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Case Report

Herlyn-Werner-Wunderlich Syndrome: a Case Report

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Abstract

Herlyn-Werner-Wunderlich syndrome (HWWS) is a rare congenital Müllerian anomaly characterized by uterus didelphys, hemivaginal septum, and unilateral renal agenesis. This case involves a 14-year-old girl exhibiting all features of the syndrome who presented with a lower abdominal mass. Despite its rarity, preoperative diagnosis is possible with a strong clinical suspicion and an MRI scan, which is considered the gold standard for diagnosis. Early diagnosis is crucial to prevent complications associated with delayed diagnosis.

Keywords: *Congenital Mullerian Anomaly; Renal Agenesis; Haematocolpos*

Introduction

Herlyn-Werner-Wunderlich syndrome (HWWS) is a rare congenital Müllerian anomaly characterized by uterus didelphys, hemivaginal septum, and unilateral renal agenesis [1].

It is typically observed in adolescents and young women after menarche, presenting with dysmenorrhea, irregular menstruation, abdominal pain, and pelvic mass [2]. Diagnosis can be challenging due to the rarity of the syndrome. A high index of suspicion in patients with renal anomalies and endometriosis is essential for detecting Herlyn-Werner-Wunderlich syndrome to avoid associated complications [3].

A new classification of the syndrome has been proposed based on the presence of complete or incomplete obstruction of the vaginal septum. This new classification allows for earlier diagnosis and treatment. The syndrome is categorized into two classifications based on vaginal obstruction: Classification 1 (completely obstructed hemivagina) and Classification 2 (incompletely obstructed hemivagina) [3].

It is important to recognize this syndrome, especially in cases of hematometra and hematocolpos with left-sided vaginal atresia and urinary tract malformations, to prevent serious complications

Clinical Case

A 14-year-old patient began menstruating at age 12, with regular cycles and dysmenorrhea but no menorrhagia. She was being monitored for iron deficiency anemia, recently discovered with a hemoglobin level of 8 after laboratory tests following a consultation with a general practitioner for fatigue and skin pallor. The patient presented to the pediatric surgical emergency department in Rabat with

right lower abdominal pain and a sensation of fullness in the lower abdomen. Upon examination, a tender mass was noted in the right pelvic region without other associated signs, and the infectious biological workup showed no abnormalities.

An urgent abdominal-pelvic ultrasound was performed, revealing the absence of the left kidney and right nephromegaly measuring 14 cm, as well as a large hematoma in the pelvic area, likely related to hematocolpos , Given the clinical picture and persistent pelvic pain, an MRI was requested.

The left uterine horn was of normal size and morphology, with a large hematoma showing high signal on T1 and intermediate signal on T2, containing some septations consistent with hematocolpos. The right hemi-cavity exhibited a functional endometrium. A serpentine structure in the left lateral uterine region showed high signal on T1 and intermediate signal on T2, with fringing, consistent with hematosalpinx (Figure 1,2).

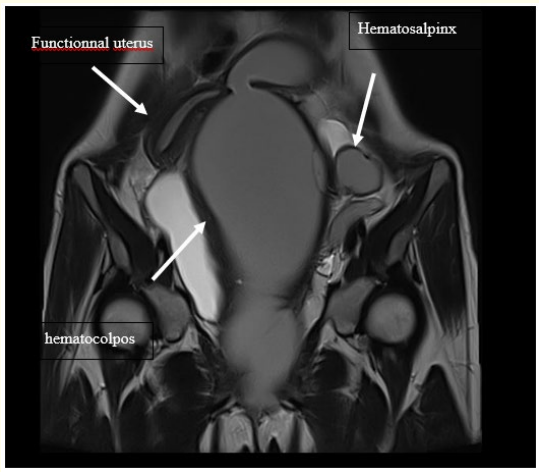

Figure 1: *MRI (T2 coronal): Bicornuate bicervical uterus with a blind hemi-vagina. The left uterine horn is of normal size and morphology, containing hematometra and a large hematoma showing intermediate signal on T2, with some septations consistent with hematocolpos. The right hemi-cavity exhibits a functional endometrium. A serpentine structure in the left lateral uterine region, showing intermediate signal on T2 and maximum thickness, contains fringing, consistent with hematosalpinx.*

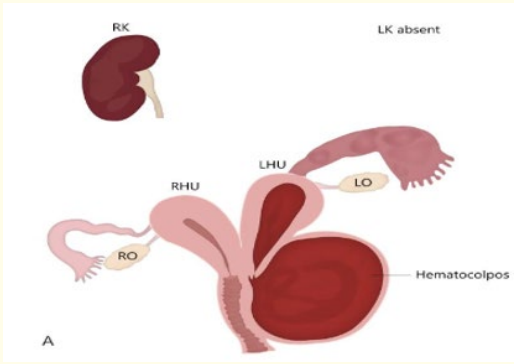

Figure 2: *Drawing of Herlin-Werner-Wunderlich syndrome. (A). HWWS, classification 1.1. with obstructed left hemivagina [4].*

Discussion

The Herlyn-Werner syndrome was initially described by Herlyn and Werner in 1971. This syndrome involves anomalies in the development of the Müllerian ducts, which are embryologically linked to the Wolff or mesonephric ducts, explaining the frequent association with urological anomalies [5].

The clinical outcomes of the two types of syndromes-completely obstructed hemivagina and incompletely obstructed hemivagina-differ significantly. Common symptoms include chronic pelvic pain, dysmenorrhea, and a palpable mass due to hematocolpos or hematoma, as observed in our patient. Less frequently, patients may experience primary amenorrhea, dyspareunia, urinary retention, spontaneous rupture of hematocolpos, infertility, and reproductive issues. Additionally, patients may present with mucopurulent discharge and intermenstrual bleeding if there is communication between the uterine cavities.

In cases with a strong suspicion of Müllerian duct anomalies, an ultrasound should be performed initially to delineate any genital tract abnormalities [6]. In our patient, the ultrasound indicated pelvic hematocolpos, which prompted further investigation with MRI. MRI is the most accurate method for diagnosing these conditions and selecting the most appropriate therapeutic options. It is superior to hysterosalpingography (HSG) and ultrasound for evaluating uterine contours, the shape of the endometrial cavity, and the vaginal septum while also detecting acute and chronic complications [6].

High-resolution T2-weighted imaging (T2WI) should be obtained in three different planes, including the uterine plane, to better understand uterine morphology and characteristics. Occasionally, the non-obstructed hemivagina may be flattened by the hematocolpos, making it difficult to identify. T1-weighted images (T1WI) typically show the intensity of the hemorrhagic fluid signal in the obstructed area in the axial plane. In some instances, this fluid may lose signal intensity from T1WI to T2WI due to the high concentration of proteins and iron from recurrent hemorrhage; this T2 shortening is referred to as the “shading effect.”

Intravenous contrast should not be routinely used but may be helpful in cases of infectious complications or incidental findings. While HSG provides valuable information about the uterine cavity, it may fail to characterize certain uterine subtypes, as non-communicating rudimentary horns are not visualized. In our case, HSG was not performed due to the availability of MRI in our department.

Laparoscopy is considered the most effective method for evaluating reproductive system anomalies [7]. However, it is typically reserved for cases where the diagnosis remains unclear after imaging or when MRI is unavailable and is not performed routinely. The decision to proceed with laparoscopy depends on several factors, including the interval between menarche and diagnosis, the severity of symptoms, and the presence of a hematoma or pyometra [7].

Laparoscopy can also be therapeutic in selected cases, such as the drainage of hematocolpos/hematometrocolpos, septectomy, or marsupialization of the obstructed hemivagina. It is highly effective in reducing pelvic pain and lowering the risk of infection and subsequent hematometra [8].

In our patient, a therapeutic laparoscopy was performed, involving drainage of the hematocolpos and septectomy of the blind hemivagina to prevent recurrence, resulting in significant post-treatment improvement.

Early detection and treatment are crucial for a favorable prognosis and preservation of fertility.

Conclusion

Pediatricians, radiologists, gynecologists, and pediatric surgeons should familiarize themselves with this syndrome, which manifests variably in patients with uterine and vaginal anomalies. Associated renal anomalies should also be considered.

Despite its rarity, a preoperative diagnosis is possible and depends on a high degree of clinical suspicion and the performance of MRI, which is considered the gold standard for diagnosis. It helps clinicians plan, stage the disease, and assess the risk-benefit of different therapeutic approaches. Early intervention is necessary to reduce the risk of endometriosis and infertility.

Bibliography

1. L Karaca., *et al*. "Herlyn-Werner-Wunderlich syndrome: a very rare urogenital anomaly in a teenage girl". *Journal of Emergency Medicine* (2014).
2. Piccinini PS and Doski J. "Herlyn-Werner-Wunderlich syndrome: A case report". *Revista Brasileira de Ginecologia e Obstetrícia* 37.4 (2015): 192-196.
3. Sanjay Mhalasakant Khaladkar., *et al*. "The Herlyn-Werner-Wunderlich Syndrome-A Case Report with Radiological Review". *Polish Journal of Radiology* 81 (2016).
4. Panaitescu AM., *et al*. "Herlyn-Werner-Wunderlich Syndrome: Case Report and Review of the Literature". *Diagnostics* 12.10 (2022): 2466.
5. Zhu L., *et al*. "New classification of Herlyn-Werner-Wunderlich syndrome". *Chinese Medical Journal* 128 (2015): 222-225.
6. Lopes Dias J and Jogo R. "Herlyn-Werner-Wunderlich syndrome: pre- and post-surgical MRI and US findings". *Abdominal Imaging* 40.7 (2015): 2667-2682.
7. Del Vescovo R., *et al*. "Herlyn-Werner-Wunderlich syndrome: MRI findings, radiological guide (two cases and literature review), and differential diagnosis". *BMC Medical Imaging* 12 (2012): 4.
8. Gholoum S., *et al*. "Management and outcome of patients with combined vaginal septum, bifid uterus, and ipsilateral renal agenesis (Herlyn-Werner-Wunderlich syndrome)". *Journal of Pediatric Surgery* 41.5 (2006): 987-992.

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