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Case Report

Complete Uterine Septum with Duplicate Cervix, Longitudinal Vaginal Septum and Unilateral Vaginal Obstruction (Herlyn-Werner-Wunderlich Syndrome): A Case Report of a Mullerian Duct Anomaly

Leili Hafizi[1], **MD**; **Seyedeh Azam Pourhoseini**[1*], **MD**; **Seyedeh Sepideh Hosseini**[1], **MD**

1 Department of Obstetrics and Gynecology,School of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran

**Corresponding author:* Seyedeh Azam Pourhoseini, School of Medicine, Mashhad University of Medical Sciences, Postal code: 9183897449, Mashhad, Iran.
Tel/Fax: 09153121341; **Email:** pourhoseinia@mums.ac.ir

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Abstract

Introduction: Herlyn-Werner-Wunderlich Syndrome (HWWs) is a rare variant of Mullerian duct anomalies. It is associated with a wide range of gynecological and obstetric complications, such as urinary incontinence, urinary retention, endometriosis, pelvic infection, pelvic pain and infertility.
Case presentation: We conducted the present study to investigate and manage HWWs with pelvic pain. The surgery was performed on the operative room of Imam Reza Hospital in April 2018.
Conclusion: The reported case is related to Herlyn-Werner-Wunderlich Syndrome, a rare congenital Mullerian duct anomaly, which it is not included in the current classification system of the Mullerian abnormalities of the American Fertility Association and is based on the typical pattern of caudal evolution towards cranial.

Keywords: Mullerian aplasia, Uterus, Septum, Cervix

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1. Introduction

Uterine anomalies have been reported in 2 to 4% of women of reproductive age (1). These anomalies may be asymptomatic or associated with various obstetric and gynecological complications (2).

Uterine septum is a consequence of impairment in canalization or reabsorption of septum between the two Mullerian ducts. The severity of the disorder range from a small central septum to a complete disorder in septal absorption, which results in a complete uterine septum with a longitudinal vaginal septum (3).

Herein, we reported a case of Herlyn-Werner-Wunderlich Syndrome, which is a rare Mullerian duct anomaly that cannot be included in the current classification system of the Mullerian disorders of the American Fertility Association and is based on the typical pattern of unilateral caudal evolution towards cranial.

2. Case Presentation

A 16-year-old multigravida woman referred to gynecologic department of Mashhad University of Medical Sciences, Iran in April 2018.

She was married for eight months. She presented with dysmenorrhea and pelvic pain two years ago. Furthermore, she complained of dyspareunia and bad smell of vagina since she was married. Gynecologic history indicated menarche at 14 years of age, followed by 2 years of irregular menses.

To evaluate the patient›s complaint, ultrasonography of uterus and adnexa was requested in which bicornuate or septal uterus was reported. According to the probable report of the bicornuate uterus, ultrasound of the kidneys and the urinary system was also requested, which revealed right kidney agenesis.

The patient was taken to the operating room for a procedure involving both laparoscopy and hysteroscopy. The surgery was done in the operative room of Imam

Reza Hospital affiliated to Mashhad University of Medical Sciences with the Olympus system and general anesthesia.

Before the surgery, pelvic examination was performed under anesthesia. In this examination, a small cervix was observed through a needle hole (the entire vaginal wall and ceiling were examined carefully for the presence of double cervical cysts-nothing was detected). On the right side of the vaginal wall, a cystic mass, which was stretched from the cervix to the edge of the hymen was observed and touched (Figure 1).

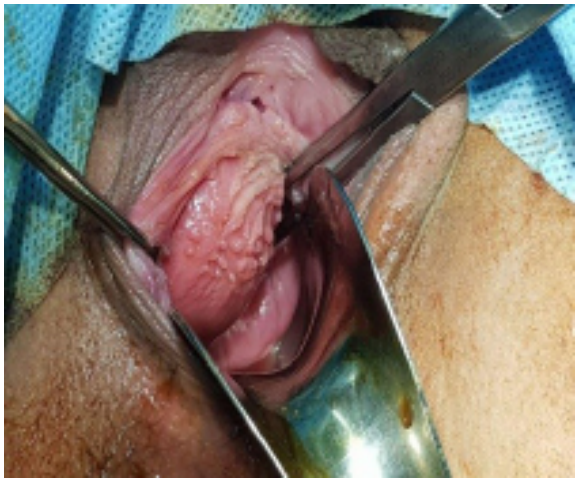

Figure 1: The figure shows cystic mass, stretched from the cervix to the edge of the hymen (right hemivaginal obstruction).

Subsequently, the patient undergone laparoscopy. Laparoscopic abdominal and pelvic explorations were normal. Adnexa was normal on both sides. Only the fundus of the uterus with a broad view (similar to the septal uterus) was observed (Figure 2).

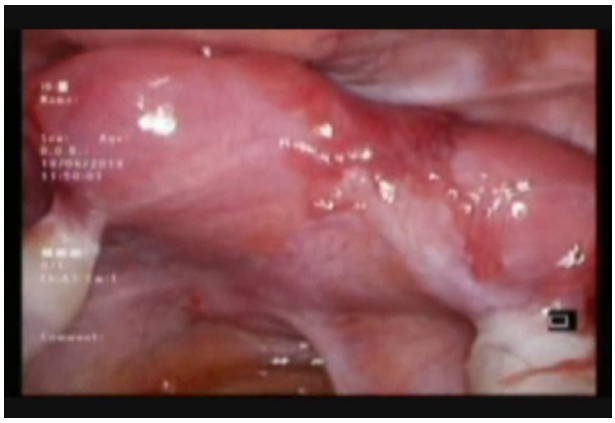

Figure 2: The figure shows Uterus.

Hysteroscopy was performed by inducing a small hole in the cervix to examine the uterus cavity. In the panoramic view, a small cavity was seen with an ostiuma indicating a hemi cavity.

Afterwards, in the right vaginal wall at the maximum bulging, a two-centimeter long incision was created and a large amount of pus was removed.

The inside of the cavity was then washed with normal saline. Following that, inside the cavity was checked with a hysteroscopy and longitudinal vaginal septum with right obstruction was confirmed. Another small cervical hole was observed at the end of the blocked right hemi vagina with vaginoscopy, which was seen by entering the right hemi uterus with the single ostiuma.

At this stage, with the diagnosis of complete vaginal septum with right hemivaginal obstruction, the longitudinal vaginal septum (Figure 3) and cervix were removed utilizing LigaSure. Subsequently, uterine septum resection was performed using an L-shape anceresectoscope. Figure 4 represents the depicted contour of the patient›s anomalies.

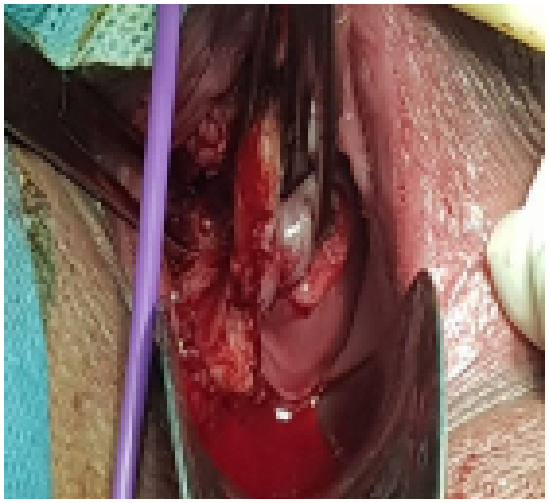

Figure 3: The longitudinal vaginal septum was removed with LigaSure.

The patient was recommended to undergo hysteroscopy again to examine the uterus cavity and cervical position after two months.

Three months later, the patient underwent the second hysteroscopy; there was a 2 -cm wide septum in the fundus resected with a resectoscope.

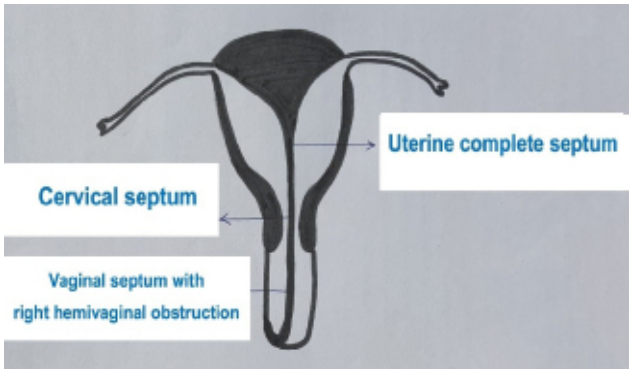

Figure 4: The figure shows Mullerian anomaly of the patient.

3. Discussion

Herlyn-Werner-Wunderlich Syndrome (HWWs) is classified as Class III Mullerian dysgenesis. It is characterized with uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis .This abnormality accounts for 5% of the total Mullerian dysgenesis (4). The Mullerian ducts of the female embryo fuse together in the midline and form the uterus, cervix and upper vagina at 8 weeks of pregnancy. Once they fail to fuse, they produce two hemiuretri and hemicervices.

Wolffian ducts play an important role in the development of internal genital organs and kidneys. They are inductor elements for adequate fusion of the Mullerian ducts. If one of the Wolffian ducts is absent, the ipsilateral kidney and ureter will fail to fuse in the midline (4).

HWWs usually present after menarche with progressive pelvic pain and dysmenorrhea secondary to haematocolpos caused by obstructed vagina (5). Regarding our subject, they presented with dysmenorrhea and pelvic pain from 2 years ago.Uterine septum is the consequence of impairment in canalization or reabsorption of septum between the two Mullerian ducts. The severity of the disorder range from a small central septum to a complete disorder in septal absorption, which results in a complete uterine septum along with a vaginal longitudinal septum (3).

Mullerian duct anomalies may be concomitant with renal or skeletal system anomalies. The kidney system anomalies are found in 20-30% of people with Mullerian duct anomalies, thus, all people with these anomalies should be examined via ultrasonography or intravenous pyelography to diagnose any renal disorders (5). In the reported patient, due to the reported bicornuate uterus, the urinary system was examined in which the unilateral renal agenesis was reported.

Even though certain studies have indicated that since canalization and wall reabsorption between the two Mullerian ducts occur after the development of the urinary system, the septal uterus is not commonly associated with renal anomalies. However, in the reported patient, one-sided renal agenesis was reported in ultrasound. Mullerian anomalies may be asymptomatic or accompanied with symptoms such as amenorrhea, pelvic pain, dysmenorrhea, dyspareunia, and undesirable obstetric complications such as frequent abortions, preterm delivery. The most prevalent indication of surgical interventions to repair these defects is the presence of pelvic pain and undesirable effects of pregnancy (6). In our patient, laparoscopy was performed due to pelvic pain and dysmenorrhea. Mangla and colleagues reported a 35-year-old patient with dyspareunia and menorrhagia whose radiology report indicated didelphys or bicornuate uterus. She underwent laparoscopy, hysteroscopy and a complete uterine septum through which a double cervix and a complete vaginal septum was diagnosed. Due to a history of dyspareunia, the vaginal septum was resected and the patient became ill with the need to evaluate further interventions (3). Regarding our patient, a complete resection of the vaginal septum was performed along with resection of uterine septum and both cervix were unified by removing the cervical septum and re-evaluation of cervical uterus was postponed to two months following the surgery.

Choosing an appropriate treatment in these anomalies has always been challenging. Previously, a complete resection of the uterine septum was supported; however, some experts have recommended the removal of septum only in patients with the history of frequent abortions due to the probable dangers such as uterine rupture, scar formation and cervical insufficiency (7). There are different opinions regarding the removal of cervical septum at the same time with the removal of uterine septum. A number of researchers refuse to remove cervical septum due to the risk of cervical failure.

In a case report of Ribeiro and co-workers, a 19-year-old woman was examined for dysmenorrhea who was diagnosed with complete uterine septum, double cervix and complete vaginal septum. The vaginal septum removal, unifying cervix and uterine septum removal was performed for the patient (7). Additionally, in our subject, due to the presence of small cervical holes and regarding the history of dysmenorrhea, we decided to remove the cervical septum. Sonawane and Bhavsar also reported a 31-year-old woman with a history of five-year-old primary infertility and dysmenorrhea, diagnosed with complete uterine septum, double cervix and complete vaginal septum.

She had undergone complete vaginal septum resection two years before, and underwent uterine septal resection surgery with resectoscope from the upper part of the inner hole of the cervix and the cervical septum remained intact (8). In our patient, septal resection of the uterus, cervix and vagina was performed in one surgical procedure simultaneously.

4. Conclusion

Unusual uterine anomalies has attracted a great deal of scientific attention and still needs to be further studied; therefore, we understand embryology better and root out these disorders. Due to the low prevalence of these anomalies, there is little scientific evidence to determine the best treatment in this regard. In «complete uterine septum with cervical duplication and longitudinal vaginal septum» anomalies, longitudinal vaginal septum resection can easily be performed, yet septum removal requires hysteroscopy skills, and is recommended in all the patients with undesirable fertility outcomes. There are different opinions about the unifying cervix concomitant with uterine metroplasty in the patients with complete uterine septum along with cervical duplication. Some specialists do not agree with it due to the risk of cervical failure, meanwhile, successful pregnancy outcomes without complications in the next pregnancy have been published in case reports and new articles after cervical septum resection supporting it.

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