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MRI Evaluation of OHVIRA Syndrome: A Case Report of a Rare Congenital Malformation

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Abstract **Case Report**

Obstructed Hemivagina and Ipsilateral Renal Anomaly (OHVIRA) syndrome is a rare congenital Müllerian duct anomaly defined by the triad of uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis. It typically presents shortly after menarche with cyclic pelvic pain and dysmenorrhea due to hematocolpos. We report an atypical case of an 18-year-old woman presenting with dyspareunia despite regular menstrual cycles. Pelvic ultrasound suggested a uterus didelphys associated with a cystic pelvic lesion. Magnetic resonance imaging (MRI) confirmed a uterus didelphys with a double vaginal cavity, including an obstructed hemivagina, and ipsilateral renal agenesis. The anomaly was classified as U3b C2 V3 according to the ESHRE system. This case highlights the pivotal role of MRI in diagnosing Müllerian anomalies and emphasizes that OHVIRA syndrome may present without classic obstructive symptoms.

Keywords: OHVIRA syndrome, Herlyn-Werner-Wunderlich syndrome, Uterus didelphys, Obstructed hemivagina, Renal agenesis, Magnetic resonance imaging.

INTRODUCTION

Müllerian duct anomalies (MDAs) arise from defects in the development, fusion, or resorption of the paramesonephric ducts during embryogenesis. Their prevalence is estimated at 5–7% in the general population and may reach up to 25% in women with reproductive disorders [1].

OHVIRA syndrome, also known as Herlyn–Werner–Wunderlich syndrome, is a rare entity characterized by the association of uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis [2].

This condition results from a combined anomaly of the Müllerian and mesonephric ducts, explaining the frequent coexistence of genital and urinary tract malformations [3].

Classically, OHVIRA syndrome presents shortly after menarche with cyclic pelvic pain, dysmenorrhea, and pelvic mass due to hematocolpos [4]. However, atypical or delayed presentations have been increasingly reported, including dyspareunia or nonspecific pelvic symptoms, which may obscure the diagnosis [5].

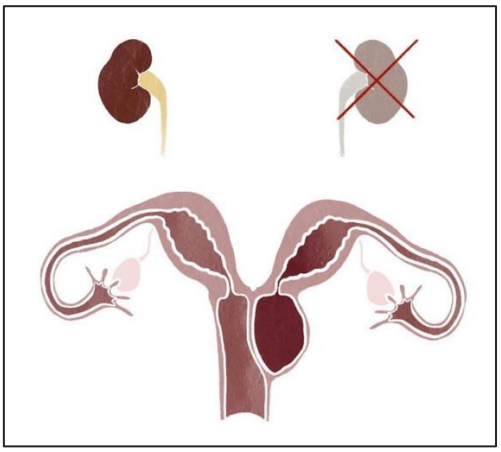

Figure 1: Illustration showing a hematocolpos in the left hemivagina, a vaginal septum, and ipsilateral renal agenesis.

Source: Quintero G., Rojas J.J., Quintero N. (2024). « Illustration showing hematocolpos in left hemivagina, vaginal septum, ipsilateral renal agenesis », dans: Herlyn-Werner Wünderlich OHVIRA syndrome… Ciencia Latina Revista Científica Multidisciplinaria, 8(3), 3727–3743. doi:10.37811/cl_rcm.v8i3.11586.

Magnetic resonance imaging (MRI) is considered the reference imaging modality for evaluating MDAs due to its high spatial resolution, excellent soft tissue contrast, and multiplanar capability, enabling precise anatomical characterization and classification [6].

Case Report

An 18-year-old married woman presented with progressive dyspareunia over several weeks. She reported regular menstrual cycles since menarche, without dysmenorrhea or chronic pelvic pain. No urinary or systemic symptoms were noted.

Gynecological examination revealed normal external genitalia. Speculum examination demonstrated a bulging mass along the lateral vaginal wall, with a slightly displaced but otherwise normal-appearing cervix. Bimanual examination confirmed a vaginal mass with mild tenderness.

Pelvic ultrasound suggested a uterus didelphys associated with a cystic pelvic lesion, raising suspicion of a Müllerian anomaly. A pelvic MRI was performed for further evaluation.

Pelvic MRI demonstrated a uterus didelphys characterized by a complete bicorporeal configuration with two well-defined and widely divergent hemi-uteri (intercornual distance = 9 cm) separated by a deep fundal cleft measuring 8 mm.

Figure 2: T2 coronal images showing two uterine cavities (arrows) and left vaginal cavity(asterisk)

Each hemi-uterus communicated with an independent cervical canal, both showing preserved physiological signal intensity and associated with mild endouterine fluid retention.

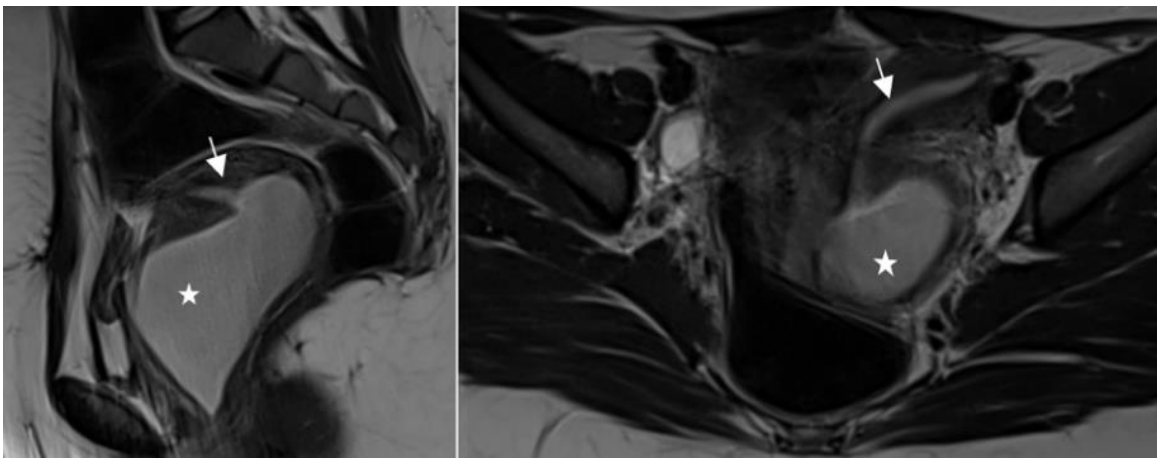

Figure 3: Sagittal and Axial T2-weighted images demonstrating the left uterine cavity (arrow) communicating with an obstructed ipsilateral hemivagina, associated with hematocolpos (asterisk)

At the vaginal level, a double vaginal cavity was identified: the left hemivagina was markedly distended and contained fluid with high signal intensity on T2-weighted images and intermediate signal on T1-weighted images, associated with thinning of the vaginal walls; this hemivagina was blind-ending due to a transverse obstructing septum, responsible for fluid retention.

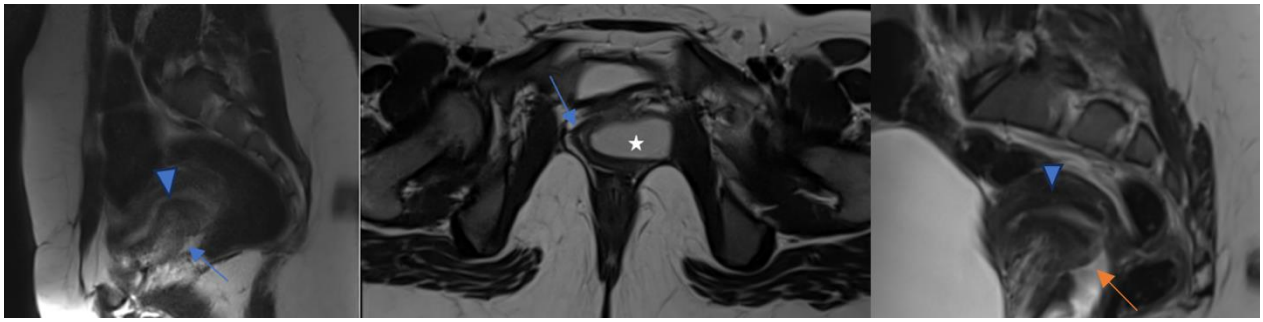

Figure 4: Sagittal and axial T2-weighted MR images demonstrate the right uterine cavity (arrowhead) communicating with an ipsilateral hemivagina, initially collapsed (blue arrow) and subsequently distended following endovaginal gel instillation. ***Additionally, the left hemivagina demonstrates distension with hematocolpos (asterisk)***

In contrast, the right hemivagina appeared collapsed, without detectable endoluminal fluid, and was patent, as confirmed after endovaginal gel instilliation.

Both ovaries were normal with a follicular pattern. The urinary bladder and the rectum showed normal morphology. No significant lymphadenopathy or peritoneal effusion was observed. Complementary imaging confirmed left renal agenesis.

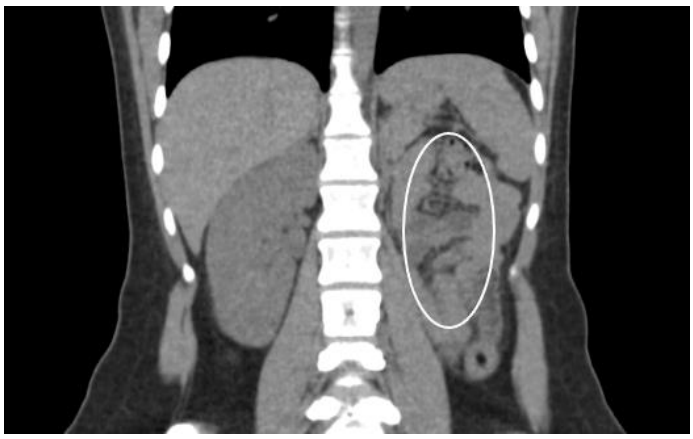

Figure 5: Coronal CT image demonstrating absence of visualization of the left kidney, ipsilateral to the obstructed hemivagina

Based on imaging findings, the diagnosis of OHVIRA syndrome was established. According to the ESHRE/ESGE classification, the anomaly was categorized as U3b C2 V3 [7].

DISCUSSION

OHVIRA syndrome is a rare congenital anomaly resulting from asymmetric development of the Müllerian ducts combined with mesonephric duct maldevelopment [3]. The presence of uterus didelphys reflects complete failure of Müllerian duct fusion, while the obstructed hemivagina results from a transverse vaginal septum, and the associated renal agenesis arises from ipsilateral mesonephric duct anomaly.

The classical clinical presentation includes cyclic pelvic pain and dysmenorrhea, typically occurring shortly after menarche due to progressive accumulation of menstrual blood in the obstructed hemivagina [4]. However, atypical presentations are increasingly recognized. Patients may present with dyspareunia, infertility, or minimal symptoms, particularly when partial drainage or delayed obstruction occurs [5]. In this case, the absence of dysmenorrhea contributed to delayed clinical suspicion.

Imaging plays a crucial role in diagnosis. Ultrasound is usually the first-line modality but may be limited in accurately defining complex anatomy. MRI provides near-complete diagnostic accuracy in identifying uterine morphology, vaginal septa, and associated renal anomalies [6]. It is also essential for classification according to systems such as ESHRE/ESGE, which facilitates standardized diagnosis and management [7].

The main differential diagnoses include other uterine duplication anomalies, such as septate uterus or bicorporeal uterus without vaginal obstruction, which lack the characteristic obstructed hemivagina and renal agenesis.

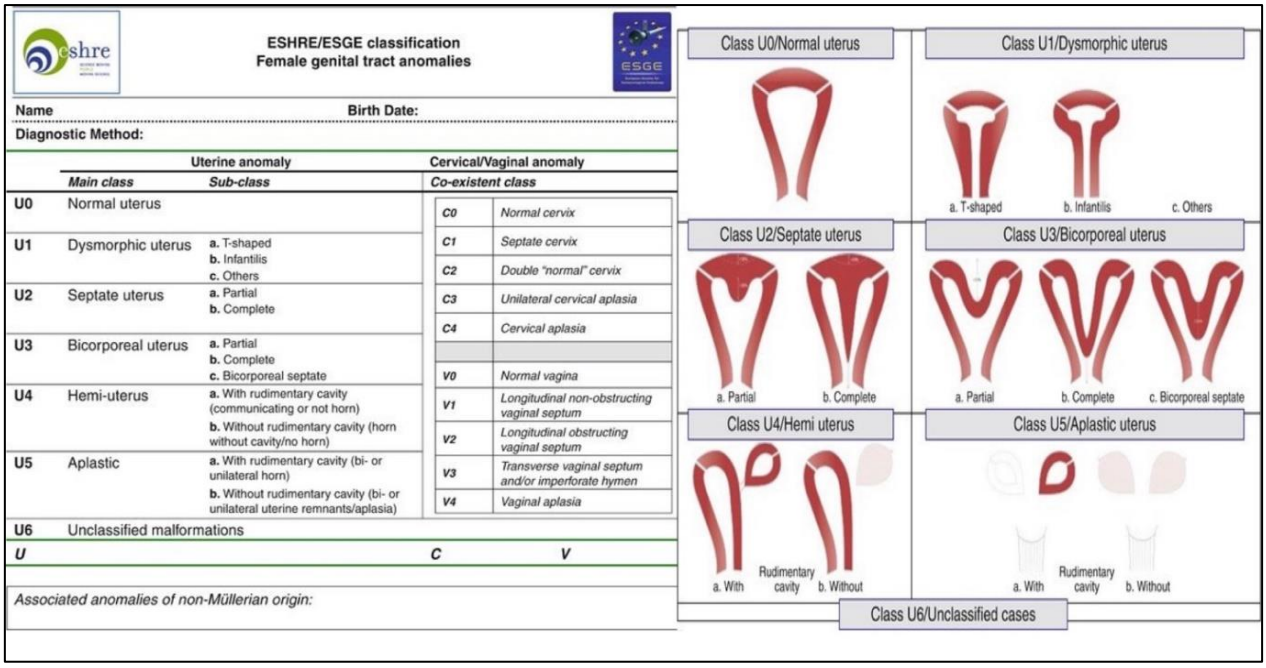

Figure 6: Classification of congenital anomalies of the female genital tract according to the ESHRE/ESGE consensus.
Source: Grimbizis G.F., Gordts S., Di Spiezio Sardo A., Brucker S.Y., De Angelis C., Gergolet M., Li T.C., Tanos V., Brölmann H., Gianaroli L., Campo R. (2019). « The ESHRE/ESGE consensus on the classification of female genital tract congenital anomalies », dans: ECR 2019 Poster C-1879: Educational exhibit, European Radiology. doi:10.26044/ecr2019/C-1879.

Early diagnosis is important to prevent complications such as endometriosis (reported in 23–38% of untreated cases) [8], chronic pelvic infection, infertility and obstetric complications.

The treatment of choice is surgical resection of the obstructing vaginal septum, which is minimally invasive and associated with excellent clinical and reproductive outcomes [9].

Conclusion

This case illustrates an atypical presentation of OHVIRA syndrome in a young woman presenting with dyspareunia and regular menstrual cycles. MRI was essential for establishing the diagnosis, accurately delineating the anatomy, and guiding management. Awareness of such non-classical presentations is crucial to avoid delayed diagnosis and prevent long-term complications.

References

1. Acien P. Incidence of Müllerian defects in fertile and infertile women. *Hum Reprod*. 1997;12(7):1372–1376.https://doi.org/10.1093/oxfordjournals.humrep.a019588
2. MR. Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA syndrome). J Pediatr Adolesc Gynecol. 2007;20(5):245–9.
3. Acien P. Embryological observations on the female genital tract. Hum Reprod. 1992;7(4):437–45.
4. Orazi C, Lucchetti MC, Schingo PM, Marchetti P, Ferro F. *Herlyn-Werner-Wunderlich syndrome*. Radiology. 2007;244(2):591–597.DOI: 10.1148/radiol.2442060425
5. Gholoum S, Puligandla PS, Hui T, Su W, Quiros E, Laberge JM. Management and outcome of patients with combined vaginal septum, bifid uterus, and ipsilateral renal agenesis (Herlyn–Werner–Wunderlich syndrome). *J Pediatr Surg*. 2006;41(5):987–992. doi: 10.1016/j.jpedsurg.2006.01.021.
6. Troiano RN, McCarthy SM. Müllerian duct anomalies: imaging and clinical issues. Radiology. 2004;233(1):19–34.
7. Grimbizis GF, Gordts S, Di Spiezio Sardo A, Brucker S, De Angelis C, Gergolet M, et al. The ESHRE/ESGE classification of female genital tract congenital anomalies. Human Reproduction. 2013;28(8):2032–2044. doi:10.1093/humrep/det098.
8. Rock JA, Jones HW. Te Linde's Operative Gynecology. 11th ed. Philadelphia: Wolters Kluwer; 2015.
9. *Quintero G., Rojas J.J., Quintero N. (2024). « Illustration showing hematocolpos in left* hemivagina, vaginal septum, ipsilateral renal agenesis », dans: Herlyn-Werner Wünderlich OHVIRA syndrome… Ciencia Latina Revista Científica Multidisciplinaria, 8(3), 3727–3743. doi :10.37811/cl_rcm. v8i3.11586.