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Successful Gestation of a Herlyn-Werner-Wunderlich Syndrome Carrier And Patient, After a Hysteroscopic Septoplasty

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ABSTRACT

Introduction: Herlyn-Werner-Wunderlich Syndrome (HWWS) is an embryological malformation that damages the urinary and genital systems. It may have clinical repercussions as progressive dysmenorrhea and compromise the patient's obstetric future, with occurrence of severe endometriosis and infertility.

Case Report: A patient, 18 years old, admitted for an amniotic fluid loss complaint. She demonstrated a previous diagnosis of HWWS during anamnesis, which was previously discovered and surgically adjusted 4 years earlier, through a hysteroscopic vaginal septoplasty. The patient needed an obstetric admission due to a clinical diagnosis of Prelabor Rupture of Ovular Membranes (PROM), with abdominal delivery after 5 days due to the development of severe preeclampsia. The procedure had no intercurrence, alongside the birth of the conceptus in good clinical conditions.

Discussion: The patients who carry that rare syndrome have didelphys uterus, low genital obstruction and unilateral renal agenesis. When the genital obstruction is complete, the symptoms are often more glaring, producing palpable and painful abdominal mass because of the hematocolpos. Regardless of the anatomic space, the retained flow is susceptible to infections and may cause abdominal sepsis. Furthermore, the patient can acquire infertility for the higher risk of endometriosis or for the anatomic distortions. The cirurgical adjustment is associated with good pregnancy rates and should always be suggested to those patients.

Conclusion: Considering the serious repercussions to the life quality of those patients, the Herlyn-Werner-Wunderlich Syndrome (HWWS) should always be present in differential diagnosis of a specialist doctor's routine. The premature diagnosis leads to a necessary adjustment and contributes to positive and favorable results in the reproductive life of such patients.

KEYWORDS: malformation, dysmenorrhea, endometriosis infertility

ARTICLE DETAILS

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INTRODUCTION

Herlyn-Werner-Wunderlich syndrome is an embryological malformation, originally described by the triad: uterus didelphys, low genital obstruction and unilateral renal agenesis. It occurs as a consequence of the Muller ducts fusion, responsible for the formation of uterine tubes, the uterus and the vagina superior part (1). Since the external genitalia has embryological origin from the urogenital sinus, its development is regular, which delays the diagnosis. Intense and progressive dysmenorrhea occurs in the cases where the obstruction is complete, alongside palpable abdominal mass in the suprapubic region (secondary to hematocolpos) (3). Premature diagnosis is essential to prevent complications as infertility, endometriosis and recurrent miscarriages. Besides, it guarantees improvement in life quality and sexual function of the afected patients.

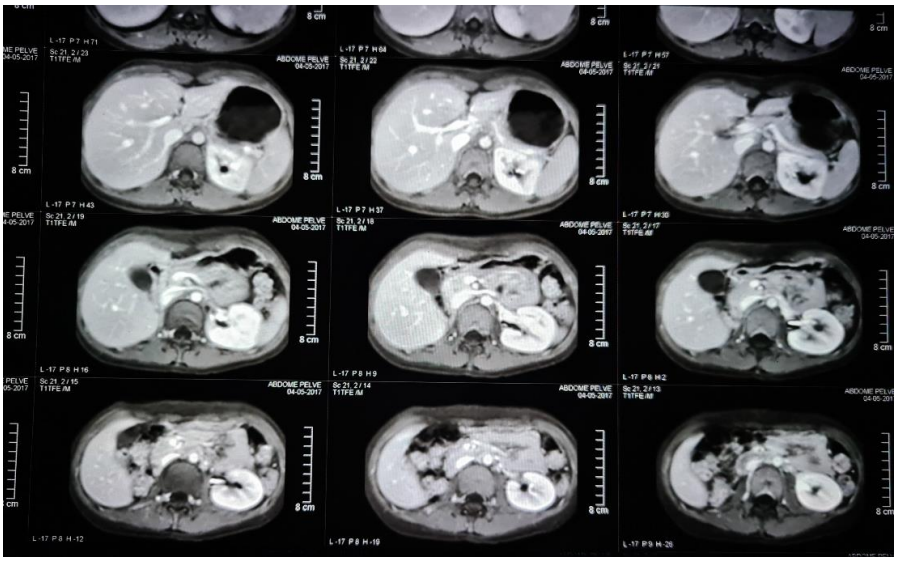
CASE REPORT

18 year-old primiparous patient, treated in the Obstetric Emergency of our service during the gestational age of 34 weeks and 5 days, according to the obstetric ultrasonography of the first trimester. She complained about amniotic fluid

loss, which started on the same day, alongside a lower back pain with irradiation to the lower abdomen and colic. During her admission, the patient informed she was a carrier of Herlyn-Werner-Wunderlich Syndrome (HWWS), having a background of surgical adjustment with hysteroscopic vaginal septoplasty 4 years earlier (at the same hospital service). She had prenatal care at a tertiary hospital without any interference.

Throughout the physical admissional exam, the uterine fundus was observed as lateralized to the left, with an abdomen painless to palpation and absent uterine dynamics. It was detected a clear fluid flowing out of the external cervical orifice after the specular examination. The fetus had good vitality on the cardiotocography exam. The pregnant patient was diagnosed with Prelabor Rupture of Ovular Membranes (PROM) and hospitalized for the clinical condition follow-up, according to the institutional protocol. Antibiotic therapy and steroids were administered for pulmonary maturation, maintaining clinical vigilance on any signs of premature labor and chorioamnionitis. Pregnancy interruption was scheduled for 37 weeks of gestation, in the absence of complications.

The patient informed she had her menarche at 12 years old, with a menstrual flow of small volume. She had a light and progressive dysmenorrhea. She realized a stronger pain after her sexarche (at 14 years old). It was so intense that made her look for an emergency gynecological assistance. The patient was hospitalized at that moment, in our service for clinical condition stabilization. The results of the physical exam, the transvaginal and urine tract ultrasonography as the total abdominal nuclear magnetic resonance (Picture 1) suggested she had HWWS. The patient was submitted to a hysteroscopic vaginal septoplasty, performed without any intercurrence and with the release of a great quantity of pus (pyometrium). She had an improvement afterwards.



PICTURE 1 - Nuclear magnetic resonance sequence on T1 showing only kidney to the left.

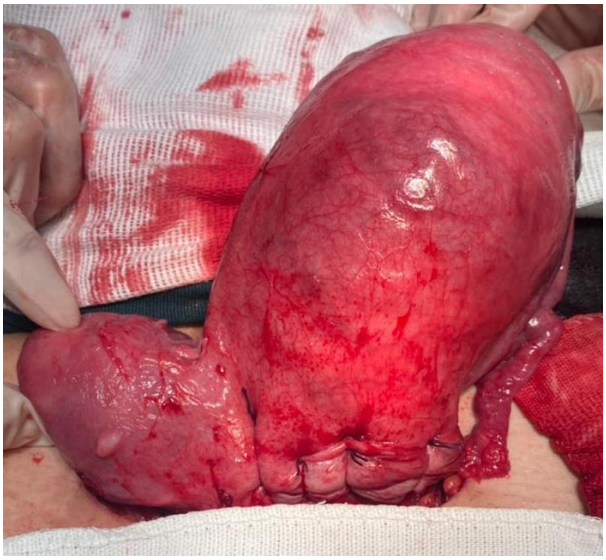
The obstetric ultrasonography demonstrated an only and alive fetus, visible on the breech, weight: 1921 grams, percentile 2, according to the chart of fetal weight of Hadlock (1), top insertion placenta, fundic body and anhydramnios. The fetus demonstrated signs of centralization (brain-placenta rate of 0.87 - p1).

The patient had pressoric elevations and cefaleia complaints (she did not have similar alterations during the prenatal care). Looking for clinical signs of serious conditions, we observed laboratorial changes (Table 01) due to the hypertensive syndrome. An urinary tract ultrasonography was performed, where an only (left side) kidney was observed without any suggestive alteration of renal injury.

Table 1: **Brief of the laboratorial changes during obstetric hospitalization**

DAY	Urea (mg/mL)	Creatinine (mg/mL)
03 °	35	0,8
04 °	35	1,4

Due to the renal comorbidity and the progressive clinical and laboratorial worsening of pre-eclampsia, we decided to induce the labor by abdominal route on the 35th week and 3rd day of gestation, with intraoperative visualization of topic gestation on the left uterine cavity; empty right accessory uterine cavity; bilaterally-present annexes without macroscopic changes (Picture 2). Procedure performed without intercurrence, with the fetus born with good vitality in conditions of joint accommodation.



PICTURE 2 - Intraoperative sight, anteroposterior of didelphys uterus, after fetus withdrawal from the left uterine cavity; rudimental uterine right cavity.

DISCUSSION

HWWS, one of the manifestations of the Müllerian Malformations, is a rare variation that was identified in 1972 for the first time. Its incidence varies from 0,1% to 3,8% in the few cases publicized by the scientific literature (2) (3) (4). It happens due to a fault in the embryological formation, affecting Müller's (paramesonephric) and Wolf's (mesonephric) ducts during the 8th week of gestation (5) (6), approximately. The abnormalities in the urinary and reproductive tracts often coexist due to the very related embryogenesis (7) (8). The renal agenesis is the most common malformation, present in 67% of the cases (8), approximately.

The medical condition is diversified, since little variations of the syndrome may happen (9). In the cases where there is a complete obstruction of the vagina through the septum, the patient presents an intense and progressive dysmenorrhea, with increase of the retrograde menstrual flow (6). Along the months, it emerges a painful and palpable abdominal mass: a consequence of the intravaginal (hematocolpos) and intrauterine (hematometrius) blood accumulation. That can generate urinary retention and constipation, usually 12-18 months after the menarche (10). Besides, alongside the hematocolpos presence, the patient might have fever, shivers, nausea and vomits (10). Regardless of the affected anatomic space, the retain flow is prone to infection, and it can rarely lead to a pelvic inflammation and tubo-ovarian abscess (10).

The diagnosis can be retarded in cases of parcial obstruction, where there is communication between both vaginal spaces (11). Therefore, the patient will have a monthly menstrual exteriorization, with a more discreet pelvic pain, usually treated with contraceptives and non-steroidal anti-inflammatory. That hides the true diagnosis and delays even more the needed therapeutic interventions (12). Besides the use of pharmaceuticals, the degree of septum elasticity (11) (13) also interferes in premature diagnosis.

The chronic pelvic pain is usually a symptom that indicates the patient has HWWS; it may be followed by different stages of endometriosis (14). It happens as a consequence of the increase of retrograde menstruation, secondary to the low obstructive condition. Thus, the endometriosis rate increases in case there is a complete vaginal obstruction (14). About 17,1% of the patients who have HWWS will be diagnosed with endometriosis, which consolidates the percentages of 17% to 35% of endometriosis in patients with just a didelphys uterus (3) (14). When the diagnosis is slow, we can observe serious anatomic distortions and pelvic adherences. Hence, we can notice cases of infertility for many reasons, which enforces the importance of a sooner diagnosis (13) (15).

After the surgical adjustment, the gestation rate is usually high and it should, therefore, be performed as soon as possible to avoid future complications (9). However, only the presence of the didelphys uterus can help us to identify some obstetric complications as: premature labor, anomalous apparitions, ovular membranes, Prelabor Rupture of Ovular Membranes and restrict intrauterine growth (13). Our case denotes exactly the possible obstetric disorders as during the brief hospitalization of the patient we diagnosed every

alteration described previously (patient admitted for PROM, with obstetric ultrasonography highlighting the breech presentation and weight on the percentil 2 of Hadlock). Accordingly, those patients require a stricter obstetric follow-up (16). In our case, the referral to a prenatal care of high risk has become indispensable to a positive obstetric scenario.

CONCLUSION

Considering the diverse and severe complications that endanger the patient's reproductive future and provoke serious maternal issues, the premature diagnoses of the Herlyn-Werner-Wunderlich Syndrome is essential for gynecologists and specialists in human reproduction. Since the surgical treatment has excellent obstetric results and it contributes to the symptoms improvement, knowledge is the key to the patient's quality of life improvement, when they carry that rare syndrome.

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