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Case Report | Olgu Sunumu

PREVIOUSLY MISSED DIAGNOSIS OF HERLYN-WERNER-WUNDERLICH SYNDROME (OHVIRA SYNDROME) AND VAGINOSCOPIC SURGERY ACCOMPANIED BY FLUOROSCOPY

HERLIN-WERNER-WUNDERLICH SENDROMU (OHVIRA SENDROMU) VE FLOROSKOPI EŞLIĞINDE VAJİNOSKOPİK AMELİYATI ÖNCEDEN ATLANAN TANI

İlknur Benli Cesur[1], Zerrin Ozcelik[1], Dilek Kaplanoglu[2], Mustafa Kaplanoglu[3*]

[1]University of Health Sciences, Adana City Training and Research Hospital, Department of Pediatric Surgery, Adana, Türkiye. [2]Yuregir Goverment Hospital, Department of Obstetric and Gynecology, Adana, Türkiye. [3]Cukurova University, School of Medicine, Department of Obstetric and Gynecology, Adana, Türkiye.

ABSTRACT
Herlyn-Werner-Wunderlich (HWW) syndrome is a rare congenital malformation of the female urogenital tract characterized by uterus didelphys and occluded hemi-vagina and ipsilateral renal agenesis (OHVIRA) syndrome. We present a 16-year-old girl with abdominal pain, a rare and unusual treatment for this syndrome. The patient who underwent laparotomy with the preliminary diagnosis of a mass of approximately 15 cm in the left uterine segment, was diagnosed with USG and MRI after reocclusion of the existing vaginal segment and recovered after fluoroscopy and vaginoscopic segmental resection by us. The diagnosis of WWH syndrome was confirmed by ultrasonography and MRI. The case presented is the first vaginoscopic hymen-sparing surgery performed with fluoroscopy in the literature for rare HWW Syndrome.
Keywords: Pelvic pain, dysmenorrhea, mullerian abnomaly, flouroscopy, vaginoscopy

ÖZ
Herlyn-Werner-Wunderlich (HWW) sendromu, uterus didelfisi ve tıkalı hemi-vajina ve ipsilateral renal agenezis (OHVIRA) sendromu ile karakterize, kadın ürogenital yolunun nadir görülen bir konjenital malformasyonudur. Bu sendromun nadir ve sıra dışı bir tedavisi olan karın ağrısı şikayeti olan 16 yaşında bir kız çocuğu sunuyoruz. Sol uterin segmentte yaklaşık 15 cm'lik kitle ön tanısı ile laparotomi uygulanan hasta USG ve MR ile mevcut vajinal segmentin reoklüzyonu yapılarak tarafımızca floroskopi ve vajinoskopik segmental rezeksiyon sonrası iyileşti. WWH sendromu tanısı ultrasonografi ve MRG ile doğrulandı. Sunulan olgu literatürde nadir görülen HWW Sendromu için floroskopi ile yapılan ilk vajinoskopik kızlık zarı koruyucu cerrahidir.
Anahtar Kelimeler: Pelvik ağrı, dismenore, müllerian anomali, floroskopi, vajinoskopi

***İletişim kurulacak yazar/Corresponding author:** Mustafa Kaplanoglu; Cukurova University, School of Medicine, Department of Obstetric and Gynecology, Adana, Türkiye.
Telefon/Phone: +90 (322) 338 60 60 **e-posta/e-mail:** mkaplanoglu@cu.edu.tr
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Introduction

Obstructed hemivagina and ipsilateral renal anomaly syndrome (OHVIRA Syndrome (OS)), also known as Herlyn Werner Wunderlich Syndrome, is a rare pathology that accounts for 0.16-10% of mullerian system anomalies. It was first reported by Purslow in 1922. Wunderlich in 1976, and Herlyn and Werner in 1983 reported similar cases and the name of this pathology began to be called "Herlyn-Werner-Wunderlich Syndrome".[1] In 2007, Smith and Laufer suggested that the name of this syndrome should be OHVIRA Syndrome as an abbreviation of existing pathologies.[2]

It has different variants that include uterine didelphis or complete uterine septum and renal anomalies (agenesis, dysgenesis).

OS is a pathology of the development of the female urogenital system. Here, the theory that the lower part of the vagina develops from the urogenital sinus, and the upper part of the vagina, uterus and tubae develops from the paramesonephric duct is insufficient to explain the development of OS. Acien's theory tried to explain this. According to Acien, the female genital tract consists of synovaginal bulbs that emerge from the distal mesonephric ducts connected to the urogenital sinus.

Therefore, pathology of mesonephric duct development may also disrupt bilateral paramesonephric duct development and cause an obstructed hemivagina and renal agenesis/dysgenesis.[3]

In our presented case, a 16-year-old patient with a regular menstrual cycle applied to the emergency department with acute abdomen. After the evaluation, an adnexal mass was detected and laparotomy was performed. In the laparotomy, uterine and possible vaginal anomalies were detected, abdominal hysterotomy was performed and hemivaginal patency was achieved. The patient applied to the pediatric surgery polyclinic again due to the recurrence of symptoms in the 4th postoperative month, and the patient was consulted to the Gynecology and Obstetrics Clinic, where it was found that the hemivaginal opening was closed. Then, vaginoscopic hymen-sparing segmental vaginal septum resection was performed using a hysteroscopy equipment. OS diagnosis, clinical treatment and especially minimally invasive treatment are presented through this case.

Case Report

A 16-year-old female adolescent patient with a gynecological age of 4 applied to the pediatric surgery clinic in an external center with acute abdomen symptoms. After the evaluation, laparotomy was performed with the preliminary diagnosis of adnexal mass. A uterine anomaly (uterus didelphis?) was detected at laparotomy. While the right uterine segment was in normal size, the left uterine segment was found to be approximately 15 cm in diameter. A hysterotomy was performed on the patient and the hemivaginal side of the abdomen was perforated, blood flow was ensured vaginally, and the operation of the patient was terminated when a Foley drain exiting the vagina was placed. The patient, whose drain was removed 1 week later, applied to the same clinic again 4 months later due to inguinal pain. In the ultrasonography and MRI evaluation, uterus didelphys, two uterine cavities, cervix, left renal agenesis and partial vaginal atresia were detected, and the diagnosis was appeared to be OS. According to the OS classification made by Zhu et al in 2015, Class 1.1 anomaly (Completely obstructed hemivagina, no communication between the two hemiuteri, hemivagina containing a blind end) was detected.[4] In the MRI evaluation, the right uterus was 41x22 mm and the endometrium was 5 mm. Left corpus uteri and cervix was full with blood and was 10x5 cm in size (hematometra), and blind vagina was full with blood (hematocolpos) and was 8x4 cm. Both ovaries were normal. Right kidney was observed in normal location and size, left kidney was not observed (agenesis) (Figure 1). The mass was thought to be due to the closure of the vaginal opening and was referred to an upper center Pediatric Surgery clinic. Due to the exacerbation of the patient's pain, who was followed up, an emergency operation was decided, but the operation could not be performed due to the Covid-19 pandemic conditions. For this reason, a drain from the abdominal wall to the uterine cavity was placed to gain time for the patient to relax and for proper surgical planning. The patient was then consulted to us. First, vaginoscopy was performed in order to plan the surgical treatment method. There, a single cervix was seen and the scar tissue of the previous vaginal septum incision was observed in the lower left part of the cervix.

It was decided to perform hemivaginal septum resection with hysteroscopy, since the patient's family particularly wanted the hymen to be preserved. Signed ınformed consent form was obtained from her family members. A hemivaginal septal resection was planned and performed by vaginoscopy for the patient whose full cavity (endometrial and vaginal) size and septum localization were determined in X-Ray scopy by giving radiopaque material preoperatively through the drain (Figure 2; Figure 3). During the procedure, the mass was filled with a radiopaque material, and the hemivaginal septum was resected directly, without damaging other tissues, accompanied by X-Ray scopy. (Figure 4). The operation was terminated with the placement of the drain coming out of the vaginal route (inflated with 80 cc of fluid) and the removal of the abdominal drain. During these procedures, the hymen was preserved. The patient was discharged on the 1st postoperative day, and the vaginal drain was removed 20 days later and the patient is being followed up by our clinic without any problems. (Figure 5).

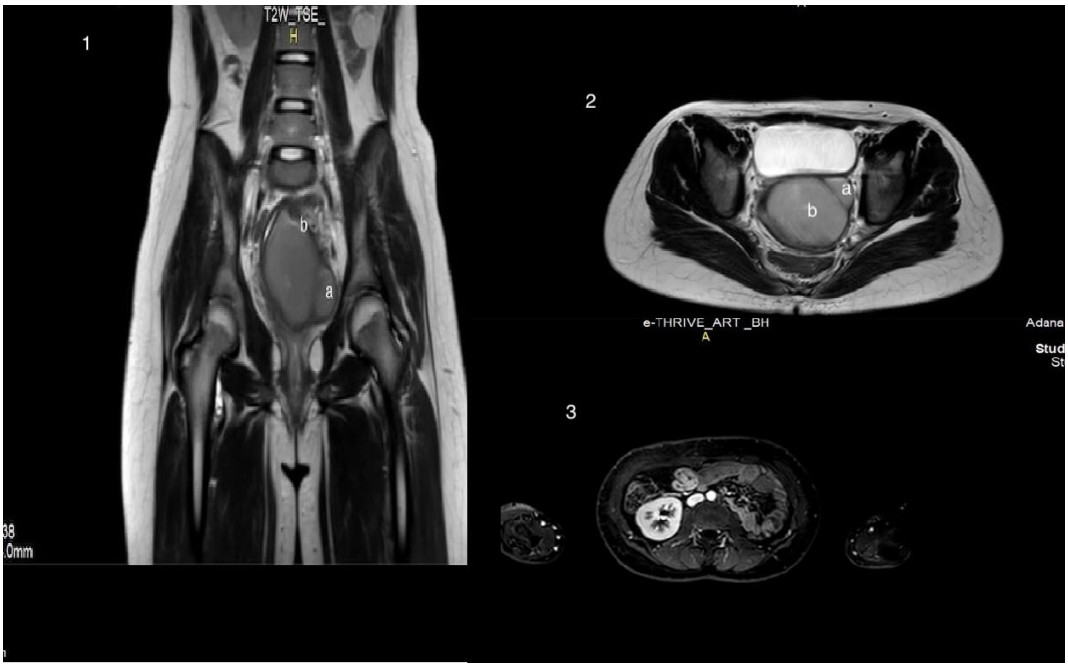

Figure 1. Longitudinal (1) and sagittal (2) MRI view of drained (a) and non-drained (b) uterine cavity and MRI view of unilateral renal agenesis (3)

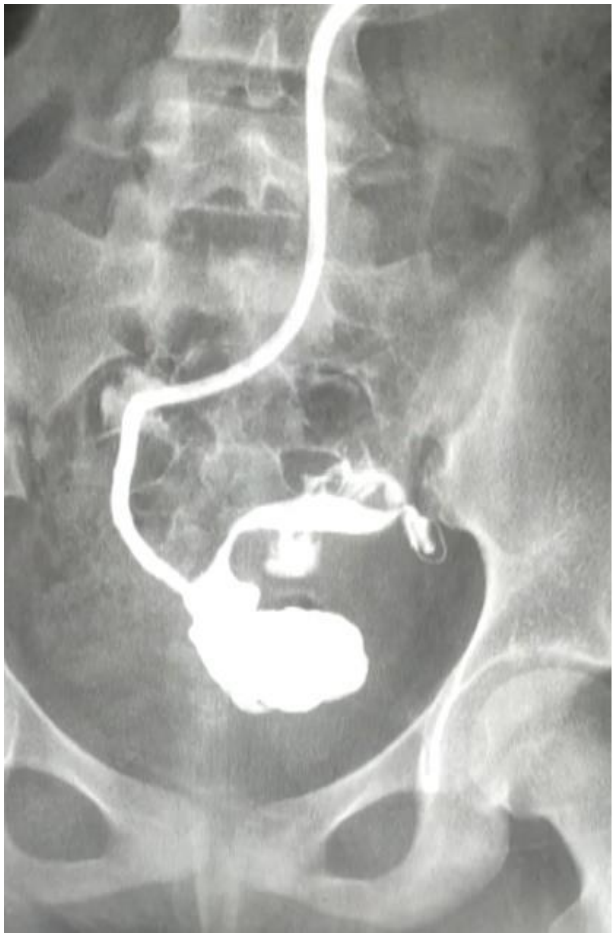

Figure 2. Radiopaque passage from dilated uterine cavity, hemivagina and single tube is observed with X-Ray Scopy)

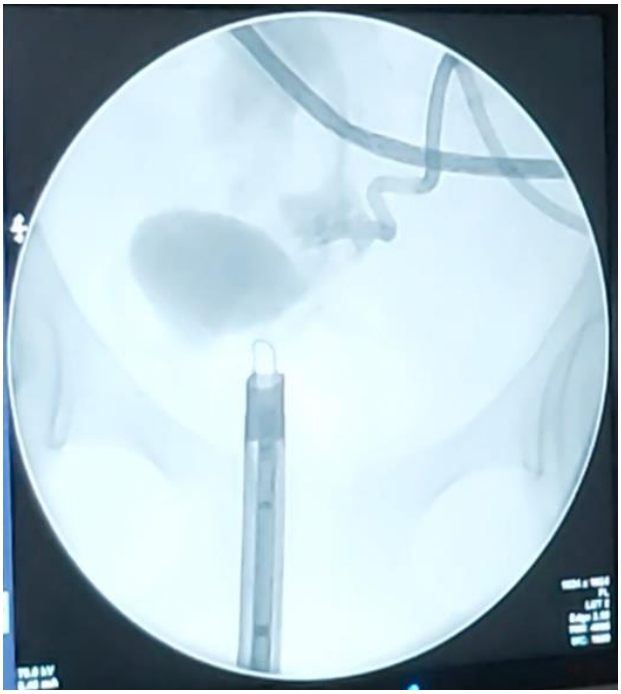

Figure 3. The segment to be dissected was determined with the help of scopy during hysteroscopy

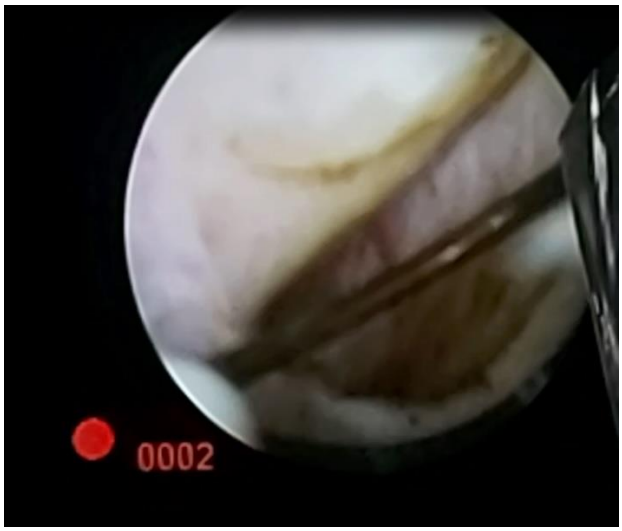

Figure 4. With hysteroscopy, the hemivaginal septum can be opened and the upper vagina can be seen. Since the coagulum was emptied with the help of a drain before, there was no coagulum discharge.

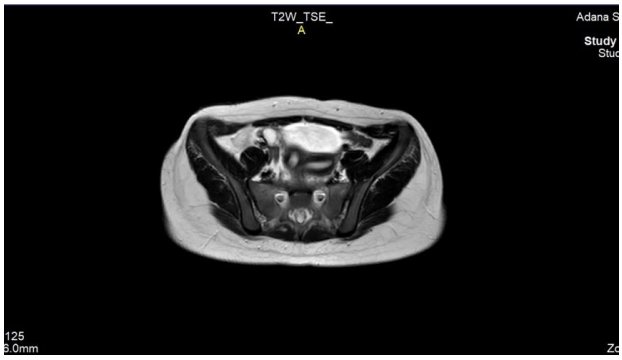

Figure 5. Post-surgical follow-up MRI evaluation. The uterine cavity is emptied, but the cavity volume tension continues

Discussion

The theoretical and widely accepted view is that the development of the female genital tract occurs by the merging of the mesonephric (Wolf) and Paramesonephric (Muller) ducts and then their canalization. According to this theory, while the paramesonephric ducts on both sides merge in the midline and form the uterus, cervix and upper part of the vagina, the lower part of the vagina is formed from the urogenital sinus.[1] However, this hypothesis falls short of explaining some vaginal and uterine developmental anomalies (such as OS). Here, all components of OHVIRA can be explained using Acien's hypothesis. Here, the mesonephric duct has a critical role in the development of the paramesonephric duct. Coordination at all stages of development is essential for full urogenital development. The mesonephric duct and paramesonephric duct are in communication with different mediators and hormones at the stage when the mesonephric duct regresses to the cranial and forms the renal system, and at every stage of the formation of the paramesonephric duct. A developmental defect in the mesonephric duct will both adversely affect kidney development and disrupt the correct positioning of the paramesonephric duct with which it is paired, and this may cause vaginal (septum, incomplete development) and uterus (didelphis) anomalies. The most common defect in the cranial regression of the mesonephric duct is cystic remnants in the vaginal wall (Gardner Cysts).[3]

Similar pathologies can also be detected in males. Zinner Syndrome is a rare congenital anomaly seen in males. This syndrome is characterized by male infertility, unilateral agenesis, seminal vesicle cysts, and ipsilateral ejaculatory duct obstruction. The name OSVIRA Syndrome OSVIRA (Obstructed Seminal Vesicle and Ipsilateral Renal Agenesis) is also suggested, based on the abbreviations OHVIRA Syndrome in females. With these findings, it has pathologies that can be explained by Acien's embryological development theory and can be considered as the equivalent of OHVIRA Syndrome in females.[5]

The most common clinical findings in OS are groin/abdominal pain, palpable abdominal mass, and acute abdominal pain with cyclic regular bleeding that begins shortly after menarche. In rare cases, it is encountered with difficulty in urination. The combined evaluation of anamnesis, ultrasonography and MRI findings is often necessary for definitive diagnosis. Laparoscopy is unnecessary for diagnosis. Obstructive hemivagina is the main cause of clinical findings. Hemoperitoneum, hematocolpos, pyocolpos, pelvic adhesion, acute abdomen, tuboovarian abscess, urinary incontinence, long-term endometriosis are possible complications.[6,7] It is clear that definitive treatment is only possible with surgery. Hemihysterectomy, hemivaginectomy or salpingectomy, which are very aggressive treatment options, should be reserved for particularly complicated cases.[8] In patients who do not have advanced complications, such as our case, the route of choice should be hymen-sparing surgery, especially if requested by families, and segmental septum resection with the help of hysteroscopy. We also suggest that in cases where a drain was placed through the abdominal wall, to prevent complications and to make the operation more comfortable, administering a radiopaque material fluoroscopically to ensure that hysteroscopy cuts the entire septum region. In the meantime, if there is no additional pathology, it is unnecessary to perform laparoscopy for the risk of endometriosis that may occur due to retrograde menstruation, especially in this group of patients. Laparoscopic surgery can be planned for patients with ongoing chronic pelvic pain and dysmenorrhea, especially for the diagnosis and/or treatment of endometriosis.

Many patients apply to emergency services with acute abdominal pain more than dysmenorrhea and are taken to surgery with a preliminary diagnosis of a mass before a definitive diagnosis can be made. However, in a limited number of cases, due to the connection between the vaginal or uterine cavities, patients may also apply due to infertility, dyspareunia, and intermittent mucous discharge. In these cases, since we cannot clearly see the semi-blood-filled vaginal cavity, the diagnosis with

ultrasonography is difficult and the diagnosis is often made with MRI.

Our presented case is a patient who had dysmenorrhea, but applied to the emergency department with abdominal pain and was taken to surgery for an adnexal mass. Here, it is aimed to give information about the stages until the final diagnosis and hymen-sparing surgery. Our case is the first case in the literature in terms of the way of diagnosis and especially surgery performed with scopy.

OS is a congenital anomaly of the urogenital system whose treatment is surgical. The diagnosis starts with suspicion, as the symptoms may differ according to the OS variants. Patient evaluation with detailed physical examination, ultrasonography and MRI is essential.[8] Although the treatment is surgical, vaginoscopic vaginal septum segmental resection should be the method to be recommended because of its low cost, ease, minimal invasiveness, short hospitalization time and effectiveness, especially in cases where hymen preservation is desired by the family, as in our presented case. In addition, if the existing vaginal and uterine cavity can be filled with radiopaque material, segmental resection under scopy will protect against possible adjacent structure injuries, especially in cases where finding a safe area for resection is difficult.

Compliance with Ethical Standards

Consent was obtained from the patient.

Conflict of Interest

The authors report no conflicts of interest.

Author Contribution

Authors contributed equally to this work.

Financial Disclosure

None

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