervention can help prevent complications and preserve future fertility.

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