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Gynatresia in Herlyn-Werner-Wunderlich Syndrome: A Case Report

Valentim Priscila Margarete Araujo Beserra[1]*, Bernardes Jamile Martins[1], Volpini Flavia Resende[1], Alencar Aline Baffa de[2], Pelanda Dandhara Rocha da Silva[2] and Chambô Filho Antônio[3]

1Medical Resident in Gynecology and Obstetrics, Santa Casa de Misericórdia Hospital, Vitória, Espírito Santo, Brazil.

2Undergraduate Medical Student, Santa Casa de Misericórdia Hospital, Vitória, Espírito Santo, Brazil.

3Full Professor and Coordinator of the Medical Residency Program in Obstetrics and Gynecology, Santa Casa de Misericórdia Hospital, Vitória, Espírito Santo, Brazil.

Institute in which the study was conducted: Department of Gynecology, Santa Casa de Misericórdia Hospital, Vitória, Espírito Santo, Brazil.

***Correspondence:**
Priscila Valentim, Medical Resident in Gynecology and Obstetrics, Santa Casa de Misericórdia Hospital, Rua Saul de Navarro 214, Apto. 401, Praia do Canto, 29055360 Vitória, ES, Brazil, Tel: +55 27 99513 5994.

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ABSTRACT

Background: *Female genital malformations are a consequence of abnormalities in the differentiation of the Müllerian ducts at different phases during embryogenesis, resulting in a variety of clinical outcomes. Herlyn-Werner-Wunderlich (HWW) syndrome is a rare congenital anomaly of the female urogenital tract involving the paramesonephric ducts (Müllerian ducts). The syndrome is characterized by the triad of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis.*

Case: *This paper describes the case of a 14-year-old girl presenting with pelvic pain, abdominal swelling and the triad that is characteristic of HWW syndrome. Diagnosis was based on the patient's clinical history and imaging findings. The patient underwent surgery involving complete drainage of a hematocolpos retained in the obstructed vaginal cavity and resection of the vaginal septum. The procedure was successful in providing symptomatic relief. Twelve months following surgery, the patient's menstrual cycles were regular, with no dysmenorrhea.*

Discussion: *Greater understanding of this syndrome is crucial in ensuring that diagnosis is reached at an early stage, thus preventing complications such as retrograde menstruation, endometriosis, infections, adhesions, infertility, and acute or chronic pelvic pain.*

Keywords

Obstructed hemivagina, Herlyn-Werner-Wunderlich syndrome, Müllerian anomalies, Pelvic pain, Renal agenesis, Uterus didelphys.

Introduction

Herlyn-Werner-Wunderlich (HWW) syndrome is a rare, obstructive congenital syndrome that involves anomalies of the paramesonephric ducts (Müllerian ducts), consequently affecting the female urogenital tract. The triad of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis characterizes the anomaly from an anatomical perspective. While the incidence of Müllerian duct anomalies is 2-3%, HWW syndrome corresponds to just 0.16-10% of these Müllerian duct anomalies [1,2].

In the female embryo, absence of the Y chromosome and of functional testis that would otherwise produce the anti-Müllerian hormone (AMH) determines the differentiation of the Müllerian

ducts. These ducts merge to form the uterus, fallopian tubes and the proximal and medial thirds of the vagina. They develop up to the 20th week of intrauterine life, encompassing a succession of events such as growth, fusion, canalization and septal resorption, giving origin to the uterus, fallopian tubes and the upper two-thirds of the vagina. Failures in the growth and fusion phases may result in uterus didelphys, while incomplete septal resorption leads to a hemivagina. Since both the genital and urinary tracts originate from the same embryonic tissue (mesoderm), these developmental failures also affect the renal system, resulting in renal agenesis [3,4].

The syndrome is generally diagnosed at puberty. Symptoms are non-specific and include complaints of acute pelvic pain possibly simulating acute abdomen such as in cases of appendicitis, as well as chronic pelvic pain, dysmenorrhea and a palpable mass in the pelvic region. The mass reflects the presence of hematocolpos or hematometra, which form as a consequence of menstrual blood retained from previous cycles and not exteriorized due to the presence of a vaginal septum [4-6].

Purslow first described the syndrome in 1922 in a young woman of 16 years of age. The patient reported abdominal swelling that increased gradually with each menstrual cycle and was associated with pelvic pain. The pain ultimately progressed to acute abdomen. Laparotomy revealed a double uterus and a pelvic mass caused by distension of the right hemivagina [7].

In 1971, Herlyn and Werner described the simultaneous occurrence of a Gartner's cyst, double uterus, blind hemivagina and ipsilateral renal agenesis. A few years later, in 1976, Wunderlich published a similar case presenting a double uterus, single vagina and hematocolpos on the right-hand side, with no communication between the right hemi-uterus and the vagina. The patient also had renal aplasia and an ipsilateral ureter [8,9].

Timely treatment is pivotal in reducing the risk of complications that include endometriosis due to retrograde menstruation, as well as infections and adhesions that can obstruct the pelvic organs, leading to infertility and acute or chronic pelvic pain [10,11]. Awareness of this syndrome and of the anomalies involved is crucial in ensuring early diagnosis, hence preventing future complications.

Ethical approval

The institution's internal review board evaluated this report under reference CAAE 40543020.0.0000.5065 and approved its submission for publication.

Case

A 14-year-old girl was seen at the Gynecology Department of the *Santa Casa de Misericórdia Hospital* in Vitória, Espírito Santo, Brazil, complaining of pain and increasing abdominal swelling over the preceding two weeks. She had undergone menarche at 12 years of age followed by a history of irregular menstrual cycles with periods of amenorrhea lasting up to three months. She had not yet had her sexual debut, was in use of no medication and had no comorbidities. Physical examination showed the presence of a hard, well-defined and painless abdominal mass extending up to the umbilicus. Pubic hair pattern was typical for the patient's age and there were no lesions in the vulva. A septum of a bluish hue was visualized, bulging and protruding through the hymenal orifice. Speculum examination was avoided to preserve the integrity of the hymen. C-reactive protein (CRP) was 2.1 mg/l, lactate dehydrogenase (LDH) 157 U/l, and beta-hCG negative. Abdominal ultrasonography showed atrophy of the right kidney, topical left kidney with normal morphology and contours, and an expansive, voluminous, predominantly cystic mass with fine debris located within the pelvic region. Computed tomography of the abdomen proved limited insofar as evaluation of the uterus was concerned; however, two lateralized uterine horns were observed, suggesting uterus didelphys (Figure 1A). The uterus was distended suggesting hematometra (Figure 1B) and hydrosalpinx was noted on the right side. The right kidney could not be visualized and the left kidney was normal (Figure 1C). These imaging findings of uterus didelphys, hematometra (on the right side), obstructed hemivagina and ipsilateral agenesis of the right kidney were suggestive of HWW syndrome.

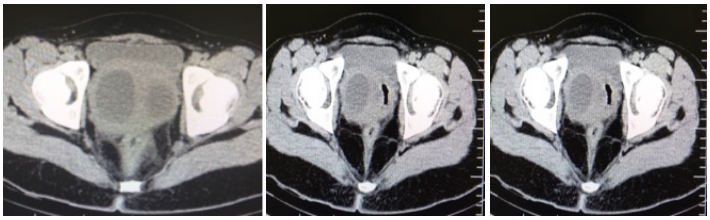

Figure 1: Computed tomography revealing two lateralized uterine horns suggestive of uterus didelphys **(A),** enlargement of the uterus suggesting hematometra **(B)**, as well as a non-visualized right kidney and normal left kidney **(C)**.

The patient underwent a surgical procedure to drain the hematocolpos and hematometra. After identification of the hymen, an incision was made in the vaginal septum and a moderate volume of blood of a chocolate-brown color was drained (Figure 2, A-D). A Foley balloon catheter was placed in the previously made incision to help define the vaginal septum and guide its subsequent resection (Figure 3, A-C).

Pelvic examination confirmed that the right cervix was open and the left closed. The hymen membrane was preserved, and the procedure was considered successful (Figure 4).

The patient was discharged from hospital two days after surgery. Postsurgical gynecological examination showed healing to be satisfactory, with no vaginal wall adhesions. The patient's menstrual cycles became regular, with no dysmenorrhea. Follow-up 12 months after surgery confirmed that the patient's menstrual cycles continued to be regular, with menstruation lasting from three to five days. She reported that she had initiated her sexual life satisfactorily, with no problems and no dyspareunia.

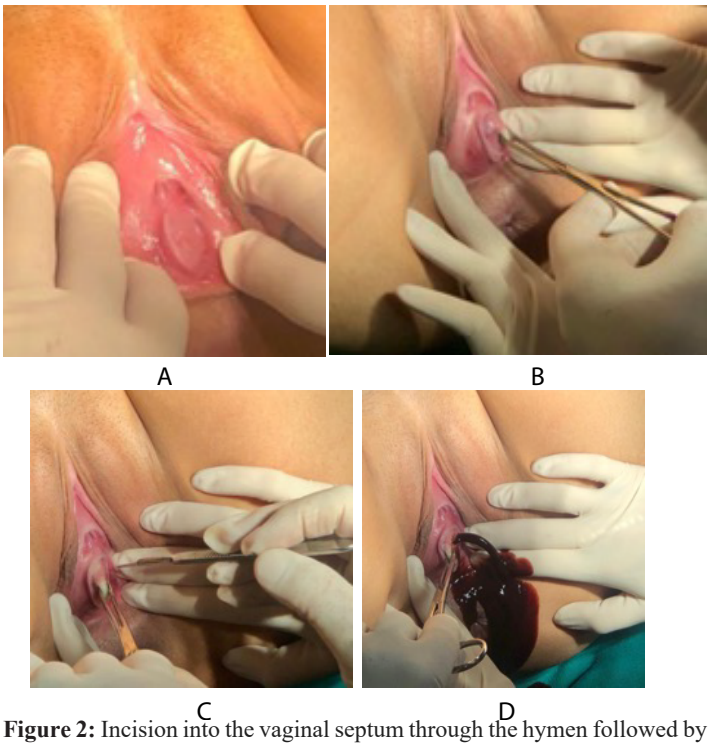

Figure 2: Incision into the vaginal septum through the hymen followed by drainage of hematocolpos **(A-D)**.

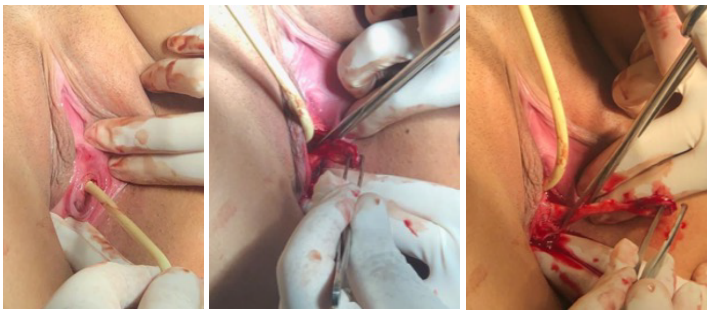

Figure 3: Resection of the vaginal septum following placement of a Foley balloon catheter **(A-C)**.

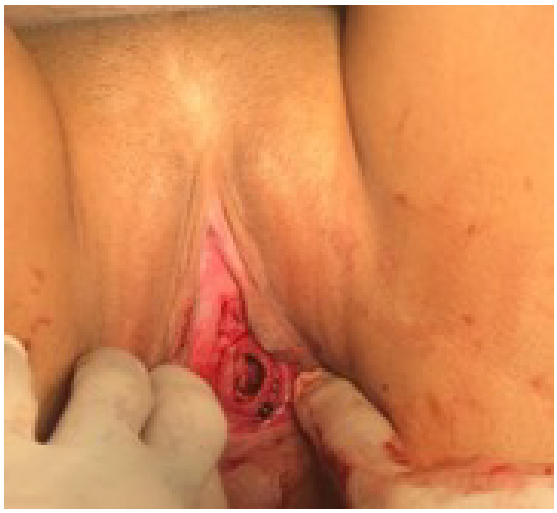

Figure 4: Preservation of the hymen.

Discussion

Patients with HWW syndrome are normally asymptomatic until menarche, when cryptomenorrhea is then triggered. Delays in diagnosing the condition may result in hematocolpos and hematometra, with pelvic pain, irregular menstrual cycles, dysmenorrhea and abdominal swelling [12].

The rare HWW syndrome exerts a significant negative impact on patients' lives; therefore, diagnosing the condition early is vital to enabling the appropriate treatment to be provided. Diagnosis of HWW usually occurs after the patient reaches menarche and menstrual abnormalities start to become apparent. These clinical signs and symptoms range from primary amenorrhea to abnormal menstrual cycles, spotting and cryptomenorrhea. In addition, acute or chronic pelvic pain may be present.

Three-dimensional pelvic ultrasound, computed tomography and magnetic resonance imaging can serve to confirm diagnosis. Excretory urography and magnetic resonance imaging are useful for diagnosing renal abnormalities. Delays in diagnosis may result in a high risk of acute complications such as pyocolpos, pyosalpinx or pelvic peritonitis, and of long-term complications such as chronic pelvic pain, endometriosis, pelvic adhesions and an increased risk of infertility and miscarriage [11,12].

Surgery should be performed as early as possible, with complete incision and marsupialization of the vaginal septum. The objective is to relieve the symptoms caused by distension of the compressed uterine horn and preserve fertility, avoiding immediate and late complications as well as ensuring a normal and satisfactory future sex life [2,5,11].

In conclusion, HWW syndrome is a rare congenital disorder that should be strongly suspected in adolescent girls who develop secondary sexual characteristics and who present with a pelvic mass, renal agenesis, menstrual abnormalities and dysmenorrhea or pelvic pain. Delayed diagnosis is common due to a lack of awareness of this syndrome. Recognizing it is fundamental in ensuring that diagnosis is made at an early stage, thus avoiding complications. With respect to treatment, in addition to the surgical approach the patient's psychological status should be taken into consideration, since the syndrome can significantly impact her reproductive and sexual future.

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