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Case Report

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Obstructed Hemivagina and Ipsilateral Renal Anomaly Syndrome (Herlyn-Werner-Wunderlich Syndrome): A Case Report

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ABSTRACT

Herlyn-Werner-Wunderlich Syndrome (HWW), also known as Obstructed Hemivagina and Ipsilateral Renal Anomaly (OHVIRA) syndrome, is a rare Müllerian duct anomaly characterized by a triad of didelphic uterus, obstructed hemivagina, and ipsilateral renal agenesis. We report a case of a 15-year-old girl presenting with acute pelvic pain, cyclic dysmenorrhea, and a palpable abdominal mass. Imaging revealed a bicornuate uterus with a blind hemivagina, hematocolpos, and hematometra. Surgical management included resection of the vaginal septum and reconstruction, resulting in prompt recovery without complications.

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Introduction

Herlyn-Werner-Wunderlich Syndrome (HWW), a rare variant of Müllerian duct anomalies, manifests as a triad of didelphic uterus, obstructed hemivagina, and ipsilateral renal agenesis, also known as Obstructed Hemivagina and Ipsilateral Renal Anomaly (OHVIRA) syndrome [1]. Common clinical features include abdominal pain, dysmenorrhea, and an abdominal mass due to hematocolpos. Management typically involves surgical intervention, including septum resection and drainage of accumulated blood [2]. This report describes a case of HWW in a patient presenting with these characteristic symptoms.

Case Presentation

A 15-year-old girl presented to our gynecological emergency department with worsening right lower abdominal pain persisting over the past six months. She experienced severe colicky pain in the lower right abdomen coinciding with menstruation. Her pain did not radiate and was not associated with fever, vomiting, or urinary symptoms. She had no significant medical or surgical history, aside from her first menstrual period occurring a year prior with regular cycles, dysmenorrhea, and cyclic abdominal pain. Local pharmacy analgesics temporarily alleviated her symptoms. She was born full-term without complications, and there were no familial congenital disease histories.

Upon admission, she was afebrile, with stable vital signs (pulse 74 beats/minute, blood pressure 115/80 mmHg). General physical examination findings were unremarkable, except for a tender mass extending to the umbilicus. She exhibited normal secondary sexual characteristics, and vulvar examination revealed a normal hymen. Rectal examination identified a mass in the Douglas pouch. Further evaluation via ultrasound (figure 1) showed a distended endometrial cavity filled with complex fluid and low-level internal echoes, prompting a provisional diagnosis of uterine didelphys, hematometra, and hematocolpos.

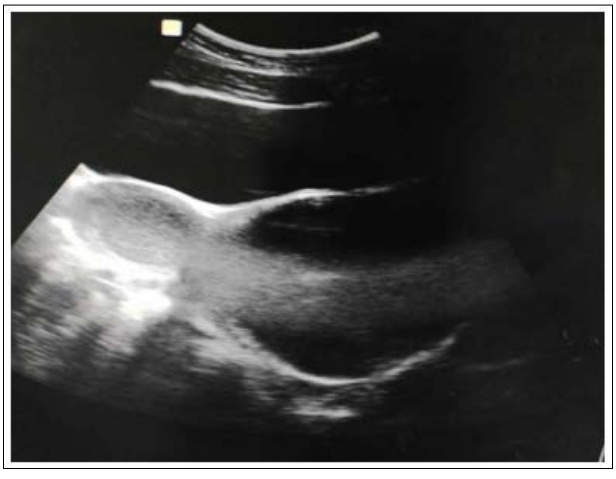

Figure 1: Ultrasound Showed a Distended Endometrial Cavity Filled with Complex Fluid and Low-Level Internal Echoes

The patient was admitted for gynecological management and symptom relief with medications (analgesics, omeprazole, paracetamol) until MRI and planned corrective surgery. Routine investigations indicated normal blood parameters with hemoglobin level at 11.1 g/dl and a white blood cell count of 9000. Pelvic MRI (figure 2) confirmed findings of a bicornuate bicervical

uterus with blind hemivagina, a 15 mm hematoma at the vaginal orifice, hematocolpos measuring 10 cm by 4 cm from the vaginal orifice, and hematometra in the uterine cavity. The left uterine horn and both ovaries appeared normal, while the right kidney was absent. MRI results suggested uterine didelphys with right hematometra due to obstructed hemivagina and ipsilateral renal agenesis (HWW syndrome).

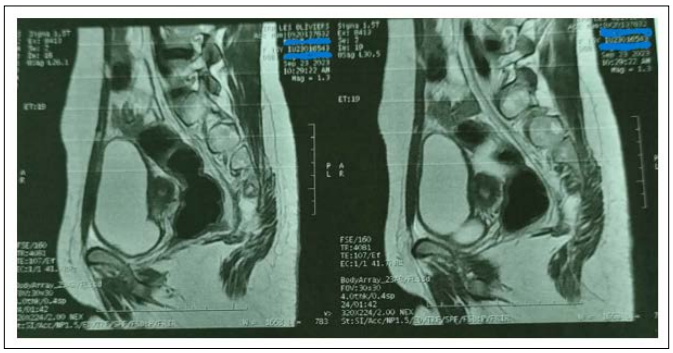

Figure 2: Pelvic MRI

Surgical Intervention Included Identification and Resection of the vaginal septum, extending to the right cervix for drainage of viscous blood. Subsequent vaginal reconstruction was performed without complications, and the patient was discharged two days post-surgery.

Discussion

Herlyn-Werner-Wunderlich Syndrome (HWW), first described in 1971 by Herlyn and Werner, is a rare urogenital anomaly in females characterized by a didelphic uterus, unilateral obstructed hemivagina, and ipsilateral renal agenesis, resulting from combined mesonephric and müllerian duct anomalies [2]. The exact incidence remains uncertain, but estimates range from 0.1% to 3.8% [1]. Patients with HWW syndrome typically remain asymptomatic until puberty. Diagnosis often occurs shortly after menarche, presenting with symptoms such as pelvic pain, dysmenorrhea, and a pelvic mass [3]. Untreated cases may lead to complications including infertility, endometriosis, pelvic adhesions, and rare occurrences of adenocarcinoma or clear cell carcinoma in the obstructed cervix or vagina [4].

Diagnostic imaging, including ultrasound and MRI, is crucial for identifying HWW syndrome due to their non-invasive nature. Ultrasound can reveal a didelphic uterus and pelvic fluid collections with low-level internal echoes contiguous with hematocolpos or pyocolpos. Additionally, features of endometriosis may appear as well-defined cystic masses with diffuse, homogeneous, low-level internal echoes, attributed to retrograde menstruation [5,6].

MRI plays a vital role in characterizing the anatomy, showing iso/high T1W and high T2W signal intensity indicative of pelvic fluid accumulation contiguous with the endocervix, didelphic uterus, and absent kidney on the affected side [1]. Laparoscopy remains the gold standard for diagnosing HWW syndrome, facilitating therapeutic drainage of hematometrocolpos, vaginal septotomy, and marsupialization. Treatment typically involves surgical intervention, such as septoplasty, to relieve obstruction and mitigate the risk of pelvic endometriosis secondary to retrograde menstruation [2].

Conclusion

Bicornuate bicervical uterus with blind hemivagina is a rare congenital malformation causing progressively debilitating pelvic pain and dysmenorrhea. The combination of pelvic ultrasound and MRI remains the diagnostic gold standard. Surgical treatment involves complete resection of the vaginal septum to facilitate continuous drainage of menstrual retention and prevent postoperative fibrosis and vaginal stenosis.

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