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Case Series

OHVIRA syndrome and its incomplete variant: multimodality imaging insights from two distinct presentations of Müllerian-renal anomalies

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ABSTRACT

Mullerian anomalies such as obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome pose diagnostic challenges due to their embryological variability and diverse clinical presentations. We present two adult cases highlighting the spectrum of OHVIRA. The first case involved a 27-year-old female with primary infertility, where imaging revealed classical OHVIRA features including uterine didelphys, obstructed hemivagina with hematocolpos, and ipsilateral renal agenesis. The second case was a 19-year-old female with vague abdominal pain and no significant menstrual complaints, in whom magnetic resonance imaging (MRI) demonstrated uterine didelphys, a single cervix, and ipsilateral renal agenesis without obstruction an incomplete variant of OHVIRA. MRI was pivotal in both cases for accurate diagnosis and surgical decision-making. Early recognition of both complete and incomplete OHVIRA variants is essential to prevent complications such as infertility, endometriosis, and chronic pelvic pain. MRI remains the gold standard for anatomical evaluation, enabling timely diagnosis and optimal clinical management.

Keywords: OHVIRA, Müllerian anomalies, Hemivagina, Renal agenesis, MRI, Vaginal obstruction, Uterine didelphys

INTRODUCTION

Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome, also known as Herlyn-Werner-Wunderlich syndrome, is a rare congenital anomaly arising from developmental failure of the Müllerian and mesonephric ducts. It is typically characterized by a triad of uterine didelphys, hemivaginal obstruction, and ipsilateral renal agenesis.[1] The exact incidence remains uncertain due to underdiagnosis, but it is most often recognized during adolescence, following menarche, when patients present with progressive pelvic pain, dysmenorrhea, or pelvic masses secondary to obstructed menstrual outflow.[2]

Clinically, OHVIRA poses a significant diagnostic challenge. Its presentation may mimic other gynecological or urological disorders, often leading to delays in appropriate management.[3] Furthermore, the anomaly may not be fully appreciated on initial pelvic ultrasound due to overlapping anatomical structures or incomplete visualization. Misdiagnosis or late diagnosis can result in serious complications such as endometriosis, pelvic adhesions, infertility, and recurrent infections.[4]

Radiologic imaging plays a pivotal role in the early and accurate identification of OHVIRA. While ultrasound may suggest the diagnosis, magnetic resonance imaging (MRI) is the modality of choice, offering superior soft-tissue resolution and multiplanar capability to delineate the complex uterovaginal anatomy and identify associated renal anomalies.[1,5] A timely and confident diagnosis not only facilitates appropriate surgical planning but also significantly reduces the risk of long-term morbidity.

In this case series, we present two adult females one with classical OHVIRA and another with an incomplete variant demonstrating the clinical and radiologic spectrum of Müllerian-renal anomalies. Through this report, we aim to highlight the critical role of MRI in diagnosis and surgical

planning, especially in distinguishing classical from variant forms.

CASE SERIES

Case 1: Classical OHVIRA syndrome in a 27-year-old female

Presenting complaint

A 27-year-old nulligravida woman presented to the gynecology outpatient department with a primary complaint of infertility for two years. She also reported mild lower abdominal discomfort and occasional dysmenorrhea, particularly during the early days of menstruation. Her menstrual cycles were regular, with no history of menorrhagia or intermenstrual bleeding. There was no prior pelvic surgery, and general and gynecological examinations were unremarkable.

Clinical background

The patient's infertility workup had been unremarkable until pelvic ultrasound raised suspicion of a Müllerian anomaly. There were no signs of pelvic inflammatory disease or endometriosis. Hormonal profile, thyroid function, and ovarian reserve markers were within normal limits.

Imaging findings

Pelvic ultrasound revealed a bicornuate uterus with two symmetric uterine horns, each containing a normal endometrial stripe. A 38×28×29 mm fluid collection with low-level internal echoes was noted along the left lateral vaginal wall, suggestive of hematocolpos. The right hemivagina was patent. Both ovaries were visualized and appeared morphologically normal.

Renal ultrasound revealed absence of the left kidney. The right kidney was ectopically located in the left renal fossa, with normal echotexture.

Hysterosalpingography (HSG) demonstrated opacification of a single right-sided endocervical canal and uterine horn, with normal contour and tubal spillage. The left horn and fallopian tube were not visualized, suggestive of a non-communicating uterine horn.

MRI pelvis revealed: uterine didelphys with two widely spaced uterine bodies and a deep fundal cleft, two distinct endocervical canals, normal right hemivagina, a well-defined mixed signal intensity collection (38 × 28 × 29 mm) in the left hemivaginal region, hyperintense on both T1- and T2-weighted images with internal blooming artifacts—consistent with hematocolpos, non-visualization of the left hemivagina, suggesting segmental vaginal aplasia, and absence of the left kidney and an ectopic right kidney located in the left renal fossa.

Diagnosis and outcome

The collective imaging findings confirmed the diagnosis of classical OHVIRA syndrome. The patient underwent transvaginal excision of the obstructing vaginal septum with drainage of hematocolpos. The procedure was uneventful. On follow-up, the patient reported complete resolution of pelvic discomfort and improved menstrual flow. She is currently undergoing fertility counseling and evaluation for assisted reproductive techniques.

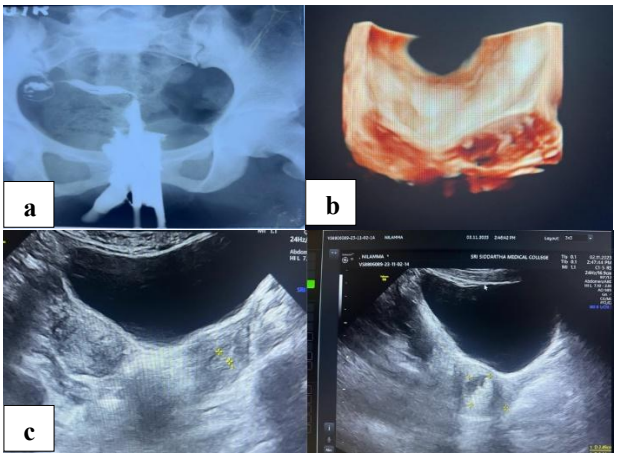

Figure 1: (a) HSG showing single right uterine horn, left uterine horn couldn't be visualized, (b) 3D reconstruction USG image showing vaginal collection with two uterine horns, and (c) USG showing two uterine horns with normal endometrial cavity with vaginal collection.

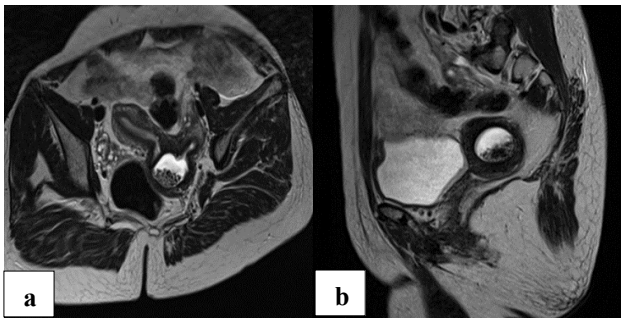

Figure 2: (a) Axial T2 MRI showing uterine didelphys with a deep fundal cleft and hematocolps, and (b) sagittal T2 MRI demonstrating hyperintense hematocolpos in the left hemivagina.

Case 2: Incomplete OHVIRA variant in a 19-year-old female

Presenting complaint

A 19-year-old woman presented with vague lower abdominal discomfort. She denied dysmenorrhea, menstrual irregularities, or urinary symptoms. There was no history of prior surgery or known congenital anomalies.

The initial clinical impression was nonspecific, and imaging revealed an incidental renal anomaly.

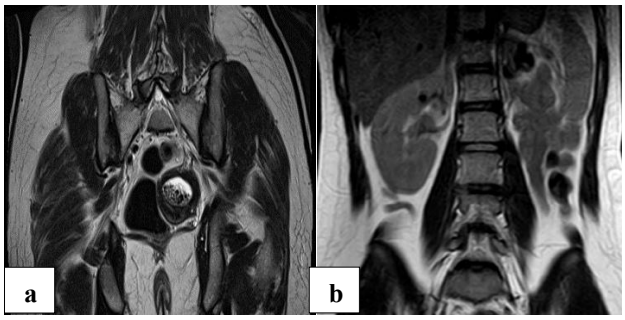

Figure 3 (a and b): Coronal T1 MRI showing absent left kidney and left hematocolps.

Clinical background

Ultrasound performed to evaluate abdominal pain revealed right renal agenesis. Given the embryological association between renal and Müllerian anomalies, further imaging with pelvic ultrasound and MRI was undertaken to assess reproductive anatomy.

Imaging findings

Ultrasound findings included: two separate endometrial echoes suggestive of a bicorporeal uterus, normal-appearing ovaries, and absence of the right kidney.

MRI pelvis confirmed: uterine didelphys with two well-formed uterine horns: right (6.0×3.0×2.9 cm) and left (6.7×3.7×3.5 cm), each horn had a separate endometrial cavity with appropriate endometrial thickness for the menstrual phase, presence of a single cervix, indicating partial fusion, no obstructing vaginal septum or hematocolpos, normal vagina and ovaries, and absence of the right kidney; the left kidney was normal in position and appearance.

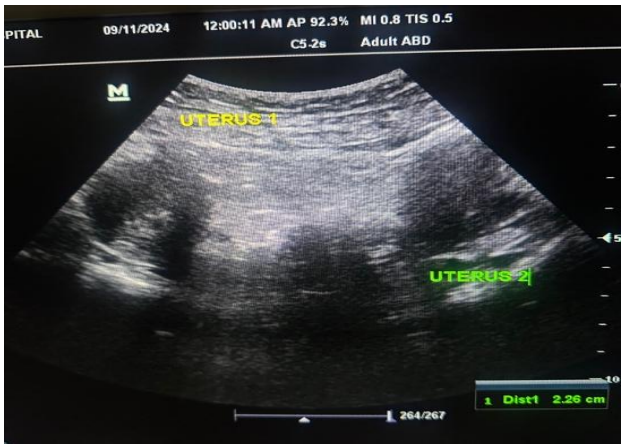

Figure 4: USG showing two distant uterine horns with normal endometrial cavity.

Diagnosis and outcome

A diagnosis of an incomplete variant of OHVIRA syndrome was established, characterized by uterine didelphys, single cervix, and ipsilateral (right) renal agenesis without vaginal obstruction. As there were no obstructive symptoms or functional impairment, no surgical intervention was required. The patient was counseled about the benign nature of the anomaly and advised regular follow-up, especially in the context of future fertility planning.

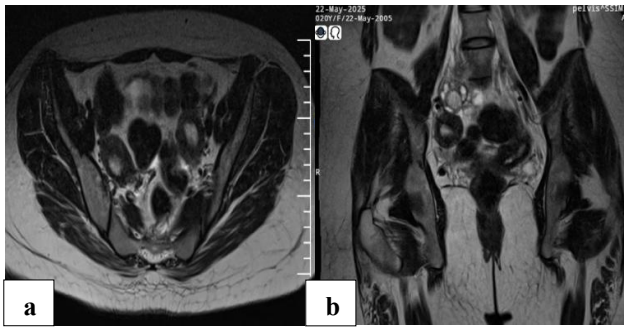

Figure 5: (a) Axial T2-weighted MRI showing two separate uterine horns consistent with uterine didelphys. Endometrial cavities are symmetric and appropriately distended for menstrual phase; and (b) coronal T2 – weighted MRI showing uterine didelphus and vaginal canal without evidence of hematocolpos or obstructive anomalies.

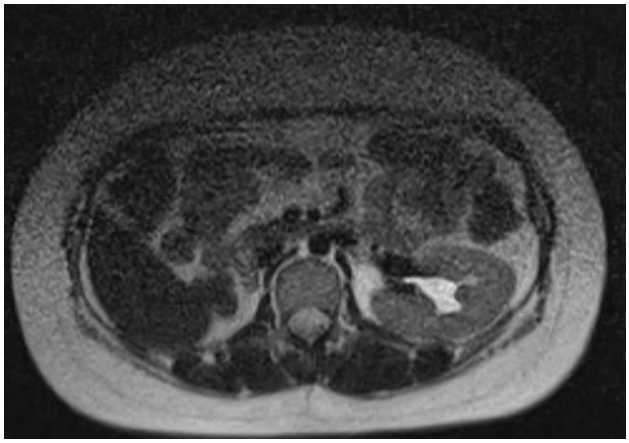

Figure 6: Axial MRI showing absent right kidney and a normally located left kidney with preserved corticomedullary differentiation and mild hydronephrosis.

DISCUSSION

In the present report, we describe two patients with Müllerian–renal anomalies within the OHVIRA spectrum one with the classical form and another with an incomplete, non-obstructive variant. The first patient presented with infertility and mild dysmenorrhea, and MRI revealed the complete triad of uterine didelphys,

obstructed hemivagina with hematocolpos, and ipsilateral renal agenesis. The second patient was asymptomatic apart from vague abdominal discomfort, and MRI demonstrated uterine didelphys, single cervix, and ipsilateral renal agenesis without vaginal obstruction.

These findings highlight that OHVIRA is not a uniform anomaly but can present with varying clinical severity. Prior studies, including those by Orazi et al and Fedele et al, note that the classical form is usually detected soon after menarche due to obstructive symptoms.[1,5] However, our first patient illustrates that diagnosis can be delayed into adulthood when symptoms are mild, which may adversely affect fertility. The incomplete variant in the second patient supports observations by Zhu et al and Smith et al, who described rare non-obstructive forms that are often detected incidentally.[2,3]

MRI played a pivotal role in both cases, confirming uterovaginal anatomy, identifying renal anomalies, and guiding management. This aligns with previous literature by Lakhotia et al and Orazi et al, which emphasize MRI's superiority over ultrasound for comprehensive anatomical evaluation.[1,4] In the classical form, MRI demonstrated a hematocolpos as a hyperintense T1/T2 collection with internal blooming, typical of chronic blood products. In the incomplete form, MRI excluded obstruction, preventing unnecessary surgical intervention.

Early recognition of obstructive OHVIRA is essential to prevent retrograde menstruation, endometriosis, and infertility. Gholoum et al reported that delayed diagnosis increases the risk of endometriosis a concern in our first patient.[9] Non-obstructive variants, such as in the second patient, generally require counseling and fertility monitoring rather than immediate surgery, consistent with recommendations by Smith et al.[3]

The contrast between these two presentations reinforces that OHVIRA encompasses a spectrum of Müllerian–renal anomalies. Maintaining a high index of suspicion, particularly in females with renal agenesis, and using MRI as the primary diagnostic tool are critical for timely, accurate diagnosis and individualized, fertility-preserving management.

Embryological basis and variants

During embryogenesis, the paired Müllerian ducts fuse to form the uterus, cervix, and upper vagina, while the mesonephric ducts contribute to ipsilateral kidney formation and influence canalization of the genital tract. Disruption in mesonephric duct formation may lead to renal agenesis and interfere with lateral fusion or vertical canalization of the ipsilateral Müllerian duct.

In the classical form, this manifests as a didelphic uterus, hemivaginal obstruction, and ipsilateral renal agenesis, as demonstrated in our first case. Incomplete or atypical variants, like the second case, may present with uterine duplication and renal agenesis but lack vaginal obstruction, occasionally featuring a single cervix. These patterns suggest that OHVIRA represents a spectrum of anomalies rather than a rigid triad. Non-didelphic variants, including bicornuate or septate uteri, have also been described in literature.

Imaging and diagnostic utility

Diagnosing OHVIRA particularly its incomplete forms can be challenging due to non-specific or absent symptoms. The second case exemplifies how such variants may be discovered incidentally.

Ultrasound remains the first-line modality, useful for identifying uterine configuration and associated renal anomalies. However, its limited ability to visualize complex pelvic anatomy necessitates further evaluation. MRI, with superior soft-tissue contrast and multiplanar capability, remains the modality of choice for characterizing uterovaginal anomalies and renal morphology.

In our first case, MRI demonstrated the classic triad and supported preoperative planning. In the second case, MRI confirmed an incomplete variant and helped avoid unnecessary surgical intervention. Thus, MRI is essential not only for diagnosis but also for guiding management across the OHVIRA spectrum.

Differential diagnosis

Several congenital anomalies may mimic OHVIRA on imaging.

Transverse vaginal septum may present with hematocolpos but usually involves a single uterus and cervix and lacks renal anomalies.

Imperforate hymen presents with cyclical pain and hematocolpos but without uterine duplication or renal agenesis; diagnosis is often made on clinical examination.

Unicornuate uterus with a non-communicating horn may mimic obstructive Müllerian anomalies and is often associated with renal agenesis. MRI typically shows a single uterus with a rudimentary horn, differentiating it from OHVIRA.

Accurate differentiation is critical, as each entity requires specific management.

Comparison with literature

Consistent with previous literature, our first case illustrates delayed diagnosis in adulthood, often due to infertility or vague pelvic symptoms. The second case highlights the under-recognized spectrum of OHVIRA variants and supports the evolving classification that includes incomplete forms.

Authors such as Zhu et al and Orazi et al have emphasized the role of MRI in classification, diagnosis, and pre-surgical mapping.[1,2] Increasing recognition of these incomplete forms has led to calls for a broader and more inclusive diagnostic definition of OHVIRA.

Clinical and surgical relevance

Early diagnosis of OHVIRA is critical to prevent complications such as retrograde menstruation, endometriosis, infertility, and chronic pelvic pain. In obstructive variants, timely surgical correction, such as septal excision, restores normal outflow and optimizes reproductive outcomes. In non-obstructive forms, conservative management and routine gynecologic surveillance are generally sufficient.

Radiologists play a central role in diagnosing both typical and atypical presentations. A thorough understanding of the embryological basis and radiologic spectrum is essential to guide treatment and avoid unnecessary interventions.

CONCLUSION

This report highlights two distinct presentations of OHVIRA syndrome one classical and one incomplete variant underscoring the diagnostic spectrum of this rare entity. While the triad of uterine didelphys, obstructed hemivagina, and ipsilateral renal agenesis defines the classical form, variants with a single cervix or no obstruction must also be recognized.

MRI remains invaluable for accurate diagnosis, characterization, and preoperative planning. Awareness of these variants is crucial among radiologists and gynecologists to ensure timely, accurate diagnosis and individualized, fertility-preserving care.

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