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A rare case of obstructed hemivagina with uterus didelphys and ipsilateral renal anomaly syndrome

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Abstract

Obstructed hemivagina and ipsilateral renal anomaly syndrome, also known as Herlyn–Werner–Wunderlich syndrome, represents a rare congenital anomaly characterized by the presence of an obstructed hemivagina with uterus didelphys and concomitant ipsilateral renal abnormalities. Typically, the clinical presentation includes cyclical abdominal pain, vaginal discharge, and/or a vaginal mass occurring post-menarche. Accurate diagnosis requires a high index of suspicion among clinicians, coupled with a comprehensive understanding of the distinctive features associated with this anomaly. Herein, we present the case of a 13-year-old female patient who complained of persistent lower abdominal pain. Magnetic resonance imaging confirmed the diagnosis of obstructed hemivagina and ipsilateral renal anomaly syndrome, and the patient was successfully treated with a laparotomy involving blood aspiration and hysterectomy. This case report highlights the significance of clinical awareness, prompt diagnosis, and timely therapeutic interventions to mitigate the adverse effects and optimize outcomes in individuals affected by obstructed hemivagina and ipsilateral renal anomaly syndrome.

Keywords

Congenital abnormalities, Müllerian anomalies, uterine anomaly, renal agenesis

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Introduction

Obstructed hemivagina with uterus didelphys and an ipsilateral renal anomaly (OHVIRA) syndrome represents a rare congenital malformation. The initial recognition of the association between renal agenesis and ipsilateral blind hemivagina was attributed to Herlyn and Werner in 1971, leading to its designation as Herlyn–Werner syndrome. Subsequently, in 1976, Wunderlich expanded the spectrum of associated malformations by incorporating uterine anomalies, resulting in the term Herlyn–Werner–Wunderlich Syndrome (HWW).[1] The reported incidence of OHVIRA is approximately 1 in 20,000 cases.[2] Typically, its clinical presentation includes cyclical abdominal pain, vaginal discharge, and/or the presence of a vaginal mass following menarche onset. In the majority of cases involving Müllerian duct anomalies, the initial clinical diagnosis tends to be inaccurate owing to the rarity of this anomaly and the presence of misleading signs and symptoms during presentation. The accurate diagnosis of Müllerian duct anomalies is crucial due to the increased incidence of associated conditions such as endometriosis, menstrual irregularities, infertility, and obstetric complications.[3]

Herein, we describe the case of a 13-year-old female with OHVIRA syndrome successfully treated with a laparotomy.

Case presentation

A 13-year-old female patient presented with recurrent lower abdominal pain that evolved into persistent discomfort radiating to the right thigh over a 9-month duration, accompanied by nausea and vomiting. Menarche occurred 2 years prior, and the patient reported irregular menstrual cycles, occurring every 2–3 months with a duration of 3–5 days. Physical examination revealed tenderness in the lower

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abdomen and the identification of a palpable pelvic mass on the right side, while the external genitalia appeared normal. The abdominal and pelvic ultrasound revealed right renal agenesis and a left-sided uterus measuring $3 \times 2.5 \times 5$ cm. The uterus exhibited a normal three-line endometrium and cervix, alongside bilateral ovaries showing multiple small follicle cystic lesions. In addition, an oval-shaped mass measuring $10 \times 7.5 \times 5$ cm was identified in the right intraabdominal region. The mass had a 1 cm thick wall containing material resembling ground glass. The decision to omit trans-vaginal ultrasound was made due to cultural considerations and the need to preserve the hymen.

Subsequently, a computerized tomography (CT) scan was done and revealed right kidney agenesis and a compensatory enlarged left kidney measuring about 13 cm (Figure 1). The imaging also indicated features suggestive of uterus didelphys coupled with unilateral (right) obstructed hemivagina and hematometrocolpos, prompting suspicion of OHVIRA syndrome. Magnetic resonance imaging (MRI) was subsequently employed to confirm the diagnosis (Figure 2).

Under general anesthesia, a perineal examination, conducted gently with the aid of two small sinus retractors, unveiled a high, thick transverse septum located just below the cervix. A low transverse abdominal incision was made, which identified the 10×7 cm right enlarged uterus filled with chocolate-like material. Aspiration of the blood was performed, followed by a hysterectomy, while the vaginal septum was intentionally preserved in situ. Finally, closure of the vaginal pouch was accomplished by suturing with the peritoneum to prevent herniation. On the third postoperative day, the patient was discharged in a healthy, pain-free condition. At the 1-month follow-up in the outpatient clinic, the patient remained asymptomatic, reporting no pain or concerns, and the incision site exhibited satisfactory healing.

Discussion

Müllerian anomalies encompass congenital alterations affecting the female reproductive tract and are frequently associated with anomalies in the renal, ureteral, and bladder systems due to their shared embryological origin.[4] OHVIRA syndrome constitutes 0.16%–10% of all Müllerian anomalies.[5]

While the precise pathogenesis of OHVIRA syndrome remains unknown, several theories have been proposed, with Acien's theory being the most widely accepted. According to Acien's theory, a primary anomaly in the Wolffian duct is the fundamental factor contributing to OHVIRA syndrome.[6] An anomaly arising from the Wolffian duct origins can result in obstructed hemivaginas. In addition, when the Wolffian duct fails to induce metanephric blastema and subsequent growth factor release crucial for the fusion of paramesonephric (Müllerian) ducts, it leads to renal agenesis and uterus didelphys.[6]

OHVIRA syndrome typically manifests during puberty, following the onset of menarche, presenting with cyclical

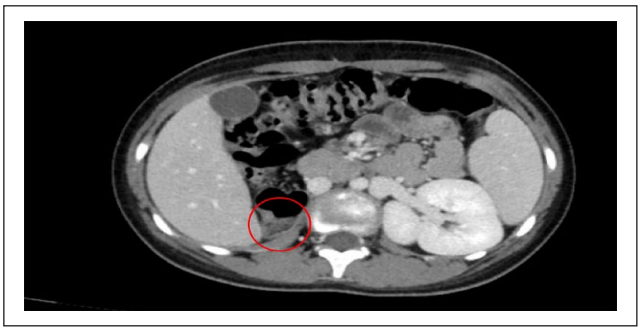

Figure 1. Computerized tomography scan of the abdomen and pelvis displaying the absence of the right kidney (red circle).

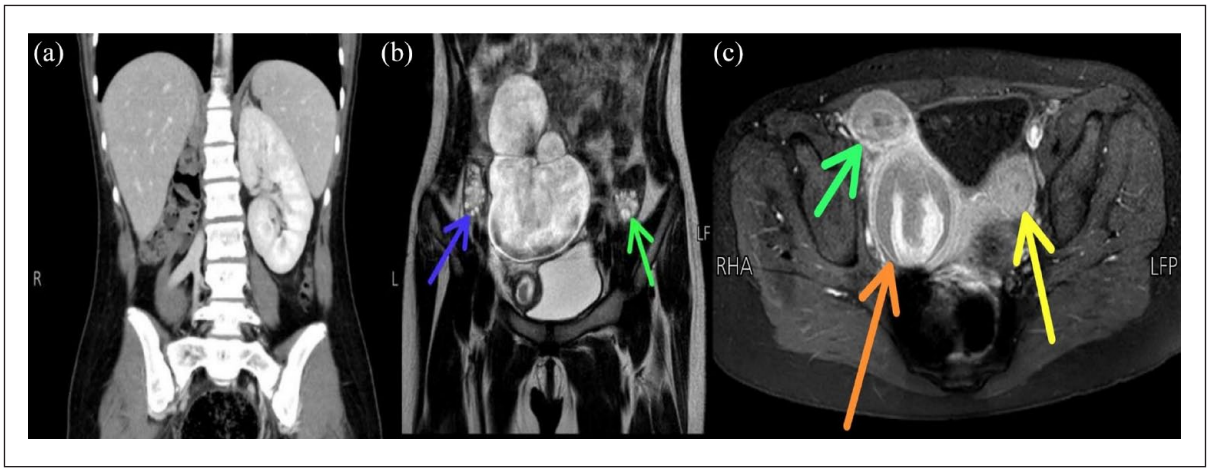

Figure 2. (a) Coronal IV contrast-enhanced image of the abdomen demonstrating the absence of the right kidney. (b) Selected T2 coronal images of the pelvis illustrating the presence of both ovaries: the right ovary (blue arrow) and the left ovary (green arrow). (c) Selected axial T1 fat-saturated magnetic resonance imaging with gadolinium highlighting two separate uterine bodies. The right uterus (green arrow) is associated with a bloody collection (hematometrocolpos—orange arrow), while the left uterine body is indicated by the yellow arrow.

abdominal pain or dysmenorrhea within a few months to a year thereafter.[7] Our patient experienced similar symptoms 1 year after menarche, characterized by severe lower abdominal pain radiating to the right thigh, accompanied by nausea and vomiting. In addition, individuals with OHVIRA syndrome may exhibit symptoms such as vaginal discharge, infertility, urinary retention, or pelvic inflammatory diseases, including tubo-ovarian abscess.[8] Notably, OHVIRA syndrome may manifest early in neonates, characterized by an orthotopic urethral meatus, a vaginal introitus located on the lateral aspect, and the presence of an interlabial mass,[9] and/or a dysplastic kidney.[10] Alternatively, presentation in the infantile period may involve an ectopic ureter and atrophic dysplastic kidney.[11] Rarely, the syndrome has been reported in adult women with a history of multiparity and recurrent urinary tract infections.[12]

The diagnosis of OHVIRA syndrome poses challenges due to its diverse clinical presentations; however, it can be achieved through a comprehensive medical history, thorough physical examination, and imaging modalities such as ultrasonography and/or MRI. Reports suggest that MRI is the preferred imaging technique for accurate diagnosis of OHVIRA syndrome.[1,13] It is important to note that the lack of visualization of a kidney on MRI does not conclusively negate the possibility of a small and/or atrophic kidney.[14] Although ultrasound is commonly used as the initial imaging modality, offering accessibility and prompt results, it may not visualize the vaginal septum. Nevertheless, it can detect renal anomalies, uterine didelphys, and hematometrocolpos.[15] In our case, a CT scan was initially performed before suspecting OHVIRA syndrome. It is worth noting that CT scans are not recommended for diagnosing this syndrome due to their lower sensitivity in visualizing uterine contours, the shape of the uterine cavity, or the location of the vaginal septum.

Surgical intervention is the primary approach for treating OHVIRA syndrome, including methods such as laparotomy, laparoscopy, or minimally invasive procedures. Among these, laparoscopy is the gold standard of treatment.[16] In our case, a laparotomy was performed due to constraints related to equipment availability. However, certain reports advocate for considering minimally invasive procedures as the initial choice over laparoscopy, emphasizing their affordability and safety, particularly concerning fertility outcomes.[13,16]

Gynecologists and pediatric surgeons should include OHVIRA syndrome in the differential diagnosis for individuals with kidney anomalies and/or acute abdominal symptoms during adolescence. Maintaining a high level of suspicion is crucial to prevent potential complications such as pelvic inflammatory disease, endometriosis, pyometra, abscess formation, and infertility.[3]

Conclusion

HWW represents a rare congenital malformation of the female urogenital tract. Due to the close developmental connection between the urinary and genital systems, it is important to routinely assess for renal abnormalities in patients with OHVIRA syndrome. This case underscores the significance of maintaining a high index of suspicion and interdisciplinary collaboration among pediatric surgeons, radiologists, and gynecologists for accurate and timely diagnosis. Accurate diagnosis and prompt intervention are essential to prevent potential complications.

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Author contributions

T.M.A., A.T.I., and N.A.A. contributed to data interpretation and drafting of the manuscript. E.M.A and L.M.H contributed to study design, data collection, and data interpretation. M.A contributed to the drafting of the manuscript and supervision of the work. All authors have read and approved the final manuscript.

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Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed consent

Written informed consent was obtained from a legally authorized representative(s) for anonymized patient information to be published in this article.

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