



CASE REPORT

Obstructed Hemivagina and Ipsilateral Renal Agenesis (OHVIRA) Syndrome: A Rare Case Report

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Abstract

Introduction: The Herlyn-Werner-Wunderlich (HWW) syndrome, also known as obstructed hemivagina and ipsilateral renal agenesis (OHVIRA), is a rare form of Mullerian Duct Anomalies (MDA). The patient with OHVIRA syndrome has dysmenorrhea, abdominal pain, and a lump in the belly. Although the exact cause and pathology of OHVIRA syndrome are unknown, early diagnosis and treatment can reduce acute symptoms and preserve normal fertility.

Case Report: This article reports a case of 15 a 15-year-old woman who complained of foul mucopurulent discharge. Ultrasound finding uterus didelphys, right obstructed hemivagina with resultant pyocolpos, and suspected of right renal agenesis. The patient was diagnosed with OHVIRA syndrome. The patient underwent vaginal septal resection, and the pyocolpos was drained.

Discussion: Intraoperatively, a completely obstructed longitudinal vaginal septum was identified on the right hemivagina, with purulent fluid consistent with pyocolpos. After septal resection, adequate drainage was achieved and communication between both hemivaginas was established. Postoperatively, the patient showed clinical improvement, with resolution of vaginal discharge and relief of vaginal obstruction. No intraoperative or immediate postoperative complications were observed. During short-term follow-up, the patient remained asymptomatic, with no evidence of recurrent obstruction or infection.

Conclusion: This case highlights several important aspects of OHVIRA syndrome, including its atypical postpartum presentation, diagnostic challenges, and unpredictable reproductive outcomes. Ultrasonography plays a key role in early detection. Vaginal septal resection and drainage are effective in relieving symptoms and preventing complications. Reporting this rare presentation contributes to improved awareness and timely management of OHVIRA syndrome in adolescents.

Keywords: OHVIRA syndrome, mullerian duct anomalies, vaginal septal resection

INTRODUCTION

Obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome is a rare form of Mullerian Duct Anomalies (MDA). The characteristic triad of this syndrome includes didelphys uterus, obstructed hemivagina and ipsilateral renal agenesis. It was initially described in 1971 by Herlyn and Werner. In 1976, Wunderlich also described a bicornuate uterus with a simple vagina and isolated right hematocervix without connection of the right uterus to the vagina in association with aplasia of the right kidney and ureter [hence, first known as Herlyn-Werner-Wunderlich (HWW) syndrome]^{1,2}. The actual incidence of this syndrome is unknown, however its occurrence is estimated to be 0.1%-3.5%^{3,4}. Of all MDAs, HWW is a rare variant that occurs in only 5% of affected individuals⁵

The cause is unknown, but it has been linked to improper development of the Mullerian and Wolff ducts during embryonic morphogenesis in the eighth week of gestation¹. HWW syndrome is classified into two types based on whether the hemivaginal obstruction is complete or incomplete: Classification 1: completely obstructed hemivagina; Classification 2: partially obstructed hemivagina. These two classifications have very different clinical characteristics⁶.

After a few months of menarche, the patient with OHVIRA syndrome appears with dysmenorrhoea, pain in the abdomen, and a lump in the belly. Although the specific cause and pathology of OHVIRA syndrome are unknown, early identification and therapy can reduce acute symptoms and preserve normal fertility⁷. The reported incidence of OHVIRA syndrome varies widely because many cases remain undiagnosed until adolescence or adulthood. Delayed diagnosis is associated with increased risks of infection, endometriosis, infertility, and adverse obstetric outcomes.⁸⁻⁹

The aim of this article is to report a rare and atypical postpartum presentation of obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome in an adolescent, to describe its clinical, imaging, and surgical findings, and to emphasize the importance of early recognition and appropriate surgical management to prevent long-term gynecologic and reproductive complications.¹²

CASE REPORT

A 15-year-old girl was referred from regional hospital with a diagnosis of Gartner's cyst. She presented with lumps from the genitals appear 1 month after vaginal delivery. She also complained of yellowish and smells discharge from the genitals. After menstruation, smelly blood spots came out. She denied either pain or bleeding during intercourse. There is no history of urinary disorders. The menarche had occurred 2 year before, her cycles were regular (28 days) and the menstrual bleeding usually lasted about 7 days with moderate dysmenorrhea and hypermenorrhea (5-7 times pad changes/day). She had been married since a year ago. She has a 1-month-old boy with a birth weight of 1800 grams. The baby boy was born alive, preterm, and delivered by a midwife. She has no history of disease or surgical intervention.

Abdominal examination was normal. At the gynecologic examination, the septum was visible

on the right proximal side of the vagina, there was a smelly greenish yellow fluid coming out of the septum, there was a multiparous portio on the left side, and there was a greenish yellow flux flowing through the posterior fornix (Fig. 1). The sondage of uterus was 5-cm-anteflexion. Bimanual vaginal examination revealed the vaginal mucosa was smooth, the size of the uterus was within normal limits, there was no palpable adnexal mass, the septum was palpable in the right hemivagina and the portio was palpable in the left hemivagina.

Ultrasonography showed no hematometra, there were two uterine cavities with two separate endometrium and the right kidney was not visualized (Fig. 2). It revealed uterus didelphys and suspect of right renal agenesis. Blood examination including liver function test was within normal limits. Regarding right renal agenesis, uterus didelphys, right obstructed hemivagina with resultant pyocolpos, the case was diagnosed as OHVIRA or HWW syndrome.



Figure 1. Pyocolpos on gynecologic examination.



Figure 2. Ultrasonography (USG) showed two uterine cavities with two separate endometrium and the right kidney was not visualized

DISCUSSION

The exact cause and pathogenesis of HWW syndrome are unknown^{4,7}. These congenital abnormalities are thought to include abnormal development of the Mullerian (paramesonephric) and Wolffian (mesonephric) ducts. In women, the Mullerian duct grows downward towards the midline, crosses the Wolffian duct, comes into contact with each other, and joins to form the uterovaginal canal, forming the fallopian tubes, uterus, and two-thirds of the upper part of the vagina. Uterine hypoplasia or agenesis, unicornuate, didelphys, bicornuate, septate, and arcuate can result from the distal segments of the Mullerian duct not developing or failing to fuse. The Wolffian duct is the kidney's origin and an inductor for Mullerian duct fusion. The cause of unilateral renal agenesis associated with imperforate hemivagina is abnormal development of the tail section of one of the Wolffian ducts^{1-9,18,23-24}.

There were no abnormalities in the appearance of the external genitalia in the reported cases. The anomaly manifests itself in the internal genitalia as a didelphys uterus, with duplication. A pyocolpos is present because the right side of the hemivagina is obstructed. The presence of an obstruction causes pain with each period, also known as dysmenorrhoea. Pain complaints can be felt continuously in more chronic conditions. Endometriosis cannot be ruled out in the long run¹⁰.

Ultrasound and MRI are both very useful imaging techniques for diagnosing this syndrome. Three-dimensional ultrasound can be used both abdominally and vaginally and allows for the viewing of images in three planes orthogonally. This technique appears to provide high-quality images comparable to those obtained by MRI, as well as high precision in the diagnosis of congenital anomalies, at a lower cost and with greater tolerance. In terms of detecting acute and chronic complications, MRI outperforms hysterosalpingography (HSG) and ultrasound. Because of its ability to display better anatomical images of the internal genital organs and high sensitivity to indicate the presence of blood, magnetic resonance imaging (MRI) is the modality of choice for the diagnosis of HWW syndrome and other congenital disorders^{11,24}.

Laparoscopy is still the gold standard for female genital tract abnormalities diagnosis, but it is only used when MRI is unavailable or fails to establish a diagnosis^{11,12}. Laparoscopy is another gold standard diagnostic modality that can provide an added benefit by allowing for hematomocolpos or hematometra therapeutic drainage, vaginal septectomy, or marsupialization. The preferred treatment for HWW syndrome is laparoscopic vaginal septum excision¹². IVP can be used to visualize urinary tracts and provide an overview of kidney function; in this case, we suggested that the renal function was unaffected based on symptoms and a renal function test, so the IVP was unnecessary.

A septectomy procedure is used to treat cases of HWW syndrome that are accompanied by hemivaginal obstruction and secondary hemato/pyocolpos. The primary goal of this procedure is to relieve pain symptoms by increasing menstrual blood flow, resulting in no more hematocolpos. It is hoped that the reproductive process will resume normally in the future. Delays in diagnosis should be avoided as much as possible because they can cause or worsen existing complications^{14,20}.

Based on the physical abnormalities discovered, it was decided to perform surgical

management of the obstructed vaginal septum with vaginal septectomy. In this patient, the vaginal septum was completely longitudinal, preventing access to all sides of the left hemivagina. Septectomy removes the entire septum along the vagina so that menstrual blood from the left hemiuterus can flow freely. Currently, only vaginal septectomy is used to correct the pyocolpos obstruction and associated pain. The patient's didelphys uterus is still not communicating. Although fertility is generally preserved, pregnancy in OHVIRA syndrome is frequently complicated by preterm birth, malpresentation, fetal growth restriction, and increased cesarean section rates^{23-22,25}

In this reported case, she did not suffer infertility but she delivered a preterm and low birth weight baby. According to some authors, pregnancy was possible in 87% of HWW syndrome cases, and 62% of them delivered at term. Premature labor occurs more frequently in patients with OHVIRA syndrome than in the general population, but the rate of miscarriage does not differ. Patients with OHVIRA syndrome can have vaginal births. However, caesarean sections are required in approximately 84% of pregnancies. Breech presentation is the most common indication, occurring in 51% of cases. A possible complication that can increase the rate of caesarean section is fetal growth restriction¹⁵

CONCLUSION

This case demonstrates that OHVIRA syndrome may remain undiagnosed even after pregnancy and delivery, as illustrated by this adolescent patient who presented postpartum with pyocolpos and vaginal obstruction. Despite having untreated OHVIRA syndrome, the patient was able to conceive and deliver a preterm, low-birth-weight infant, highlighting the variable reproductive outcomes associated with this condition. Ultrasonography was sufficient to establish the initial diagnosis in this case, showing uterus didelphys, obstructed hemivagina, and suspected ipsilateral renal agenesis. Surgical management by vaginal septal resection and drainage resulted in complete resolution of symptoms without immediate complications.

This case emphasizes the importance of clinical suspicion in adolescents with atypical vaginal discharge or postpartum complications. Early recognition and timely surgical intervention are crucial to prevent infectious sequelae, gynecologic morbidity, and adverse obstetric outcomes in patients with OHVIRA syndrome.

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