Case Report

DOI:

Unilateral Renal Agenesis and Hemivaginal Obstruction in Uterus Didelphys: A Case Report of OHVIRA Syndrome

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Abstract:

Obstructed hemivagina with ipsilateral renal anomaly (OHVIRA) syndrome is a rare congenital anomaly involving uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis. We present a case of a 15-year-old girl who presented with lower abdominal pain and progressive dysmenorrhea since menarche. Transabdominal ultrasonography suggested uterine duplication with left-sided hematocolpos and absent left kidney. MRI confirmed uterus didelphys with complete vaginal and cervical duplication, left-sided hematometra, hematocolpos, and hematosalpinx. A CT scan confirmed left renal agenesis. Surgical management involved left hemihysterectomy, salpingectomy, and resection of the vaginal septum. The patient experienced immediate relief postoperatively and was counseled on long-term reproductive outcomes. This case highlights the importance of early recognition of OHVIRA syndrome in adolescent females with cyclical pelvic pain. Timely imaging, particularly MRI, and appropriate surgical intervention can prevent complications and preserve fertility.

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Introduction:

Obstructed hemivagina with ipsilateral renal anomaly (OHVIRA) syndrome – also known as Herlyn–Werner–Wunderlich (HWW) syndrome – is a rare congenital Müllerian anomaly characterized by uterine duplication accompanied by an obstructed hemivagina and agenesis or dysplasia of the ipsilateral kidney.[1,2]

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The condition arises from a combined malformation of the paramesonephric (Müllerian) and mesonephric (Wolffian) ducts during embryogenesis, typically between the 8th and 12th weeks of gestation.[3,4] It is estimated to occur in approximately 1 in 20,000 females, though this may be underreported due to asymptomatic or misdiagnosed cases.[5]

Clinically, most patients remain asymptomatic until menarche. Then, symptoms such as cyclical abdominal or pelvic pain, dysmenorrhea, and a palpable pelvic mass due to hematocolpos or hematometra often emerge.[6,7] Vaginal discharge, irregular menstruation, or urinary tract symptoms may also be present, depending on the degree of obstruction and presence of ectopic ureters or infection.[8] Menstrual flow may appear normal due to unobstructed outflow from the contralateral hemivagina, contributing to diagnostic delay.[6]

Transabdominal ultrasonography is usually the initial imaging done which may show uterine duplication, hematocolpos, and renal agenesis.[9] However, magnetic resonance imaging (MRI) is the gold standard for definitive diagnosis, providing excellent delineation of uterine anatomy, vaginal septum, and renal structures.[10,11] Occasionally, computed tomography (CT) may incidentally detect the renal anomaly but is not preferred due to radiation exposure and lower soft-tissue contrast.[12] Laparoscopic or vaginoscopic evaluation further confirms the diagnosis and guides management. [13,14]

Treatment typically involves surgical excision or incision of the obstructing vaginal septum to relieve hematocolpos and restore normal menstrual outflow.[1,15] Timely intervention prevents complications such as endometriosis, infection, and infertility. When appropriately managed, most patients retain normal reproductive potential.[5,14]

Case Description And Management

A 15-year-old girl presented with the complaints of lower abdominal pain and worsening dysmenorrhea that began shortly after menarche at the age of 13. Her menstrual cycles were irregular, and she often experienced pelvic discomfort, which had gradually increased in severity over the past two years. On query, no associated urinary symptoms or previous history of surgery were found.

Given her persistent pain and irregular menstruation, a transabdominal ultrasound was performed. The scan revealed two separate, divergent uterine horns with a large fundal cleft. The endometrial cavities and cervices were clearly separated. The left cervix appeared dilated, containing moderate fluid collection with low-level internal echoes. Mild collection was also noted in the left endometrial cavity. Additionally, the left fallopian tube was dilated and tortuous, also containing low-level internal echoes (Figure 1). The right uterine horn and cervix appeared normal (Figure 2). The left kidney was not visualized, while the right kidney was normal in size, measuring approximately 12.2 cm. Based on these findings, OHVIRA syndrome was suspected.

For further evaluation of the anatomy, an MRI of the pelvis was done, which also revealed complete duplication of the cervix, uterus, and vagina. Hyperintense collections were seen on both T1- and T2-weighted images within the left endometrial cavity, cervix, and upper vagina—suggestive of hematometra and hematocolpos. The left fallopian tube was also dilated and contained similar hyperintense material, consistent with hematosalpinx (Figure 3,4,5).

A CT scan of the abdomen and pelvis was done to assess the urinary system and confirmed the MRI findings. It showed two uterine horns consistent with uterus didelphys, along with left-sided hematometrocolpos, hematosalphinx, and complete agenesis of the left kidney (Figure 6, 7).

After completion of the diagnostic modalities, the patient underwent surgery. A left hemihysterectomy along with left salpingectomy was performed, followed by transvaginal resection of the obstructing vaginal septum. The procedure was uneventful otherwise. Postoperatively, the patient experienced immediate relief from pelvic pain and was discharged in stable condition. She was counseled about the diagnosis, future fertility implications, and the importance of routine follow-up.

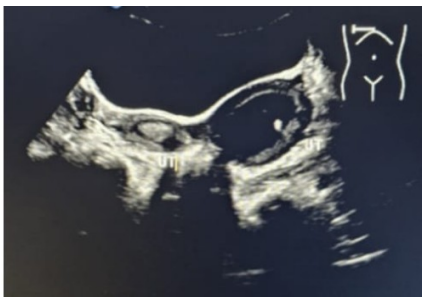

Fig 1: Two separate uterine horn and cervix, moderate collection in left side.

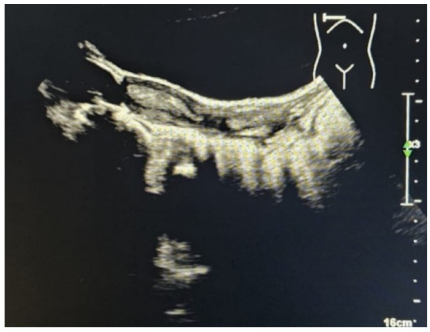

Fig 2: Normal appearance of right uterine horn, cervix.

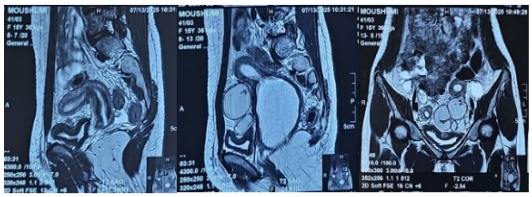

Fig 3, 4, 5: Showing uterine didelphys, hematometrocolpos & hematosalpinx (left), and normal appearance of uterine horn, cervix, vagina on the right.

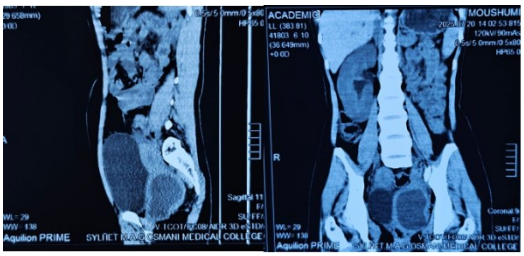

Fig 6, 7: Two uterine horns, uterus didelphys, left hematometrocolpos, left hematocolpos and left renal agenesis.

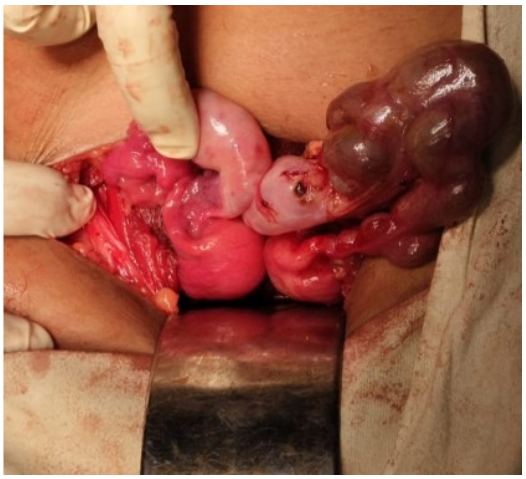

Fig 8: Per-operative visuals of uterus didelphys

Discussion:

Congenital anomalies of the female reproductive tract associated with urinary system malformations present unique diagnostic and management challenges.[5,6] Among these, obstructed hemivagina with ipsilateral renal anomaly (OHVIRA) syndrome is a particularly rare entity that has been increasingly recognized since its first descriptions in the 20th century. Classically, it was termed as *Herlyn–Werner–Wunderlich* (HWW) syndrome in the early 1970s.[16] Wunderlich (1976) and others reported the triad of uterus didelphys (or bicornuate uterus with double cervix), an obstructed hemivagina, and ipsilateral renal agenesis.[16] In 1992, the acronym OHVIRA (Obstructed Hemivagina and Ipsilateral Renal Anomaly) was introduced to emphasize that the ipsilateral kidney abnormality may include not only agenesis but also dysplastic variants.[14] In modern usage, OHVIRA and HWW refer to the same anomaly. The term OHVIRA reflects the embryologic link between the reproductive and renal findings, while HWW is based on the classic clinical triad. [14,16]

The estimated incidence of OHVIRA/HWW syndrome is approximately 1 in 20,000 females.[5] Müllerian anomalies overall occur in about 0.8–4% of the population, and OHVIRA comprises a small fraction of these.[5,12] Most reported cases involve uterus didelphys. One review found this in over 85% of cases, although bicornuate or septate uteri can rarely occur. Likewise our case, patients typically present after menarche, when normal menstruation from the unobstructed side masks the anomaly until retained menses in the blind hemivagina cause pain or a pelvic mass.[6,7]

OHVIRA syndrome is the result of an embryological arrest of the Mullerian and mesonephric ducts at eight weeks of gestation.[1] The mesonephric duct is essential for both kidney development and normal fusion of the ipsilateral Müllerian duct. Failure of the Wolffian duct on one side leads to renal agenesis or dysplasia, while also displacing the ipsilateral Müllerian duct laterally and preventing its normal fusion and canalization with the contralateral side.[3,4] This results in a duplicated uterus (usually didelphys) with one obstructed hemivagina. The absence of the Wolffian duct underlies the consistent ipsilateral occurrence of the renal anomaly in relation to the vaginal obstruction.[3,4]

No classification exists solely for OHVIRA, but most cases fit into general Müllerian anomaly systems. In the ASRM (AFS) classification, OHVIRA corresponds to Class III (uterus didelphys) with an obstructing longitudinal vaginal septum.[17] In the ESHRE/ESGE classification, it is typically described as U3bC2V2, corresponding to a didelphys bicorporeal uterus with double cervix and longitudinal obstructing vaginal septum.[17] Some authors also distinguish between complete and partial obstruction types.[9]

Our patient presented with post menarcheal cyclical lower abdominal pain, with dysmenorrhea. OHVIRA should be suspected in such girls with cyclical pelvic pain, a pelvic mass, or unusual discharge, especially if imaging shows a single kidney, which was

evident in the imaging modalities performed by us.[7,8] Ultrasound is usually the first imaging modality and can identify uterine duplication, hematocolpos, and renal agenesis.[9] However, MRI is the gold standard, providing detailed soft tissue contrast and clear delineation of uterine anatomy, septal location, and associated anomalies.[1,10] Diagnostic laparoscopy or vaginoscopy may be done when imaging is inconclusive or as part of surgical management.[13]

The mainstay of treatment is surgical removal or incision of the obstructing septum to restore normal menstrual outflow and prevent complications.[1,11] This is usually performed transvaginally; in adolescents or those with an intact hymen, hysteroscopic or vaginoscopic approaches are preferred to preserve anatomy.[15] Laparoscopic assistance may be required in complex cases.[11] Rarely, hemi-hysterectomy is performed if the obstructed horn is non-communicating and inaccessible. When treated early, OHVIRA has an excellent prognosis, with relief of symptoms and preservation of fertility.[5,12] However, delayed diagnosis may lead to complications such as endometriosis, adhesions, and reduced fertility.[7,12] Pregnancy is possible, but patients remain at increased risk for miscarriage, malpresentation, and preterm delivery.[7,18] Careful antenatal monitoring is recommended for women with a history of OHVIRA.[18]

Conclusion:

In summary, this case reveals the importance of considering OHVIRA syndrome in adolescent girls presenting with post-menarcheal cyclical pelvic pain and menstrual irregularities. A high index of suspicion, supported by detailed imaging, particularly MRI and prompt surgical management can result in excellent outcomes, as seen in our patient.

Acknowledgement:

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Conflict of interest

We do not have any conflict of interest.

Ethical approval

Ethical approval was not needed because this is a case report. However, informed written consent was obtained from the patient for preparation of this manuscript.

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