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(CASE REPORT)

Atypical presentation of OHVIRA Syndrome in an adolescent with chronic pelvic pain: A case report

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Abstract

Introduction: Obstructed Hemivagina and Ipsilateral Renal Anomaly (OHVIRA) syndrome, also known as Herlyn–Werner–Wunderlich syndrome, is a rare congenital Müllerian duct anomaly. It typically presents after menarche with cyclic pelvic pain secondary to menstrual blood retention.

Case Presentation: We report the case of a 15-year-old adolescent girl presenting with chronic pelvic pain in the absence of menstrual or urinary symptoms. Imaging revealed **uterus didelphys** associated with an obstructed right hemivagina complicated by hematometra and hematocolpos, along with an ipsilateral ectopic pelvic kidney. Surgical excision of the vaginal septum with marsupialization was performed, resulting in complete resolution of symptoms.

Conclusion: OHVIRA syndrome should be considered in adolescents presenting with chronic pelvic pain, even in the absence of menstrual abnormalities. Magnetic resonance imaging (MRI) plays a pivotal role in diagnosis, and early surgical management provides symptom relief while preserving reproductive potential.

Keywords: OHVIRA Syndrome; Müllerian Anomaly; Hematocolpos; Adolescent

1. Introduction

Müllerian duct anomalies result from abnormal development, fusion, or resorption of the paramesonephric ducts during embryogenesis.

Obstructed Hemivagina and Ipsilateral Renal Anomaly (OHVIRA) syndrome is a rare congenital disorder characterized by uterine duplication, unilateral vaginal obstruction, and an ipsilateral renal malformation [1,2]. The syndrome results from a concomitant developmental defect of the Müllerian and mesonephric ducts.

OHVIRA syndrome typically presents after menarche with progressively worsening cyclic pelvic pain, dysmenorrhea, and occasionally a pelvic mass due to retained menstrual blood [3]. However, atypical presentations without overt menstrual abnormalities may occur, leading to delayed diagnosis and an increased risk of complications such as infection, endometriosis, and infertility [4,5].

We report an atypical presentation of OHVIRA syndrome in an adolescent girl with chronic pelvic pain but no menstrual abnormalities.

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2. Case Presentation

A 15-year-old nulliparous adolescent girl with no significant medical or surgical history was referred for evaluation of chronic pelvic pain that had been evolving for several months.

The pain was not associated with dysmenorrhea, menstrual irregularities, urinary symptoms, or gastrointestinal complaints.

On clinical examination, the patient was alert, hemodynamically stable, and in good general condition (**ECOG Performance Status 0**).

Abdominal examination revealed right-sided pelvic tenderness associated with asymmetry and a palpable right lateralized pelvic swelling.

A vaginal examination was not performed to preserve hymenal integrity. Rectal examination revealed a firm, bulging anterior pelvic mass.

2.1. Imaging Findings

2.1.1. Pelvic Ultrasound

Pelvic ultrasonography demonstrated a large heterogeneous hypoechoic oblong mass with a ground-glass appearance, measuring approximately 160 × 90 mm, with a thickened wall.

An adjacent rounded structure with similar echogenic characteristics was identified, suggesting blood retention involving the right uterine cavity and a blind-ending hemivagina.

These findings were suggestive of a uterine malformation, most likely **uterus didelphys with an obstructed right hemivagina**.

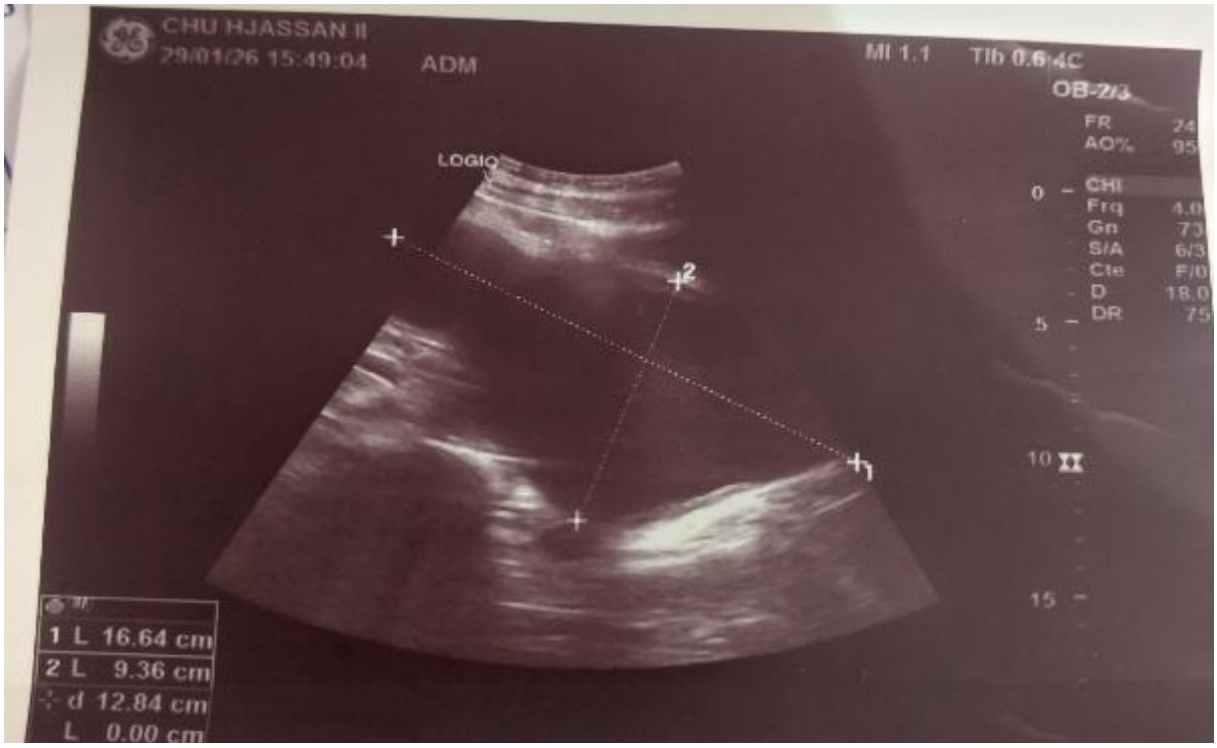

Figure 1 Heterogeneous hypoechoic pelvic mass with a ground-glass appearance (160 × 93 mm), suggestive of blood retention associated with uterus didelphys and an obstructed right hemivagina.

2.2. Magnetic Resonance Imaging

Pelvic MRI demonstrated two completely separate uterine cavities and cervices, consistent with **uterus didelphys**.

The right uterine cavity and cervix were markedly distended and filled with hemorrhagic content displaying high signal intensity on both T1- and T2-weighted images, without diffusion restriction or post-contrast enhancement.

This cavity was continuous with a blind-ending right hemivagina, consistent with right-sided hematometra and hematocolpos.

The left uterine cavity measured 28 × 51 mm and displayed preserved zonal anatomy with a 10-mm endometrium. It communicated with a normally developed cervix and vagina.

Both ovaries were normal in size and position, with a physiological follicular appearance.

MRI also revealed a right ectopic pelvic kidney measuring approximately 4 cm. Ultrasonography confirmed a hyperechoic kidney with preserved corticomedullary differentiation.

No intraperitoneal fluid collection or bladder or rectal abnormalities were identified.

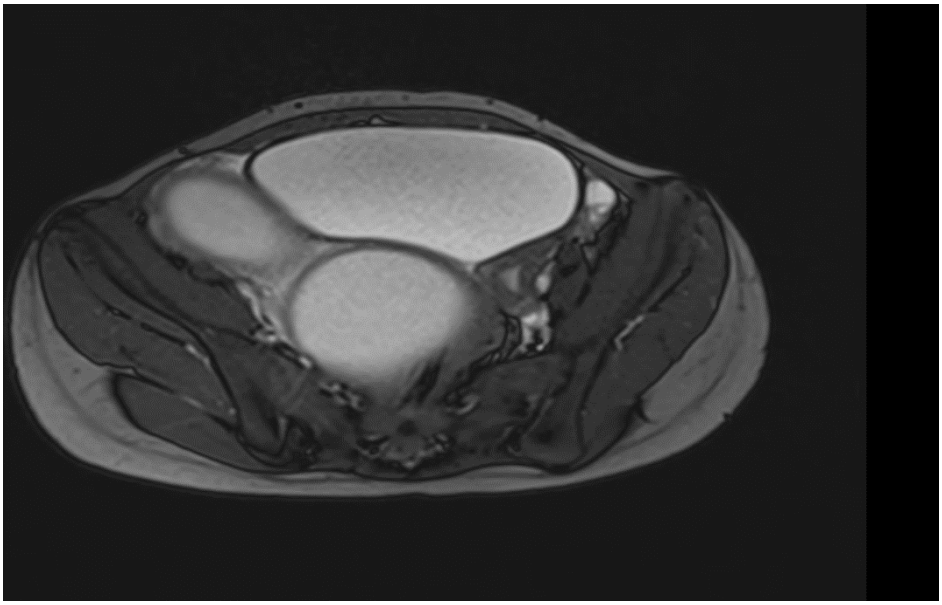

Figure 2 Axial pelvic MRI demonstrating uterus didelphys with marked distension of the right hemivagina and right hemiuterus secondary to an obstructed hemivagina.

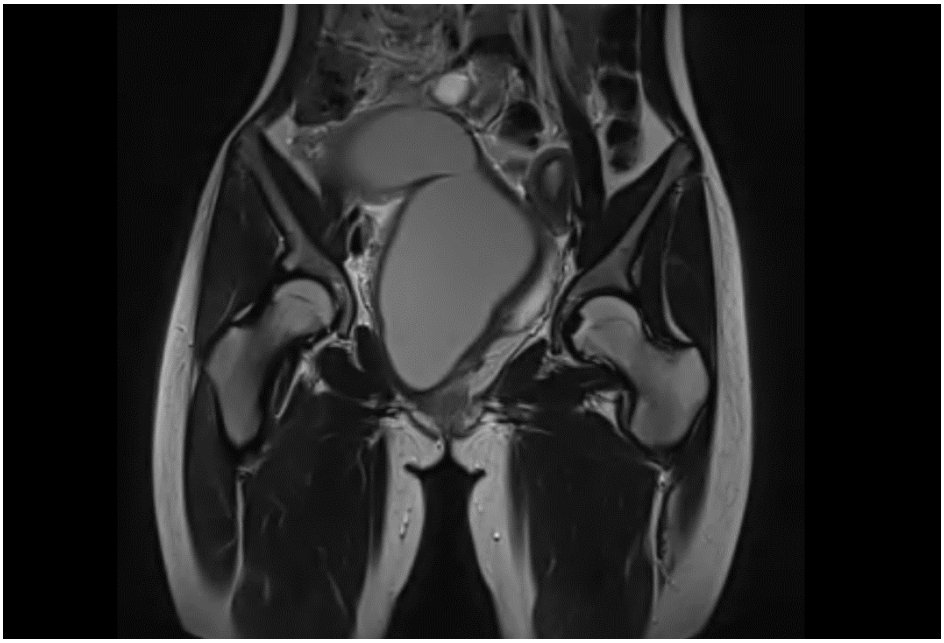

Figure 3 Coronal T2-weighted pelvic MRI showing uterus didelphys with marked distension of the right hemivagina and right hemiuterus due to obstruction, resulting in hematometra and hematocolpos (hyperintense fluid signal). The left hemivagina and left hemiuterus appear non-distended.

2.3. Diagnosis

Based on the clinical and radiological findings, a diagnosis of OHVIRA syndrome was established, consisting of **uterus didelphys**, an obstructed right hemivagina complicated by hematometra and hematocolpos, and a right ectopic pelvic kidney.

2.4. Management

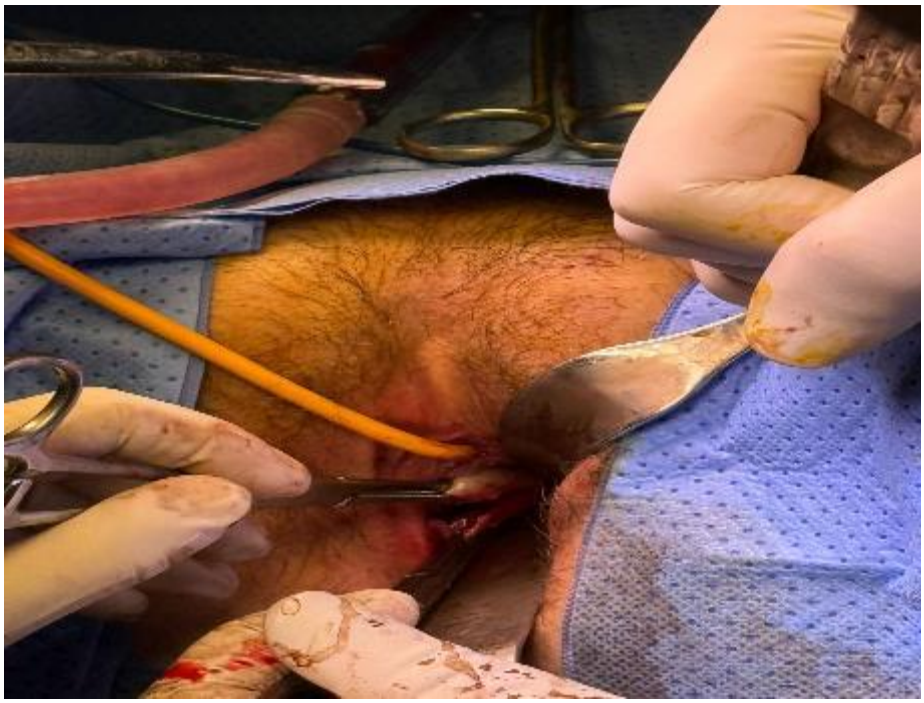

Figure 4 Incision of the bulging right vaginal septum allowing evacuation of hematocolpos and hematometra.

Following informed consent, surgical management was performed under locoregional anesthesia with sedation in the lithotomy position.

Vaginal examination revealed an intact annular hymen and a bulging vaginal mass corresponding to the hematocolpos.

The bulging area was incised using a cold scalpel, allowing drainage of thick chocolate-colored fluid.

The obstructing vaginal septum was widely excised, and marsupialization of the anterior and posterior vaginal walls was performed using absorbable sutures (Vicryl® 1).

Hemostasis was achieved, and a Foley catheter was inserted to maintain vaginal patency. Secondary hymenal tears related to the procedure were noted.

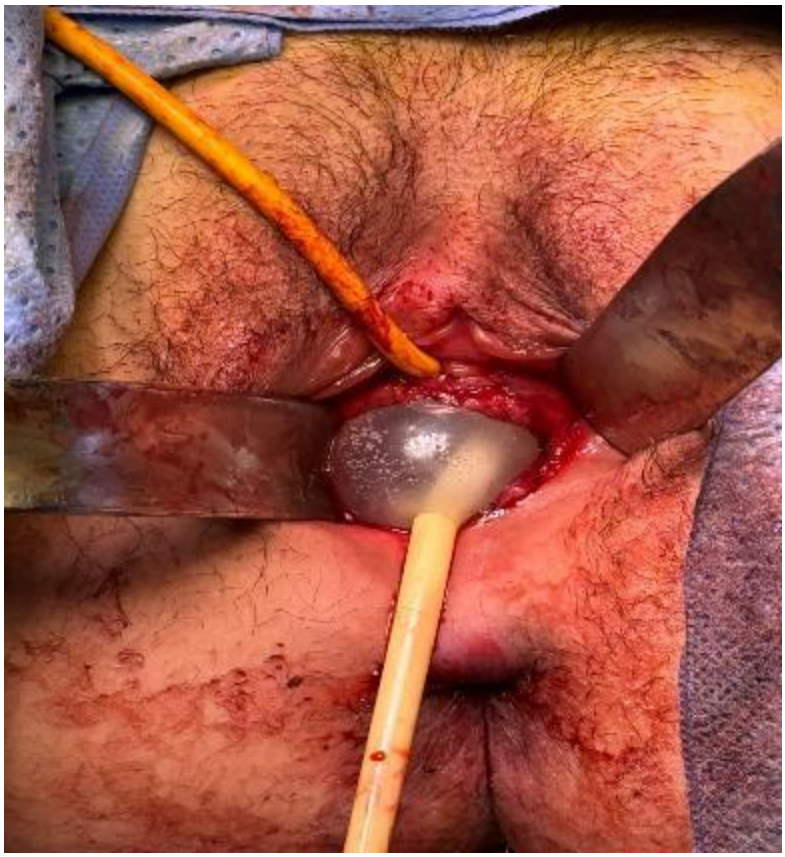

Figure 5 Placement of a Foley catheter to maintain vaginal patency.

2.5. Outcome and Follow-Up

The postoperative course was uneventful.

The patient experienced complete resolution of pelvic pain.

Follow-up was scheduled to monitor menstrual regularity, vaginal patency, and long-term gynecological and reproductive outcomes. The patient is currently being followed by the Nephrology Department for further evaluation of the associated renal anomaly.

3. Discussion

OHVIRA syndrome is a rare Müllerian–mesonephric anomaly resulting from abnormal embryological development of the Müllerian and Wolffian ducts [1,6].

Although classically associated with **uterus didelphys**, an obstructed hemivagina, and ipsilateral renal agenesis, anatomical variants such as an ectopic kidney have increasingly been reported [2,6].

Clinical presentation is frequently delayed because of atypical symptoms. While cyclic pelvic pain and dysmenorrhea are common, some patients experience apparently normal menstruation through the unobstructed hemivagina, thereby masking the underlying pathology [3–5].

Delayed diagnosis may result in complications including endometriosis, pelvic infection, adhesions, and infertility [7,8].

MRI remains the gold-standard imaging modality because it provides accurate anatomical characterization and facilitates detection of associated urinary tract anomalies [2,9].

Early diagnosis enables prompt surgical management, which is essential for symptom relief and fertility preservation.

3.1. Surgical Management

The treatment of choice for OHVIRA syndrome is transvaginal excision of the obstructing vaginal septum [7,8].

Simple incision and drainage are associated with a high recurrence rate due to restenosis. Wide excision combined with marsupialization is currently recommended because it ensures durable vaginal patency and reduces postoperative complications [5,8,10].

Gradual decompression of large hematocolpos collections is recommended to prevent retrograde reflux and vasovagal reactions [7].

3.2. Postoperative Management

Postoperative management includes temporary placement of a vaginal stent or Foley catheter to prevent adhesions, antibiotic prophylaxis when indicated, and long-term follow-up to assess menstrual function and reproductive outcomes [5,10].

A multidisciplinary follow-up approach is recommended because of the associated renal abnormalities [1].

3.3. Fertility and Obstetric Outcomes

Reproductive outcomes following surgical correction are generally favourable, with numerous reports of spontaneous pregnancies [8,11].

However, these pregnancies are associated with increased risks of preterm birth, malpresentation, and caesarean delivery [11].

Timely surgical intervention remains a key determinant of reproductive prognosis.

4. Conclusion

OHVIRA syndrome should be considered in any adolescent presenting with chronic pelvic pain, even in the absence of menstrual abnormalities.

MRI plays a central role in diagnosis.

Early surgical management consisting of vaginal septum excision and marsupialization provides excellent symptom relief while preserving reproductive potential.

Compliance with ethical standards

Disclosure of conflict of interest

The authors declare no conflicts of interest.

Statement of informed consent

Written informed consent was obtained from the patient for publication of this case report and the accompanying images.

Author Contributions

All authors contributed to patient management, manuscript preparation, and critical revision of the manuscript. All authors read and approved the final version.

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